The Journal of Laryngology & Otology

cambridge.org/jlo

Proceedings of the 155th Semon Club, 11 November 2019, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Secretary: Mr Sherif Haikel, Royal National Throat, Nose and Ear Hospital, University College London Hospitals NHS Trust
Chairpersons: Miss Irumee Pai, Miss Elfy B Chevretton, Guy's and St Thomas' NHS Foundation Trust
Invited panel for pathology: Dr Ann Sandison, Guy's and St Thomas' NHS Foundation Trust
Invited panel for radiology: Dr Steve Connor, Dr Ata Siddiqui, Guy's and St Thomas' NHS Foundation Trust
Section Chairs: Rhinology and Anterior Skull Base: Mr. Shahzada Ahmed, University Hospital Birmingham
Otology and Lateral Skull Base: Mr. Christopher Skilbeck, Guy's and St.Thomas' NHS Foundation Trust
Head And Neck Section: Miss Lisa Pitkin, Royal Surrey County Hospital
Paediatric Section: Mr. Christopher Pepper, Guy's and St.Thomas' NHS Foundation Trust

Online Only Abstract Selection

Cite this article: Haikel S *et al*. Proceedings of the 155th Semon Club, 11 November 2019, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK. *J Laryngol Otol* 2021;E2:1–16. https://doi.org/10.1017/S0022215121001158

The Professor Leslie Michaels prize for the best presentation of the meeting was awarded jointly to Saleh Okhovat for 'A very rare nasal tumour' and Jessica Lunn for 'A rapidly expanding facial mass: the challenges in pathological diagnosis and surgical management'.
Mr Haikel and Miss Chevretton have edited the proceedings of the meeting to reflect the discussion of each case by the expert panel and audience during the Semon Club meeting.

Rhinology and anterior skull base section

Chairperson: Mr Shahzada Ahmed

A very unusual nasal tumour

K A Sadler, S Okhovat, L Melia
From the Queen Elizabeth University Hospital, Glasgow

Introduction

Germ cell tumours are typically found in testicles and ovaries with occasional manifestation in extragonadal midline sites, most commonly the retroperitoneum. We present a case of a man diagnosed with a sinonasal malignancy found to be a primary germ cell tumour.

Case report

A 32-year-old male presented to the emergency department with periorbital cellulitis, headaches and fevers on a background of worsening sinusitis. He was clinically septic, with proptosis, chemosis and restricted extraocular movements. Endoscopic examination identified a nasal mass with mucopurulent discharge.

Radiological findings

Computed tomography and magnetic resonance imaging scans of the sinuses demonstrated a heterogeneous mass centred on the ethmoid air cells, with intracranial extension and frontal lobe oedema. Bilateral intra-orbital extension with indentation on the medial recti was noted. The craniocervical junction and cerebrospinal fluid spaces were unremarkable. Positron emission tomography and ultrasound scans found no evidence of testicular pathology or nodal or distant metastasis.

Histological findings

Microscopy showed extensive infiltration with poorly differentiated tumour cells. Immunohistochemistry staining for octamer binding transcription factor 3/4, placental alkaline phosphatase, Cytokeratin antigen marker (CAM 5.2) and anti pan-cytokeratin antibody cocktail (AE1/3) were positive, favouring a germ cell origin. Biochemically, there was marked elevation of serum beta-human chorionic gonadotropin, with normal lactate dehydrogenase and alpha-fetoprotein levels.

© The Author(s), 2021. Published by Cambridge University Press

Management

The patient completed four cycles of vasoactive intestinal peptide (VIP) chemotherapy (etoposide, ifosfamide and cisplatin) followed by anterior skull base resection of the residual germ cell tumour three weeks after chemotherapy. Locoregional and craniospinal radiotherapy (RT) was initiated to optimise disease control. The patient had a complete response to treatment, with resolution of orbital symptoms, normal tumour markers, and no clinical or radiological signs of recurrence.

Discussion

Multimodality treatment for sinonasal germ cell tumours with VIP chemotherapy, anterior skull base resection and consolidation with craniospinal RT can offer an excellent chance of oncological success and cure.

Conclusion

Early and effective cross-specialty communication was vital in making the diagnosis and initiating therapy in this very rare cancer presentation.

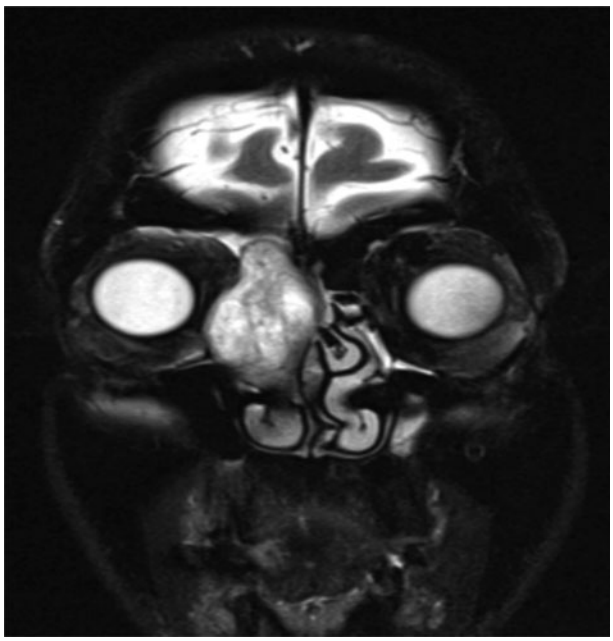

Fig. 1. Pre-operative T2-weighted magnetic resonance imaging scan (coronal plane) showing the tumour measuring 40 × 23 mm and obstructing the frontal sinus drainage pathway with intra-orbital extension and bowing of the nasal septum.

Metal-working fluids exposure and a rare fronto-ethmoid lesion

A Nassimizadeh, S Ahmad, A Sandison
From the Queen Elizabeth Hospital Birmingham

Introduction

Sinonasal neoplasms represent a rare yet histologically diverse group of tumours with aetiology attributed to occupational and social factors. The present case report describes a rare fronto-ethmoid lesion suspected of being cancer.

Case report

A 63-year-old man presented with a 2-year history of right-sided facial discomfort, medial canthal swelling and nasal obstruction. Past medical history was unremarkable, and he worked as an engineer with open machines where he was exposed to metal-working fluids. Nasendoscopy confirmed a mass arising from the right middle meatal opening.

Radiological findings

Magnetic resonance imaging (MRI) showed evidence of a soft tissue mass filling the right anterior ethmoid labyrinth, extending through the frontal sinus ostium down to the middle meatus inferiorly (Figure 1). The lesion enhanced with gadolinium on the T1-weighted MRI scans. There were no aggressive features and diagnosis of a benign right ethmoid sinus tumour was favoured.

Surgery

A right-sided Draf IIc frontal sinus drill out plus a spheno-ethmoidectomy and excision of right-sided nasal tumour was performed, and the sample was sent for histology.

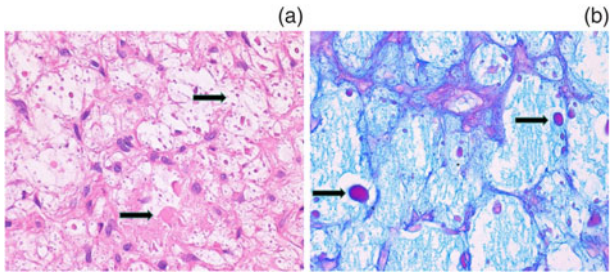

Fig. 2. Histological evaluation of the right-sided nasal tumour with (a) haematoxylin and eosin stain section (Arrows indicate foamy histiocyte top and eosinophilic material bottom), and (b) Alcian blue special stain section (Arrow indicates eosinophilic material). (×400).

Histological findings

There was no marked variation in cellularity and no significant inflammatory cell infiltrate. Dr Sandison demonstrated many foamy pink stained cells that were cluster of differentiation 68 positive macrophage type cells (Figure 2a). There was no atypia, mitosis or necrosis, but there was eosinophilic material of uncertain aetiology. Dr Sandison explained that Alcian blue stains did not show any mucin-producing cells; however, histiocytic pink cells remained present on special staining (Figure 2b). Dr Sandison stressed the rarity of this case and concluded that because of the absence of overt malignant features, a reactive granulomatous lesion to a foreign body was the favoured diagnosis. Mr Ahmed added that based on the patient's clinical history, the reaction was most likely to be to metal-working fluids, and he should be discharged with advice on refraining from close contacts with these oils.

Discussion

Mr Ahmed and Dr Sandison commented on the complex nature of the histology, which reinforced the importance of a multidisciplinary approach in the management of these

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

patients. Mr Ahmed mentioned the importance of second opinions in rare or unusual cases. In this case, the original and subsequent pathology reports differed significantly. Dr Sandison also advised the patient to undergo a chest X-ray to assess any lung involvement given the inflammatory response seen in the sino-nasal cavities.

Unilateral serosanguinous nasal discharge for one year

C W Lee, S Di Palma, S Derbyshire
From the Royal Surrey NHS Foundation Trust

Introduction

Human papillomavirus (HPV)-related multiphenotypic sinonasal carcinoma is a rare tumour reported to present as a large, destructive sinonasal mass with high-grade histological features but paradoxically behaving in an indolent manner.

Case report

A 57-year-old male presented to the emergency department with a 1-week history of frontal headache and a background of a 1-year history of unilateral (left-side) serosanguinous nasal discharge. A soft tissue mass was identified within the left nasal cavity on nasendoscopy. Under anaesthetic, the patient underwent an examination of the nose and endoscopic debulking of the nasal tumour with unresectable residual disease in the nasopharynx.

Radiological findings

Magnetic resonance imaging (MRI) of the head and computed tomography (CT) scan of the sinuses showed an expansile lesion within the left nasal cavity with complete opacification of the left paranasal sinuses. No bony destruction was seen. Positron emission tomography-CT imaging of the whole body showed no evidence of metastases.

Histological findings

Nasal biopsy showed a histologically malignant tumour with areas of necrosis and mitoses. The tumour was composed of basal type round blue cells arranged in cribriform, lobular and solid growth patterns with some glandular structures. Immunohistochemistry showed strong positivity for pool of keratins (AE1/AE3) and p16. Molecular analysis failed to demonstrate MYB gene re-arrangement leading to a diagnosis of HPV-related multiphenotypic sinonasal carcinoma. This was consistent with an HPV-related malignant tumour with epithelial features, and not an adenoid cystic carcinoma.

Management

The patient received a course of radical radiotherapy to the nasopharynx.

Discussion

Dr Connor and Mr Ahmed emphasised the importance of pre- and post-operative MRI in the management of sinonasal carcinoma. The MRI scan allows staging, detection of intracranial spread and exclusion of lesions such as meningoceles that mimic intranasal lesions. Mr Ahmed noted that on CT imaging this lesion was deemed unresectable, but MRI imaging is necessary to confirm this. Dr Ahmed asked if subtyping of these tumours was possible. Dr Sandison said it was possible but difficult. She mentioned that although HPV positivity was a good prognostic indicator for oral cancers, the situation for sinonasal cancers remains unknown. Mr Ahmed said there was some evidence that the Gardicil® HPV vaccine may reduce laryngeal papillomatosis. Miss Chevretton suggested that the 9 valent vaccine (currently only available privately in the UK) potentially may be more appropriate at reducing nasal lesions when compared to the 4 valent vaccine that is currently available to vaccinate teenagers.

Conclusion

With a vast array of different possible types of tumours of the nose, paranasal sinuses and skull base with widely differing prognosis, distinguishing histology was essential in formulating the best management plan for the patient.

Severe nasal destruction including recurrent epistaxis, rhinorrhoea, crusting and facial pain following cocaine use

R Phillips, M King, J A McGilligan
From the Brighton and Sussex University Hospitals NHS Trust

Introduction

Cocaine is a common illicit drug associated with cocaine-induced midline destructive lesions and levamisole-induced vasculitis. In cocaine-induced midline destructive lesions, cocaine causes severe destruction of the osteocartilaginous structures of the nasopharynx. Levamisole-adulterated cocaine is associated with systemic vasculitis presenting with rash, constitutional symptoms and varying degrees of oto-laryngological involvement.

Case report

A 36-year-old gentleman presented with recurrent epistaxis, nasal crusting, offensive rhinorrhoea and mid-facial pain. He developed arthralgia, fevers, sweats, weight loss and lymphadenopathy. Examination showed a large nasal septal perforation and a likely livedo reticularis rash on his left thigh. Urine was positive for cocaine and levamisole.

Radiological findings

Dr Connor commented that the computed tomography scan showed a destructive midline nasal process with involvement of the nasal septum, turbinates and medial maxillary antral walls. The palate was intact.

Histological findings

Biopsies showed a non-specific inflammatory lesion with areas of necrosis. There was no arteritis or giant cell reaction. Dr Sandison noted no evidence of malignancy or granulomata. Earlier biopsies had been suspicious for immunoglobulin

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

(Ig) G4-related disease because of increased IgG4 positive plasma cells. Dr Sandison explained that levamisole is an anti-helminthic agent that causes vasculitis and granulocytosis, and it was taken off the market in 2000.

Management

Abstinence was encouraged. The patient was given steroids initially but did not tolerate methotrexate or mycophenolate. He sustained a good response to rituximab. Debridement gave lasting symptomatic relief.

Discussion

Mr Ahmed highlighted the importance of proving abstinence before embarking on immunosuppressive therapy. The short half-life of levamisole and cocaine make proving their use difficult.

Conclusion

In midline nasal destruction, cocaine and levamisole use must be considered. As abstinence is the mainstay of treatment, and attempts should be made to ensure this prior to starting immunosuppression. The literature suggests renal or pulmonary involvement may be indications for immunosuppression.

Otology and lateral skull base section

Chairperson: Mr Christopher Skilbeck

Is it all in my head? An uncommon cause of hearing loss

J Bastianpillai, E Warner, M Wareing

From the Royal London Hospital, Barts Health NHS Trust

Introduction

Temporal bone tumours are rare entities and can present insidiously. Neoplasm should be excluded in unilateral sensorineural hearing loss.

Case report

A 41-year-old lady presented with left-sided moderate mixed hearing loss, worsening over six-months with no other symptoms. Otoscopy showed a dull left tympanic membrane. The patient's hearing improved to mild sensorineural hearing loss in the following six months but deteriorated to a severe hearing loss a year later, associated with a bulging tympanic membrane. Blood results were unremarkable aside from a mildly raised parathyroid hormone.

Radiological findings

Dr Connor demonstrated extensive petromastoid air on the initial magnetic resonance imaging scan which is suggestive of left apical petrositis. Later, computed tomography and magnetic resonance imaging (MRI) scans showed a destructive low-attenuating soft-tissue density in the left petrous bone involving the middle-ear cavity with a cochlear fistula and no pathological enhancement. The ossicles were intact. The lesion was poorly enhancing on the fat-saturated post-gadolinium MRI scans.

Histological findings

There were prominent neural bundles surrounded by a giant cell lesion. The bland multinucleate giant cells (cluster of differentiation 68 positive) are in keeping with a giant-cell tumour (Figure 3). However, a second opinion described an osteoclast-rich lesion, and immunostaining showed a lack of G34W expression (amino-acid substitution), virtually excluding giant-cell tumour.

Management

Management consisted of subtotal surgical resection of the lesion post-biopsy.

Discussion

Though initially thought to be a temporal bone giant-cell tumour, this was later excluded given the absence of G34W mutation. No specific diagnosis beyond an osteoclast-rich lesion had been found; however, Dr Sandison highlighted that a small

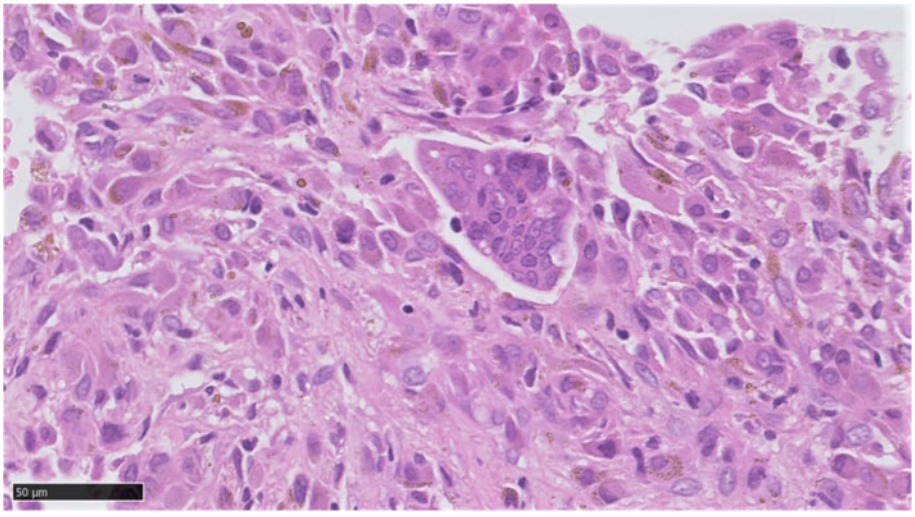

Fig. 3. Histological evaluation of the left middle ear and temporal bone biopsy demonstrating a large osteoclast-type multinucleate giant cell (H&E; ×40).

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

number of giant-cell tumours do not express this mutation, and it may be worth considering further opinions.

Conclusion

Our case highlights the importance of serial imaging if the clinical situation changes and that although radiology and histology are important, a diagnosis is not always straightforward.

A painful, destructive lesion of the temporal bone

A Vinoo, G Potter, E Stapleton
From the Manchester Royal Infirmary

Introduction

There has been a rise of necrotising otitis externa cases in recent years. We describe an elderly patient presenting with a painful discharging ear. The patient was initially diagnosed with necrotising otitis externa, but her pain persisted despite treatment. She turned out to have a different diagnosis, therefore demonstrating the importance of considering differential diagnoses when initial treatment fails.

Case report

A 79-year-old lady presented with a 6 month history of an extremely painful, discharging ear, on a background of childhood mastoid surgery, dead ear and grade 3 facial palsy.

Radiological findings

Computed tomography demonstrated an otic destructive lesion of the left temporal bone, likely representing an infective process with osteomyelitis of the labyrinth. Dr Connor demonstrated calcification in the middle ear, some spongiosis, fistulation of the semicircular canals and calcific obliteration of the cochlea.

Magnetic resonance imaging demonstrated enhancing material in the mastoid cavity, and avid enhancement of the cochlea, labyrinth and internal auditory meatus.

Histological findings

Biopsy of the ear canal granulations showed granulation tissue. The temporal bone biopsy showed cholesteatoma with foreign body giant cell response.

Management

Initially diagnosed with necrotising otitis externa after biopsy of ear canal granulations, the patient commenced systemic antibiotics. The pain did not settle, and open biopsy of the enhancing material in the mastoid showed cholesteatoma. Canal wall down mastoidectomy with blind sac closure was performed, relieving the severe otalgia.

Discussion

Cholesteatoma rarely presents with severe pain and extensive destruction, which the panel agreed on. Dr Connor pointed out that radiologically this was primarily a chronic middle-ear process that spread medially to involve the inner ears. Necrotising otitis externa is uncommon in non-diabetic patients.

Headaches, fevers and deficit of a second language

W Flynn, H Zhang, J Ahmed
From the Royal London Hospital, Barts Health NHS Trust

Introduction

Isolated clival tuberculosis without involvement of the craniovertebral junction is rarely reported and presents a diagnostic challenge.

Case report

A 30-year-old male banker from India presented with fevers, headaches and having lost the ability to speak English, which was his second language. He had lost 12 kg of weight in the preceding 4 months. He was pyrexial, tachycardic and confused. Neurological examination was otherwise unremarkable.

Radiological findings

Dr Connor demonstrated a large area of abnormality with an epicentre at the clivus and extension into the sphenoid sinus and petrous apices on magnetic resonance imaging (Figures 4 and 5). He described associated disease of the leptomeninges, dura, 3rd and 5th cranial nerves, and the basilar artery. Computed tomography depicted calcified mediastinal, hilar and mesenteric lymph nodes suggesting previous granulomatous disease.

Management

Anti-tubercular chemotherapy was initiated empirically with corticosteroids. Endoscopy allowed biopsy of a friable lesion attached to the inter-sphenoid septum. Histological findings showed non-specific granulomatous changes. Acid-fast bacilli were isolated from cerebrospinal fluid 35 days after initial lumbar puncture. Progressive communicating hydrocephalus developed, requiring

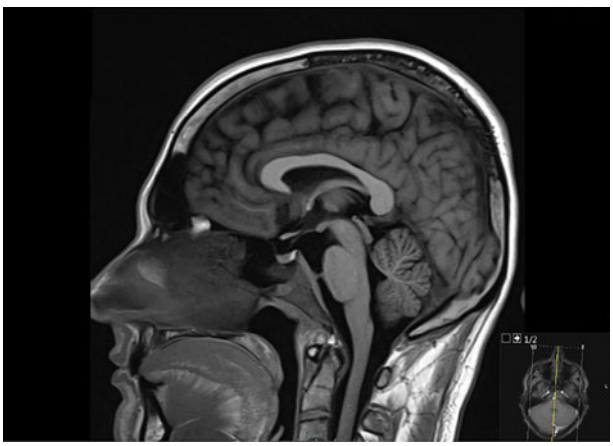

Fig. 4. Sagittal T1-weighted turbo inversion recovery magnitude magnetic resonance imaging of the head. The image depicts an abnormal low T1 signal in the clivus and pathological tissue stranding the prepontine cistern.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

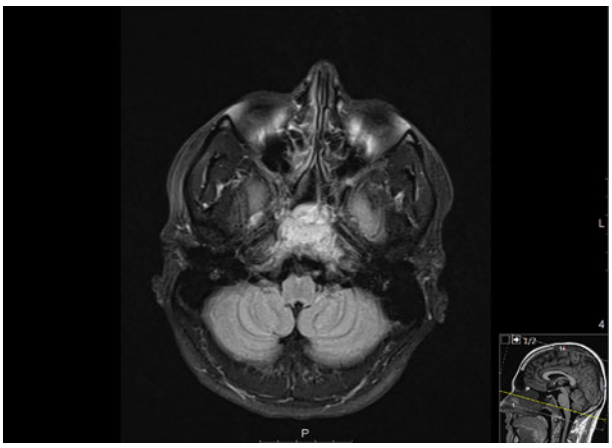

Fig. 5. Transverse T2-weighted turbo inversion recovery magnitude magnetic resonance imaging of the head. The image shows a large abnormality centred on the clivus and extending into the sphenoid sinus.

insertion of a ventriculoperitoneal shunt. Symptoms completely resolved, and the patient was discharged after 10 weeks.

Discussion

Dr Connor noted that a fossa navicularis resulting from notochord remnants could serve as a route of infectious spread from the nasopharynx to the clivus in children. However, no evidence of this could be seen on imaging in this case. Dr Connor advised further imaging only if there was clinical deterioration.

Conclusion

Tuberculous osteitis of the clivus can cause potentially lethal complications including meningitis, hydrocephalus and infectious vasculitis; however, appropriate and timely management can lead to a good outcome.

A chronic discharging ear concealing a complex diagnosis

K Hamlett, E Stapleton, S Freeman
From the Salford Royal Infirmary

Introduction

Chronic ear discharge is a common presentation. During surgery for chronic otitis media, it is advisable to send samples for histology, especially in the presence of unusual findings.

Case report

A 70-year-old man underwent a combined approach tympanoplasty for chronic otitis media in 2015. In 2017, he underwent revision surgery for recurrent ear discharge and a middle-ear polyp. There was extensive disease throughout the middle ear and mastoid, with dense adherence to the facial nerve. He re-presented in 2019, coughing up blood. Flexible nasal endoscopy showed a post-nasal space mass.

Radiological findings

Computed tomography of petrous bones in 2015 suggested chronic otitis media without evidence of bony erosion. Repeat imaging in 2017 showed an enhancing lesion in the left middle-ear cavity, with features suspicious of facial nerve schwannoma or paraganglioma. Magnetic resonance imaging in 2019 confirmed rapid advancement of inoperable disease into the middle cranial fossa and post-nasal space mass (Figure 6).

Histological findings

Histopathology in 2017 indicated neuroendocrine adenoma with a Ki-67 index of 25 per cent (Figure 7). In 2019, a post-nasal space mass biopsy yielded 'recurrent neuroendocrine adenoma' with a Ki-67 index of 50 per cent, suggesting malignancy. A third opinion concluded a likely sinonasal

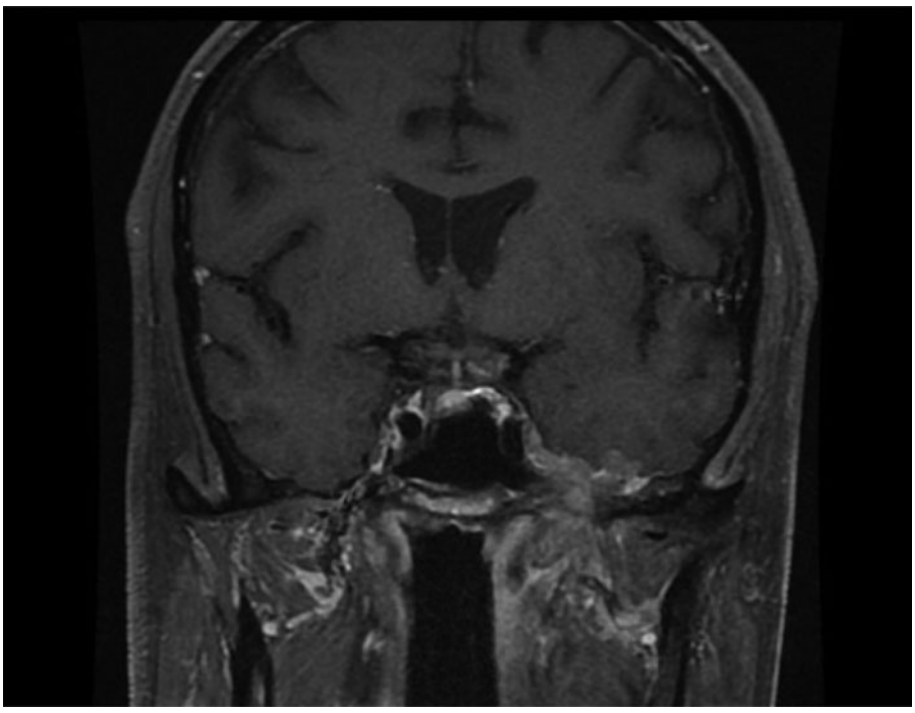

Fig. 6. Coronal T1-weighted MRI image of recurrent disease showing left middle fossa involvement, dural enhancement and nodular involvement into brain parenchyma.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

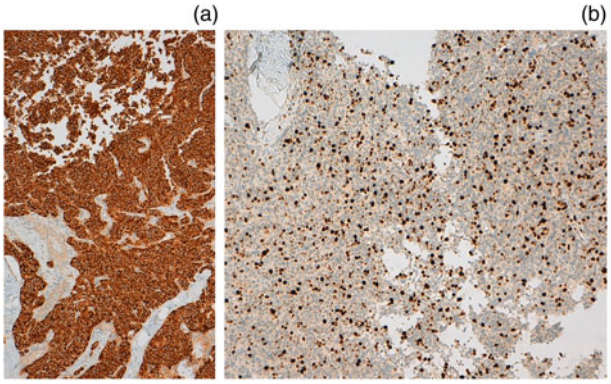

Fig. 7. Histopathology from revision canal wall down tympanomastoidectomy with (a) synaptophysin staining and (b) ki-67 staining (×50).

neuroendocrine carcinoma extended from the tip of the Eustachian tube into the middle ear.

Management

The skull base multidisciplinary team (MDT) advised palliative chemo-radiotherapy because of unresectable disease.

Conclusion

This case demonstrated the importance of careful histological analysis and MDT relationships in the management of cases in which unexpected malignant transformation occurs. Dr Sandison agreed there were early histological signs of malignancy. Miss Pai highlighted the importance of sending histology samples in routine surgery for chronic otitis media. Miss Stapleton and Mr Skilbeck said they would correlate all clinical findings before deciding on the need for sending histology samples.

Recurrent left ear infections with episodic left facial weakness, taste disturbance and imbalance

C L Dalton, A Kay, R Irving

From the Queen Elizabeth Hospital, Birmingham

Introduction

Middle-ear adenomatous neuroendocrine tumours are rare, slow-growing adenomas, on a continuum with carcinoid tumours.

Case report

A 53-year-old female presented with recurrent left ear infections and an episode of left-side facial weakness, taste disturbance and imbalance. Otoscopy showed a tympanic soft tissue mass extending into the attic.

Radiological findings

Imaging showed a middle-ear mass involving the canal, anterior epitympanum and ossicles. After initial surgery, abnormal enhancement of the left geniculate ganglion extending along the tympanic facial nerve indicated disease progression. Subsequent imaging demonstrated spread to the parotid gland and finally to the middle fossa floor and medial temporal lobe.

Histological findings

Dr Sandison demonstrated the trabecular and glandular architecture typical of middle-ear adenoma, including the epithelial nature and neuroendocrine differentiation seen on immunostaining (Figures 8 and 9). She then outlined tumour spread to the parotid gland, lymph nodes, geniculate ganglion and dura.

Management

Initial combined-approach tympanoplasty was intended to eradicate the middle-ear adenoma. Recurrence presenting as episodic facial nerve paralysis led to subtotal petrosectomy and vertical facial nerve exploration. Fluctuating facial paralysis continued, with ensuing left parotid enlargement. Left total petrosectomy, parotidectomy, and extra-cranial facial nerve resection and reanimation were performed. Subsequent skull base recurrence required a middle fossa resection followed by radiotherapy. The patient is now radiologically clear of disease.

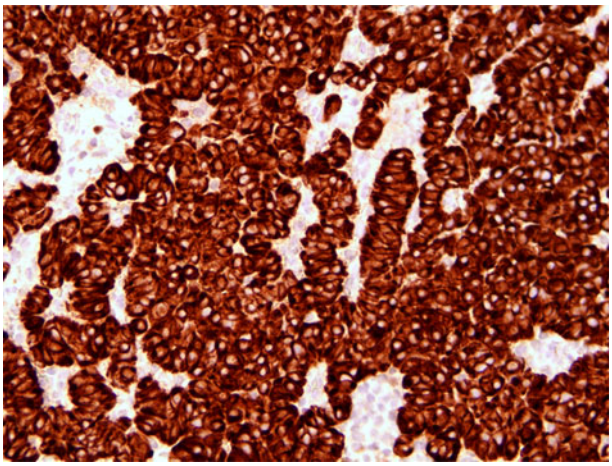

Fig. 8. High power magnification of cytokeratin immunostaining of the middle-ear tumour specimen showing its trabecular architecture and confirming its epithelial nature. (Cytokeratin immunostain; ×20).

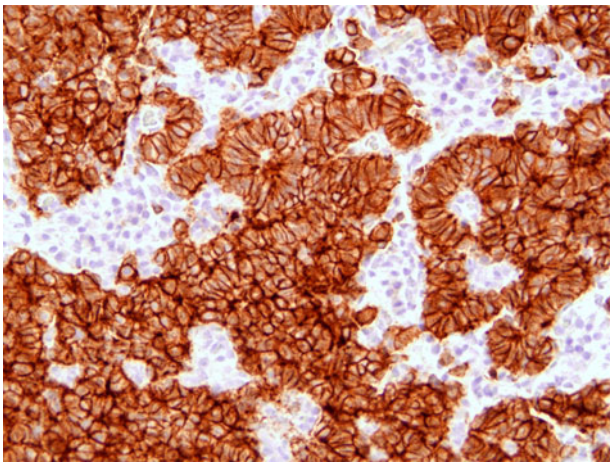

Fig. 9. High power magnification of cluster of differentiation 56 immunostaining of the middle-ear tumour specimen showing its trabecular architecture and supporting a neuroendocrine differentiation. (CD56 Immunostain; ×20).

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

Conclusion

Locally aggressive disease and late recurrences are uncommon in middle-ear adenomatous neuroendocrine tumours but can pose diagnostic and management challenges, as demonstrated here. A multidisciplinary approach and vigilant, lifelong follow-up are paramount.

A unilateral 6th nerve palsy secondary to a rare skull base lesion

J Lunn, M Jaafar, S Gane

From the Royal National Throat, Nose and Ear Hospital, London

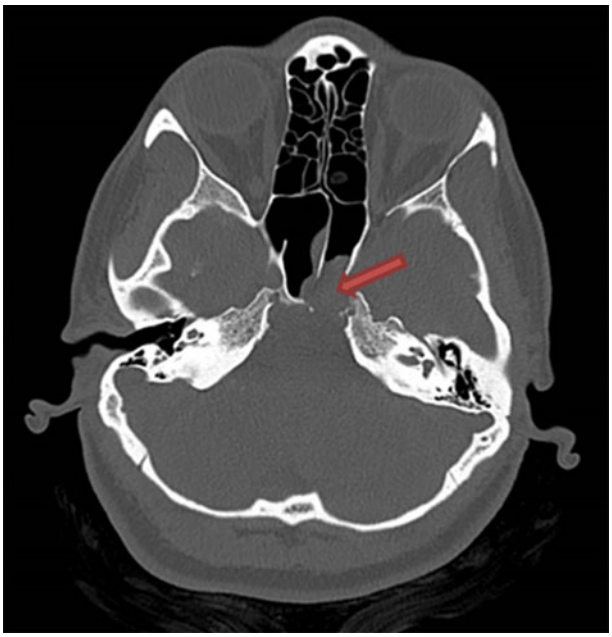

Fig. 10. Axial plane computed tomography scan of paranasal sinuses (lesion demonstrated by arrow).

Introduction

Giant cell tumours of bone are locally aggressive benign tumours predominantly occurring in long bones in early adulthood. We describe the rare case of a skull base giant cell tumour of bone presenting as an abducens nerve palsy.

Case report

A fit and well 30-year-old presented with a 4-week history of a progressive left-sided headache and diplopia. Examination showed an isolated abducens nerve palsy. Imaging and right sphenoidotomy with biopsy were performed.

Radiological findings

Dr Connor commented that the lesion appeared to be slow growing and originating from the clivus. He also noted there was bony erosion over the carotid artery and highlighted the importance of excluding an aneurysm (Figure 10).

Histological findings

Microscopy showed spindle cells and scattered multinucleated giant cells (Figure 11). Dr Sandison noted that these morphological findings have a wide range of differential diagnoses, and therefore immunostaining was required to diagnose an H3G34W-mutant giant cell tumour of bone (G34W refers to a mutated site of the histone variant H3.3 and is characteristic of this tumour).

Management

Following discussion in skull base and sarcoma multidisciplinary team meetings, the patient underwent endoscopic resection followed by radiotherapy.

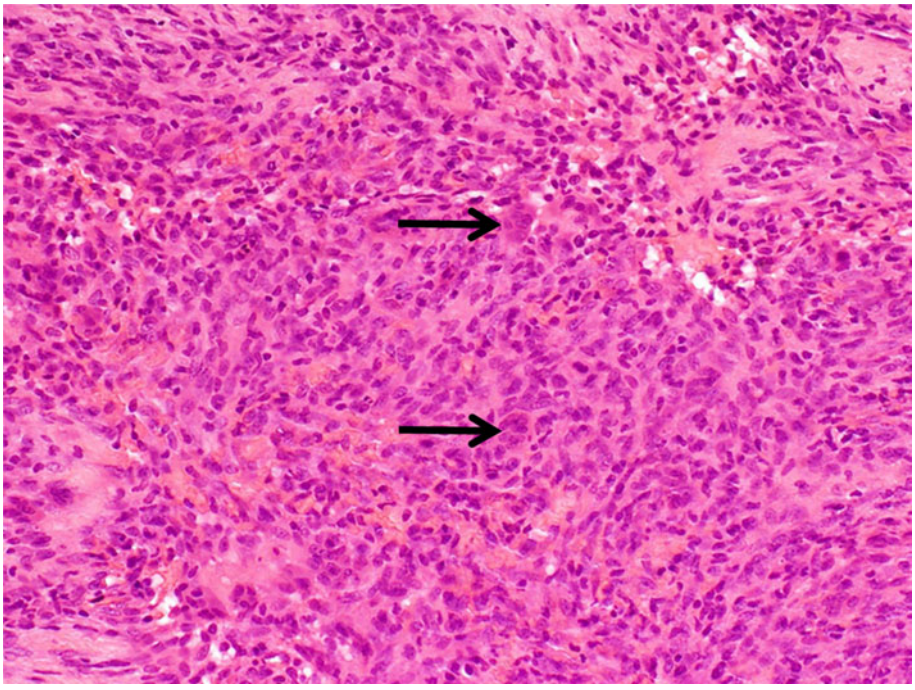

Fig. 11. Histology slide with arrows showing multinucleated giant cells. (H&E; ×10).

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

Discussion

The roles of adjuvant radiotherapy and monoclonal antibodies were discussed. Dr Sandison advised that although these lesions are generally radiosensitive, disease progression and radiation-induced sarcoma are potential sequelae in children. Radiotherapy would also make revision surgery more difficult if disease re-occurred.

Conclusion

Skull base giant cell tumours of bone are rare. Surgical excision is the treatment of choice but provides challenges because of the proximity of neurovascular structures.

Head and neck section

Chairperson: Miss Lisa Pitkin

Stridor in a 79-year-old man who lived with a 'sick old bird'

R Grounds, N Chaston, J D Wasson
From the East Kent Hospital University NHS Trust

Case report

The patient presented as an ENT out-patient with hoarseness and following examination was listed for urgent microlaryngoscopy. No overt malignancy was found, but the appearance of the vocal folds and subglottis was noted to be highly irregular. Biopsies were taken, but the histology returned only squamous papilloma. The patient had recently also been referred to the respiratory team with shortness of breath, with a known history of chronic obstructive pulmonary disease, and had started on a course of oral steroids. Of note, his pet parrot had recently died despite being treated by the vet for aspergillosis. The patient re-presented to accident and emergency three weeks later with biphasic stridor, was listed for urgent repeat laryngo-bronchoscopy and found to have copious creamy white debris obstructing the airway (Figure 12, intra-operative photograph). Further biopsies and microbiology swabs were taken, and the patient was intubated and taken to the intensive care unit but sadly died seven days later from respiratory failure due to aspergillosis.

Microbiology and histology

The second biopsies taken demonstrated spindle cell squamous cell carcinoma. The swabs cultured *Aspergillus fumigatus*. It is likely that the patient had an evolving carcinoma, but whether this was driven by, or simply colonised by aspergillus is not clear.

Discussion

Aspergillus epiglottitis is a rare but recognised disease that is usually terminal. It usually occurs in those with concomitant malignancy or immunocompromised status.

Conclusion

Where highly atypical lesions are seen, in addition to biopsy, early discussion with microbiology about treatment with antifungal agents should be considered.

Dyspnoea, dysphagia and dysphonia in a 66-year-old female

S Linton, A Vinoo, N Mani
From the Manchester Royal Infirmary

Introduction

Craniocervical paragangliomas are uncommon neoplasms arising from extra adrenal paraganglia cells. The three common craniocervical paragangliomas are: carotid body tumours,

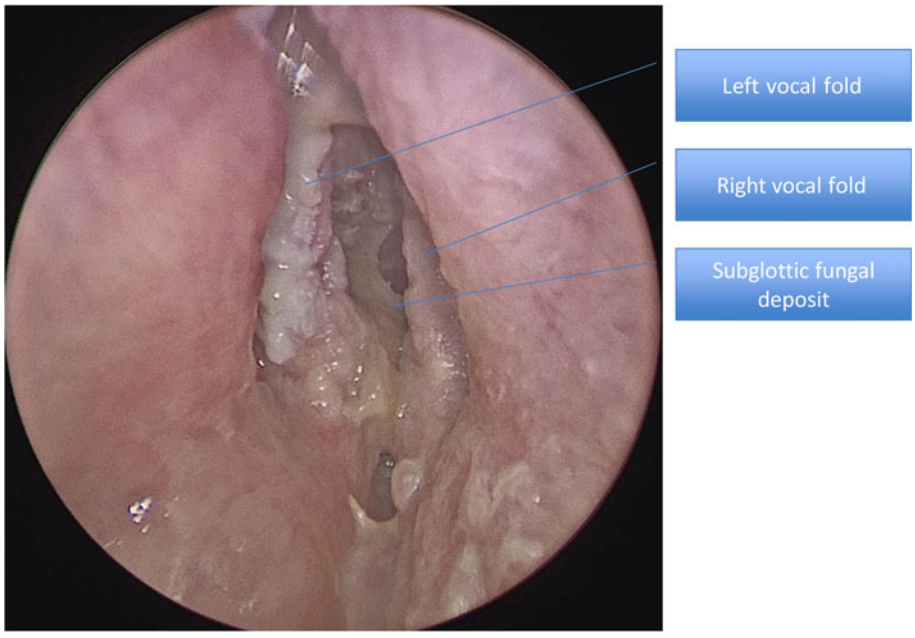

Fig. 12. Intra-operative photograph showing creamy white debris obstructing the airway.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

jugulotympanic paragangliomas and vagal paragangliomas. Symptoms and signs are due to mass effect (cranial nerve deficit, dysphagia, dysphonia) or catecholamine production (headaches, diaphoresis and palpitations). Diagnosis includes history, examination, laboratory investigations (presence of excess catecholamines) and imaging (magnetic resonance imaging (MRI), computed tomography (CT) scan or octreotide scintigraphy). Treatment is either conservative management, surgery or radiotherapy.

Case report

A 66-year-old female presented with acute dyspnoea, dysphagia and dysphonia. Examination showed a large right parapharyngeal swelling extending inferiorly into the neck. There was no associated lymphadenopathy. Flexible nasendoscopy showed the mass narrowing the supraglottic inlet; however, the larynx appeared normal. Cranial nerve examination showed a right hypoglossal nerve palsy. Urinary and serum catecholamine levels were normal.

Radiological findings

The MRI and CT neck scans showed a 5.5 × 5.3 × 3.3 cm right neck mass arising from the post-styloid parapharyngeal space, partly encasing and displacing the internal carotid artery anterolaterally and compressing the hypoglossal nerve proximal to its exit from the hypoglossal canal. There was no intracranial extension (Figure 13). A positron emission tomography scan showed no other abnormalities. Dr Siddiqui demonstrated fatty atrophy of the tongue as a result of the denervation effects of the hypoglossal nerve palsy.

Histological findings

Pathology demonstrated tumour cells positive for cluster of differentiation 56, synaptophysin, chromogranin with surrounding sustentacular cells positive for S100. These morphological and immunohistochemical features were in keeping with a vagal paraganglioma.

Management

On the basis of airway compromise and cranial nerve involvement, the patient underwent surgical resection. A combined approach via a lip-split mandibulotomy was used for access and safe control of the internal carotid artery and internal jugular vein. She had an expected high vagal nerve palsy postoperatively, resulting in voice weakness and dysphagia. She recovered well but required a feeding gastrostomy postoperatively because of unsafe swallow.

Discussion

The panel reviewed the management of cervical paraganglioma (i.e. conservative approach, surgical resection or radiotherapy). They emphasised the importance of testing for catecholamine production ahead of any surgical excision. Additionally, the panel highlighted that small, benign, well medialised, pre-styloid parapharyngeal lesions may be excised using transoral robotic surgery. This lesion, given its size, location and intimate relationship to the internal carotid artery, was therefore not suitable for such an approach.

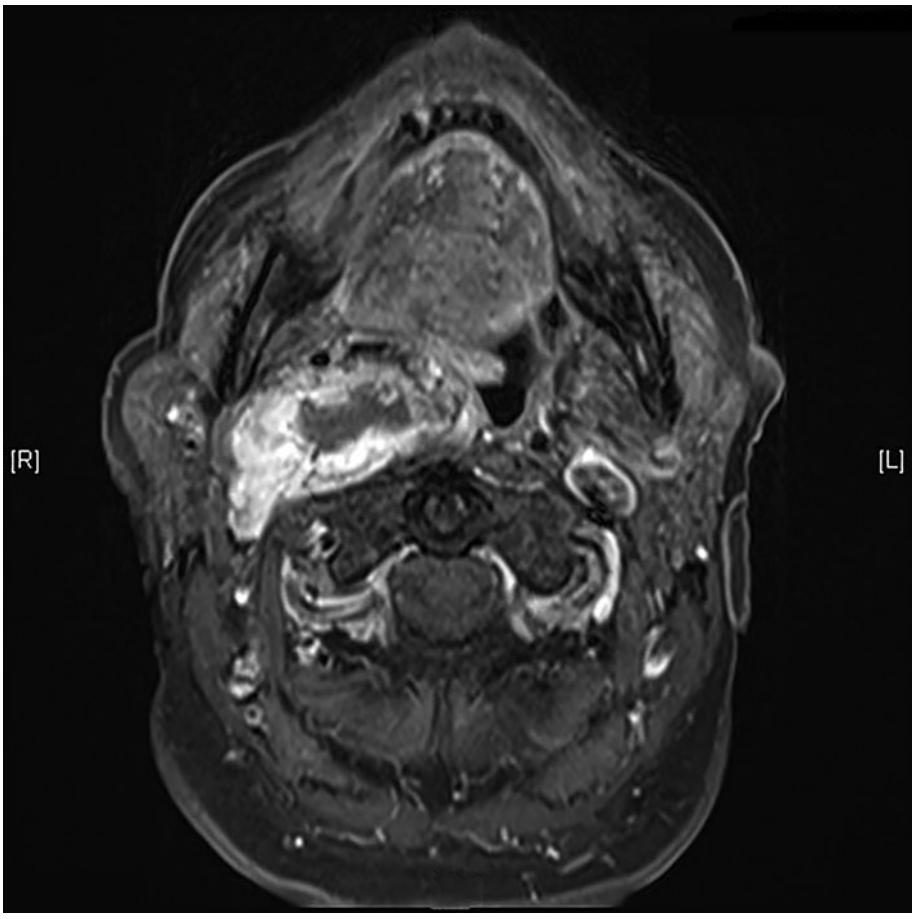

Fig. 13. Magnetic resonance imaging of the neck showing a large right vagal paraganglioma (T1-weighted post-contrast fat suppressed axial cut). R = right; L = left.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

Recurrent neck and facial swelling in a laryngectomy patient

G Wellstead, B Wong, S Burrows
From the Norfolk and Norwich University Hospital

Introduction

Cutaneous spread from head and neck squamous cell carcinoma (SCC) is extremely rare (less than 1 per cent). We report a case of multiple cutaneous metastases in a patient with pyriform fossa and floor of mouth primary disease.

Case report

A 57-year-old male diagnosed with T_2 N_2b squamous cell carcinoma (SCC) of the left pyriform fossa and synchronous T_1 SCC of the anterior floor of mouth was treated with surgery (laryngectomy, bilateral neck dissections, pectoralis major flap reconstruction and excision of floor of mouth lesion) followed by radiotherapy.

Two years after, he presented several times with acute head and neck swelling. Despite extensive investigation, no cause was found. He gained limited relief with steroid and diuretic treatment. However, eight months later he developed cutaneous lesions along his laryngectomy scar, which were biopsied.

Radiological findings

Multiple computerised tomography (CT) and magnetic resonance imaging (MRI) scans showed progressive lymphoedema of the neck and face in keeping with post-radiotherapy changes.

Histological findings

Biopsies taken from cutaneous lesions showed SCC, in keeping with cutaneous lymphatic spread from the original tumour.

Management

The patient is currently undergoing palliative chemotherapy. His lymphoedema has settled with this.

Discussion

This was an atypical presentation for skin metastasis from head and neck SCC. Imaging (CT and MRI) were performed to exclude recurrence, metastasis, internal jugular vein thrombosis or lymphatic obstruction, but the panel felt that a positron emission tomography scan would be a better modality.

Conclusion

Although rare, cutaneous metastasis from head and neck SCC should be considered as a differential diagnosis when initial investigations do not identify a cause.

An unusual case of stridor

D Chudek, I Wilson, A Kinshuck
From the Liverpool Head and Neck Centre, Aintree University Hospital

Introduction

We present a case of a 78-year-old female with shortness of breath and stridor who was found to have a subglottic mass of unusual histology.

Case report

The patient presented to the ENT clinic with a 5-month history of worsening shortness of breath and stridor. Her past medical history included rheumatoid arthritis and systemic lupus erythematosus. Nasoendoscopy showed fully mobile vocal folds with an irregular, narrowed subglottis. She underwent endoscopic airway assessment and a smooth, firm subglottic mass arising from the left lateral wall of the trachea was found.

Radiological findings

The CT and MRI neck scans showed focal thickening in the subglottic region bulging into the laryngeal lumen and measuring roughly 4 mm with a non-sinister appearance.

Histological findings

Histologically, the lesion stained positively with Congo red and demonstrated an apple green birefringence when viewed through crossed polaroids, confirming the diagnosis of localised amyloid.

Management

The mass was resected with carbon dioxide laser and sent for histology, and 40 mg of methylprednisolone acetate was injected into the subglottis and submucosa.

Discussion

Amyloidosis is a rare disease, with head and neck involvement being even more unusual. However, if present, it is usually found above the level of true vocal folds and presents with dysphonia. Therefore this patient's presentation with stridor and the anatomical location of the deposit is particularly unusual. Localised amyloidosis is treated by surgical resection, which can be repeated as the deposit re-accumulates.

Conclusion

At two months post-operatively, our patient's stridor had not returned, and her exercise tolerance improved. She will be followed up with regular larynx surveillance to monitor for deposit re-accumulation.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

An unusual source of a neck lump: colorectal cancer metastasis to a cervical lymph node

O J Wright, A Bashyam, L Pitkin
From the Frimley Park hospital

Introduction

Colorectal cancer is the third most common cancer worldwide; approximately 20 per cent of patients already have metastases at time of diagnosis. The most common metastatic sites for colorectal cancer are liver, lung, bone, brain and peritoneum. Cervical lymph node metastases in colorectal cancer are rare and associated with a poor prognosis.

Case report

A 73-year-old female patient presented with a rapidly enlarging left level IV neck lump, four years after a right hemicolectomy for a T_4N_2MO poorly differentiated caecal adenocarcinoma.

Radiological findings

Whole-body positron emission tomography-computed tomography showed elevated tracer uptake in two left level IV lymph nodes and no evidence of further metastatic disease.

Histological findings

Dr Sandison demonstrated a glandular tumour on histology which could have originated from a sinus or lung primary tumour. Subsequent immunohistochemistry demonstrated a colonic primary.

Management

The patient went on to have a level II, III and IV neck dissection with histopathological examination and immunohistochemical staining showing metastatic caecal adenocarcinoma. At two-month review, the patient was disease free and had recovered well from surgery.

Discussion

Dr Sandison highlighted the utility of immunohistochemical analysis in confirming the primary site and suggested the addition of prostate cancer to the list of infraclavicular malignancies that can metastasise to cervical lymph nodes.

Conclusion

This case describes an unusual pattern of disease spread. Although most metastatic lymphadenopathy originates from head and neck primaries, clinicians should always consider the possibility of more remote disease and not underestimate the importance of a detailed past medical history and systems review.

A rare cause of a hoarse voice

R Vasanthan, S Khosla, D Walker
From the Royal Surrey County Hospital NHS Foundation Trust

Introduction

Most laryngeal tumours are malignant, and the majority of benign laryngeal tumours are papillomas. This is a case of an oncocytic papillary cystadenoma of the larynx, a rare benign minor salivary gland tumour.

Case report

A 51-year-old male presented with a hoarse voice, with a significant smoking and alcohol history. Initial flexible nasendoscopy showed a large left vocal fold tumour.

Radiological findings

Computed tomography and magnetic resonance imaging (MRI) confirmed a $23 \times 16 \times 30$ mm tumour of the left vocal fold, extending to the supraglottis, with an associated supraglottic laryngocoele. Dr Siddiqui demonstrated the lesion with cystic features on MRI (Figure 14).

Histological findings

Dr Sandison highlighted the key histological features of the specimens from the larynx, with a cystic lesion lined by oncocytic cells with papillary architecture and concluded that this was a good illustration of oncocytic papillary cystadenoma of minor salivary gland origin (Figure 15).

Management

Biopsies were obtained via endoscopically assisted suspension laryngoscopy. The multidisciplinary team advised conservative management due to the benign condition, and the potential impact on quality of life from definitive management. The importance of lifestyle changes was emphasised to the patient particularly with regards to cessation of smoking. Interval scanning and clinical monitoring continue.

Discussion

Miss Pitkin asked whether smoking cessation may lead to regression of the lesion. Dr Sandison felt that it probably would not.

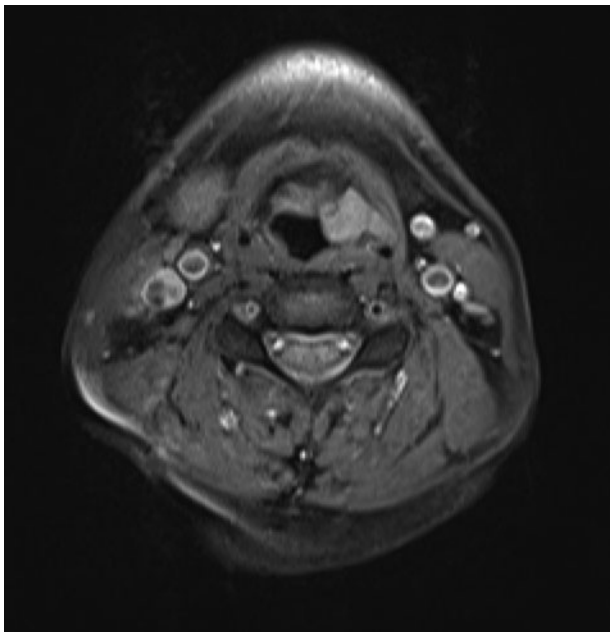

Fig. 14. Magnetic resonance imaging of the neck in the axial plane showing a left side cystic laryngeal lesion.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

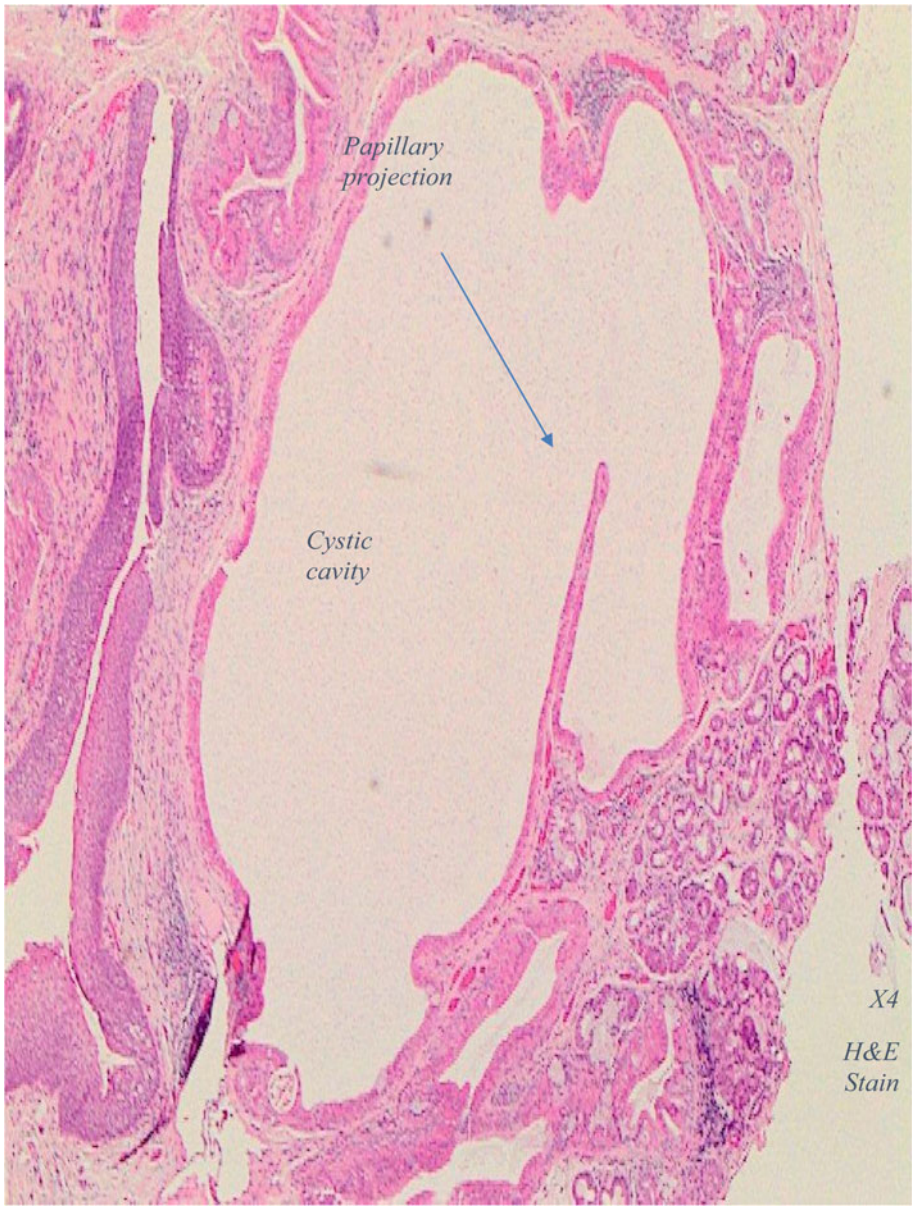

Fig. 15. Histological evaluation of cystic lesion lined by oncocytic cells with papillary architecture from laryngeal specimen. (H&E; ×4).

Conclusion

Oncocytic papillary cystadenoma of the larynx is a rare benign tumour, which can be managed conservatively or with surgical excision.

Paediatric section

Chairperson: Mr Christopher Pepper

An unusual case of paediatric acute sinusitis

J French, J Rudd, M Felton
From the Evelina Children's Hospital, London

Introduction

We present a case of paediatric acute sinusitis that was resistant to initial treatment and proved a challenging diagnosis.

Case report

A 14-year-old female was admitted following partially treated sinusitis after six-weeks of intermittent fever, otalgia and rhinorrhoea. Deterioration despite broad-spectrum intravenous antibiotics and intra-nasal topical medications prompted re-assessment, establishing symptoms of polyarthralgia, weight loss and haemoptysis. Examination included oral and nasal ulcers, blood-stained nasal discharge, bilateral tympanic membrane perforations and vesicular skin lesions. Inflammatory markers and antineutrophil cytoplasmic antibodies were persistently elevated. Haemoptysis and acute renal failure prompted intensive care admission.

Radiological findings

Computed tomography of the head and sinuses showed left-sided sinusitis with the possibility of infected contents within the left maxillary antrum (Figure 16). Dr Siddiqui identified subtle bone thickening suggestive of chronic changes. Both chest X-ray and computed tomography of the chest showed consolidation consistent with pulmonary haemorrhage, suggestive of pulmonary vasculitis.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

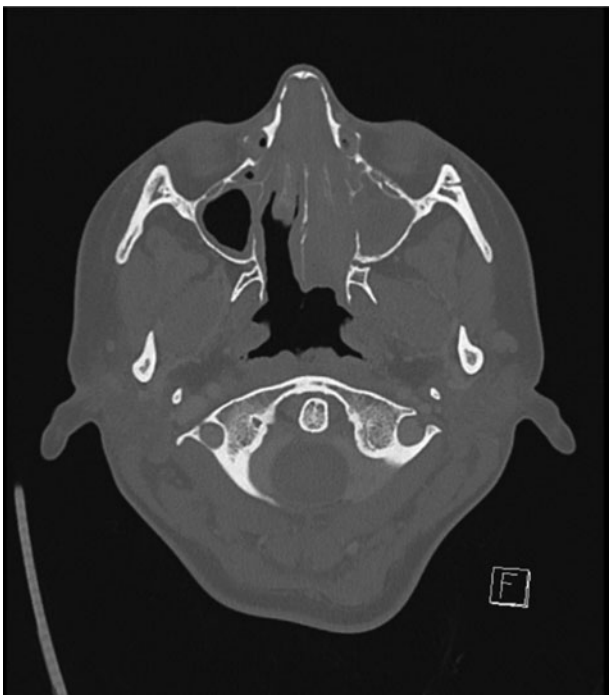

Fig. 16. Axial plane computed tomography scan of the sinuses showing left maxillary sinusitis.

Histological findings

The diagnosis of granulomatosis with polyangiitis was confirmed on renal biopsy, which demonstrated 50 per cent segmental sclerosing lesions, acute changes on healthy background kidney. Nasal biopsy showed inflammatory changes.

Management

A protracted hospital admission involving the paediatric intensive care unit included treatment with rituximab, cyclophosphamide, prednisolone and intra-nasal corticosteroids.

Discussion

Discussion of this case included the potential value of antral washout in this scenario and also prompt re-evaluation of the differential diagnosis.

Conclusion

Although granulomatosis with polyangiitis is difficult to differentiate from infectious aetiologies, early recognition is imperative to optimise outcomes and minimise critical organ involvement. This case highlights the importance of a thorough history and broad differential diagnosis.

A rapidly expanding facial mass: the challenges in pathological diagnosis and surgical management

J Lunn, K Karamali, H Daya

From the St George's Hospital, London

Introduction

Myxoid fibroblastic-type tumours are soft tissue tumours that can be further sub-classified to predict their behaviour. This varies significantly from benign but locally aggressive lesions to malignant lesions with metastatic potential.

Case report

A fit and well two-year-old girl presented with a three-month history of a rapidly enlarging left facial swelling, epiphora and increased snoring. Physical examination showed a left sided facial mass extending from her medial canthus and lateral nasal wall to her midface and inferomedial orbit.

Radiological findings

Computed tomography (CT) and magnetic resonance imaging (MRI) showed a well-defined mass centred on the inner canthus of the left eye, extending into the nasal cavity and expanding the nasolacrimal duct with evidence of local destruction and remodelling of the nasal cavity (Figure 17).

Histological findings

The final histology showed a locally aggressive neoplasm of primitive myxoid fibroblastic type that was difficult to classify but showed no malignant behaviour.

Management

The patient underwent complete surgical excision via midfacial degloving with sub-ciliary approach.

Discussion

This is a rare tumour type particularly in the head and neck of children. The difficulties with histological diagnosis and sub-

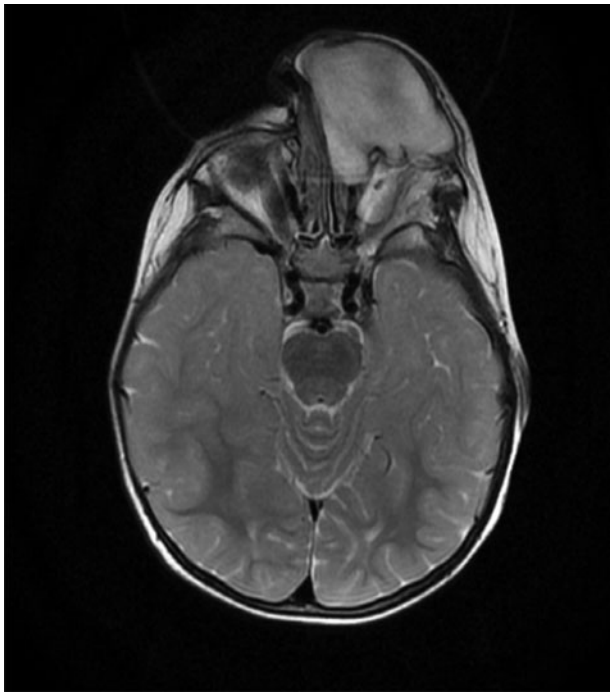

Fig. 17. Axial plane magnetic resonance imaging (T2-weighted post-contrast) of face.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

typing were emphasised by Dr Sandison. She questioned whether we initially considered more common soft tissue tumours such as rhabdomyosarcoma. Dr Siddiqui emphasised the importance of both CT and MRI to assess the extent of sinonasal tumours.

Conclusion

Morphological and clinical overlap between different myxoid fibroblastic-type tumours can generate difficulties with accurate diagnosis and therefore prediction of tumour behaviour. A careful specialist and multidisciplinary approach to the decisions about management is required.

Paediatric posterior cervical mass

J Collins, J Rudd, C Pepper

From the Evelina Children's Hospital, Guy's and St Thomas Foundation Trust

Introduction

Cervical neck masses are common in children, with the majority of cases being benign. Anatomical location, time frame and other symptoms can help discriminate between diagnoses. Here, we discuss a case with an unusual presentation given the final diagnosis.

Case report

A four-year-old boy was referred to a tertiary service with a nine-month history of increasing posterior neck mass. He was previously treated with antibiotics with no effect. He had no prominent local or systemic symptoms and was otherwise well. He had a normal birth and developmental history. He was fully vaccinated, with no foreign travel history or tuberculosis contacts. Examination showed a 3 cm firm, non-tender, non-fluctuating posterior triangle lymph node, with no overlying skin changes and no other lymphadenopathy.

Radiological findings

Dr Siddiqui reported the radiology as being non-specific. Computed tomography and ultrasound both showed an enlarged soft tissue mass, likely a lymph node, with central necrosis. There were no other enlarged lymph nodes or metastases.

Histological findings

Dr Sandison also reported non-specific findings of granulomatous inflammation, including multi-nucleated giant cells with no malignant cells.

Management

Excisional biopsy showed a deep, matted, necrotic lymph node with frank pus. Microbiological culture showed *Mycobacterium avium*.

Discussion

Mr Pepper noted despite extensive investigation there was still an unclear diagnosis prior to surgery. Given the location of the mass, it is a surprising diagnosis of non-tuberculous mycobacterium. This usually presents in cervicofacial nodes, with prominent skin changes and often fistula formation. Dr Sandison commented that it was fortuitous to gain a diagnosis as often cases do not have positive microbiological samples. Surgical excision is curative in most cases.

Ex-utero intrapartum treatment procedure for congenital airway obstruction from an oropharyngeal mass

S McKenna, J Rudd, C Pepper

From the Evelina Children's Hospital, Guy's and St Thomas' NHS Foundation Trust, London

Introduction

A rare case of congenital airway obstruction managed with ex-utero intrapartum treatment procedure.

Case report

A 38-year-old underwent pre-natal ultrasound assessment identifying a foetal facial mass. Magnetic resonance imaging (MRI) at 31 and 33 weeks demonstrated growth of the lesion from 48 to 68 mm. Ex-utero intra-partum treatment procedure was planned, and intubation was possible whilst on maternal circulation despite the oropharyngeal tumour expanding through the mouth. Subsequent tracheostomy and MRI scan preceded debulking at one week.

Radiological findings

The MRI at one week of age showed a 9.3 cm solid multi-cystic mass filling the oral cavity and extending through the mouth with deep, high para-nasopharyngeal and left lateral skull base involvement (Figure 18).

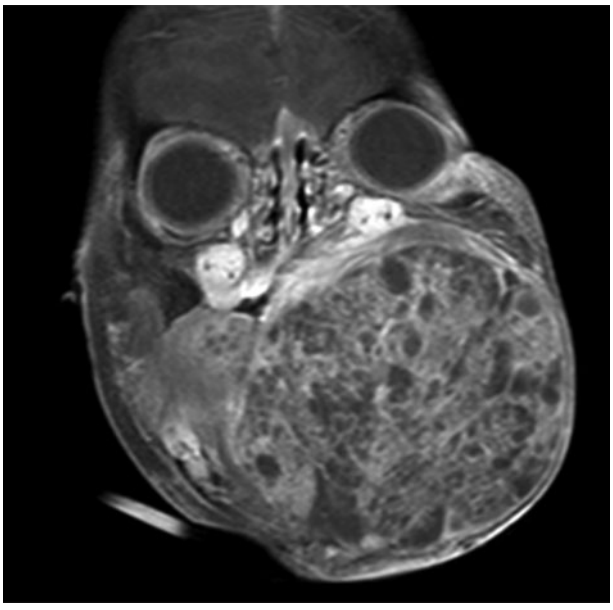

Fig. 18. Coronal T1-weighted magnetic resonance imaging of the head showing a cystic oral mass.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press

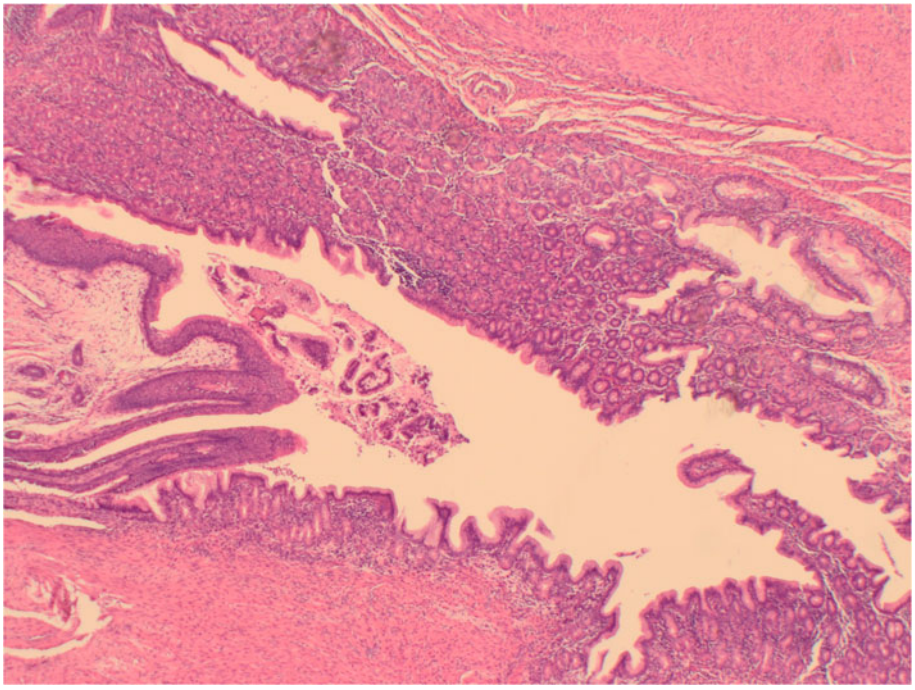

Fig. 19. Histopathology showing mature teratoma with evidence of skin, cartilage, bone, skeletal and smooth muscle, glandular tissue (respiratory and gastric), and several areas of immature neuronal elements. (H&E; ×5).

Histological findings

Teratoma with mature tissues and immature neuronal elements with areas of ulceration and haemorrhage were identified (Figure 19).

Management

Further debridement was performed at five months to limit mandibular expansion. The patient was developmentally normal with tracheostomy and gastrostomy but facial deformity improves with growth. Consideration has been given for future mandibular fixation.

Discussion

Mortality is high without the ex-utero intrapartum treatment procedure in the management of such cases, and planned delivery should consider the risk of spontaneous labour. Appropriate counselling regarding travel distance from the treating unit is therefore important. Improved data collection on ex-utero intrapartum treatment procedures nationally would help inform a more joined-up service across the country.

Conclusion

Prenatal imaging, multi-disciplinary team assessment and meticulous planning are essential for caesarean section and ex-utero intrapartum treatment procedure.

https://doi.org/10.1017/S0022215121001158 Published online by Cambridge University Press