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ONLINE ONLY ABSTRACT SELECTION

Proceedings of the 142nd Semon Club, 10 November 2011, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Chairman: Miss Elfy B Chevretton, Guy's and St Thomas' NHS Foundation Trust

Secretary: Mr Sherif Haikel, Guy's and St Thomas' NHS Foundation Trust

Invited panel for pathology: Dr Ann Sandison, University College London

Invited panel for radiology: Dr Steve Connor and Dr Ata Siddiqui, Guy's and St Thomas' NHS Foundation Trust

The Professor Leslie Michaels prize for the best presentation of the meeting was awarded to K Varadharajan for 'A Patient with VIth and VIIth Cranial Nerve Palsy'

Otology section

Chairman: Mr Rupert Obholzer

A rare mass in the middle ear

C Abbassi, R Natt, J Tahery

From the Countess of Chester Hospital, Chester

Introduction

Neuro-endocrine tumours of the middle ear are rare and often present with nonspecific symptoms.

Case report

A 36-year-old woman presented with an 18-month history of hearing loss and right otalgia. Her past medical history consisted of a left mastectomy (in 2006) and an appendicectomy for carcinoid tumour (in 1990). Otoscopy revealed a retracted right tympanic membrane, and audiometry confirmed a moderate conductive hearing loss (60 dB). Initial management involved the insertion of a grommet.

Radiological findings

A high resolution computed tomography (CT) scan of the right temporal bone revealed soft tissue debris associated with the ossicular chain, and lack of pneumatisation of the mastoid air cells.

Management

An exploratory cortical mastoidectomy and middle-ear exploration were performed. A yellowish mass extending from the mesotympanum to the mastoid antrum was found and excised.

Histological findings

The lesion appeared to have features of middle-ear adenoma showing neuro-endocrine differentiation. A staging CT scan did not show any distant neuro-endocrine tissue. Screening hormonal tests were negative. Metaidobenzylguanidine and octreotide scanning was normal. An annual chromogranin-A scan was organised as follow up.

Discussion and lessons learnt

Dr Connor and Dr Sandison held the opinion that this lesion was most likely a primary middle-ear adenoma rather than a primary carcinoid tumour. Dr Sandison felt that metastasis from the previous appendix carcinoid tumour was highly unlikely. Endocrine and ENT follow up is required to exclude the presence of other neuro-endocrine tissue and to detect any early recurrences.

'Speckled neck': a unique cause of paroxysmal lingual swelling and neck pain

A Qureshi, R Obholzer, M Gleeson

From Guy's Hospital, London

Introduction

A 42-year-old woman was referred with persistent left neck pain and intermittent lingual swelling. In 2006, she had undergone resection of a left cervical paraganglioma for similar symptoms.

Case report

On examination, the patient had a left-sided vagal palsy and no palpable neck mass. Magnetic resonance imaging showed a left cervical tumour. Intra-operatively, a pigmented, parapharyngeal tumour was found deep to the common carotid artery, with multiple speckled, soft-tissue metastases. The tumour appeared to be arising from the cervical sympathetic chain and extending into the skull base. The tumour could not be excised completely.

Radiological findings

Magnetic resonance imaging demonstrated a lesion centred at the level of the first cervical vertebra, lying medial to the internal carotid artery and partly encasing it, and extending inferiorly into the carotid bifurcation. It had a low signal on T2-weighted images, with small internal signal voids. Some left-sided cervical nodes were also visible.

Histological findings

Expert review of the previous and current histology indicated that the lesion removed in 2006 was a paraganglioma while the lesion removed in 2011 was a malignant melanocytic schwannoma. The two were not interlinked.

Management

The patient was referred to a specialist melanoma oncologist.

Discussion

Expert radiological review concluded that, although appearances were of a paraganglioma, the tumour was in an unusual location. The surgeon who had initially operated on the patient in 2006 recalled that even at first resection it had been an unusual tumour which had extended into the skull base. Nonetheless, the patient's symptoms had resolved following initial surgery. The more recent tumour was not thought to be a malignant change in the residual paraganglioma.

Conclusion

Unusual symptoms plus a parapharyngeal mass may be due to a melanocytic sympathetic schwannoma.

A patient with VIth and VIIth cranial nerve palsy

K Varadharajan, L Poynter, R Kanegaonkar

From the Medway Maritime Hospital, Gillingham

Introduction

Malignant otitis externa is a relatively uncommon condition with potentially life-threatening intracranial complications. We present a patient with malignant otitis externa which culminated in sigmoid sinus thrombosis associated with *Aspergillus flavus* infection.

Case report

A 71-year-old, diabetic man was reviewed several times in the ENT emergency clinic for persistent, left-sided otitis externa. After developing a left facial palsy (House–Brackmann grade 4) associated with labile blood sugar levels, he was admitted and recovered after treatment with intravenous ciprofloxacin. Ten days later, he re-presented with drowsiness, recurrence of the left-sided facial palsy and additional left abducens palsy.

Radiology

An initial computed tomography (CT) scan of the mastoids showed extensive soft tissue changes extending from the external auditory canal to the temporal bone, consistent with malignant otitis externa. A magnetic resonance imaging (MRI) scan performed on re-presentation showed a parapharyngeal soft tissue swelling together with left jugular bulb and sigmoid sinus thrombosis.

Management

An emergency left mastoid exploration with left sigmoid sinus clot evacuation and skull base debridement was performed. A T-tube was inserted and a mass of 'white, cheesy' material was removed from the middle ear and sent for histology.

Histology

The specimen was consistent with *A flavus* infection. Under microbiological advice, the patient was started on voriconazole.

Conclusion

Mr Obholzer emphasised the importance of considering malignant otitis externa in elderly, diabetic patients with persistent otitis externa. Despite full resolution of facial and abducens nerve palsies within 10 weeks of antifungal therapy commencement, it was agreed that antifungals should be continued for several months to prevent recurrence of infection. Dr Connor suggested that progress should be monitored with CT and MRI scans as well as a galium white blood cell scan.

A five-year history of pulsatile tinnitus resolving immediately after mastoid exploration surgery

G Chawdhary, A Lamyman, C Milford

From the John Radcliffe Hospital, Oxford

Introduction

Pulsatile tinnitus presents a diagnostic challenge, and can reflect anything from normal variation to significant pathology.

Case report

A 49-year-old woman presented with a 5-year history of left-sided, pulsatile tinnitus, with a normal ENT examination.

Radiological findings

A high resolution computed tomography scan of the petrous bones revealed a left temporal bone lesion with bone lysis, expansion and cortical erosion. The condylar fossa, anterior mastoid, zygomatic process and external auditory canal wall were involved.

Management

The patient underwent exploration of the left mastoid. At operation, abnormal soft tissue was found and removed. Immediately post-operatively, the patient's long-standing tinnitus was reported to have disappeared.

Histological findings

Macroscopically, the lesion was composed of brown tissue fragments. Microscopically, it contained giant cells, areas of osteoid and chondroid formation, and 'chicken-wire' calcification. Morphological and immunohistochemical staining (positive for S100, podoplanin, DMP-1 and Ki-67) indicated a chondroblastoma.

Discussion and lessons learnt

Dr Sandison commented that the histology showed classic features of chondroblastoma, and agreed with the diagnosis. Unfortunately, the patient's tinnitus returned after a few weeks, necessitating a second operation. The tumour was radically excised, with clear bone margins. At the time of writing, the patient was under long-term follow up to monitor for recurrence. Pulsatile tinnitus must be taken seriously and may represent serious or uncommon pathology.

A case of a discharging ear whilst eating

S Yapa, M Yardley, J Ray

From the Royal Hallamshire Hospital, Sheffield

Introduction

Sialo-aural fistulae are rare and usually arise as an acquired disease process or surgical complication. This case demonstrates the difficulty in establishing the diagnosis and deciding on appropriate management.

Case report

A 65-year-old man presented to the emergency clinic with a 10-week history of right-sided, clear ear discharge that increased when eating food. He was initially diagnosed with otitis externa but failed to respond to repeated courses of topical antibiotic treatment. There were no obvious

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fistulae within the ear canal; however, the discharge tested positive for amylase and was confirmed as saliva. Botox injections and a Jacobson's neurectomy failed to settle the problem. Subsequent examination revealed instability of the anterior canal wall with the jaw open, together with serous discharge.

Radiological findings

An initial sialogram was normal. Post-neurectomy, primary reporting of subsequent computed tomography and magnetic resonance imaging scans described some fluid within the mastoid air cells and the middle-ear cleft, with no bony defect around the salivary gland. However, secondary reporting noted a small protrusion of the temporomandibular joint capsule into the external auditory meatus, with no extension of parotid gland or fistula formation evident. Despite this, Dr Connor commented that there may have been a neck of parotid tissue extending up to the defect in the joint capsule.

Discussion

Dr Connor's comments, along with the positive amylase result, make a salivary gland fistula the likely diagnosis.

Management

A right-sided bony meatoplasty was performed. A defect in the anterior aspect of the external auditory meatus bone was identified and an interposition cartilage graft inserted. One year after surgery, there was no sign of recurrence.

Rhinology section

Mr Justin Kong

A rare cause of unilateral nasal obstruction

Y Poon, B Fu, J Lim

From the Medway Maritime Hospital, Gillingham

Background

Glomus tumours are benign neoplasms of the glomus body, a component of skin involved in regulating body temperature. They are commonly found in the subungual region of fingers, but can rarely occur in extra-digital sites. We report the 24th case of an intra-nasal glomus tumour in the English language medical literature, and one of the largest reported.

Case report

A 71-year-old woman presented with right nasal obstruction. Flexible nasendoscopy revealed a large, fleshy mass filling the right nasal cavity and pushing the septum to the left. Following computed tomography scanning, endoscopic excision of the tumour was performed.

Radiology

Computed tomography of the paranasal sinuses showed a 5.2×3.3 cm, soft tissue mass occupying the right nasal cavity, displacing the nasal septum to the left and extending posteriorly into the nasopharynx.

Histopathology

Histopathology showed a diffuse proliferation of bland cells with pale cytoplasm and round to ovoid nuclei, interrupted by vascular channels. A diagnosis of a benign glomus tumour was made. Dr Sandison suggested a haemangiopericytoma as one of the possible differential diagnoses.

Management

The procedure was uneventful and the patient was discharged home the next day. At two-month follow up, the operated site remained free of residual or recurrent disease.

Discussion

Glomus tumours often present with recurrent epistaxis; interestingly, this symptom was absent in our case. Management with complete local resection is generally curative.

Invasive lesion of the maxillary antrum

G Wong, Z Awad, R Farrell

From Northwick Park Hospital, London

Introduction

It is rare to diagnose vascular lesions in patients presenting with a mass in the maxillary antrum, and even more extraordinarily when such a lesion invades the surrounding paranasal sinuses and the orbit, causing significant proptosis. We consider such a case and also review the literature on invasive arteriovenous malformation in the maxillary sinus.

Case report

A 77-year-old man presented with a 1-year history of an enlarging, right maxillary mass. This had displaced his right orbit, causing telecanthus and hypertelorism. He was an ex-smoker with atrial fibrillation and no past surgical history.

Radiological findings

A computed tomography scan of the sinuses demonstrated a T_4 lesion of the right maxillary sinus with breaching of all bony confines and erosion of the right sphenoidal sinus. Magnetic resonance imaging confirmed invasion of the right orbit with displacement of the orbital structures and proptosis; it also showed infiltration into surrounding muscle and the nasal septum. Computed tomography angiography demonstrated a feeding vessel from the right maxillary artery. However, Dr Connor commented that the mass appeared to be mainly soft tissue, and that he would normally have expected more blood vessels to be visible in an arteriovenous malformation.

Histological findings

Dr Sandison obtained the opinion of Professor Cyril Fisher (from the Royal Marsden Hospital, London). He believed that the vascular spaces indicated a vascular malformation or an organising thrombus.

Management

The patient underwent debulking of the tumour, and has since been managed conservatively.

Discussion

Dr Sanderson concluded that, even in the face of a large mass, the diagnosis of an arteriovenous malformation is sometimes not obvious. In the presented case, we propose that the growing mass arose due to intermittent bleeds from a small arteriovenous malformation which was contained within an organised thrombus.

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An unusual site for a maxillary ameloblastoma recurrence

S P Williams, A Lamyman, C A Milford
From the John Radcliffe Hospital, Oxford

Introduction

Ameloblastomas are benign but locally aggressive, odontogenic, epithelial neoplasms. The commonest site of origin is the mandible, but approximately 20 per cent originate in the maxilla.

Case report

A 54-year-old man presented to the clinic as part of routine surveillance following a maxillectomy for a benign ameloblastoma 3 years earlier. Histology had shown complete excision, with no evidence of invasion beyond the maxilla. The patient now described a six-week history of swelling in the maxillectomy cavity, and a pedunculated, 1.5 cm lesion was seen in the superior aspect of the cavity.

Radiological findings

Contrast-enhanced magnetic resonance imaging revealed enhancing soft tissue abnormalities in the left ethmoid and both frontal sinuses. Computed tomography showed smooth expansion of the sinuses with preservation of bone.

Histological findings

Biopsies of irregular tissue around the left ethmoid were consistent with recurrent ameloblastoma showing both follicular and plexiform architecture. The tumour was composed of basaloid cells and showed peripheral palisading and ovoid, elongated nuclei, without evidence of mitotic activity or pleomorphism.

Management

Resection was performed via a standard osteoplastic flap and left external ethmoidectomy. Histological analysis confirmed macroscopic and microscopic clearance, with no malignant transformation. Eighteen months later, the patient showed no radiological or clinical signs of recurrence.

Discussion

It was noted that an osteoplastic flap was chosen rather than an endoscopic approach, as resection under direct vision was thought to offer a lower risk of any subsequent recurrence. Ameloblastoma has a high propensity to recur following inadequate surgical removal.

Conclusion

Ameloblastomas are rare, and presentations at extra-gnathic sites are exceptional. We report the first documented case in the literature of recurrent ameloblastoma in the frontal sinuses.

An unusual cause of Cushing's syndrome

C E Rennie, R De Souza, P Bouloux
From the Royal Free Hampstead NHS Trust, London

Introduction

Ectopic adrenocorticotrophic hormone (ACTH) secreting tumours are the second most common aetiology of Cushing's syndrome after pituitary adenomas, and are usually located in the chest or abdomen.

Case report

A 21-year-old man presented in 2001 with features of Cushing's syndrome. Endocrine testing suggested ACTH secretion of pituitary origin, and an inferior petrosal sinus venous sample seemed to confirm underlying Cushing's disease. The patient underwent trans-sphenoidal surgery and adenomectomy, but without evidence of cure. Due to persistent hypercortisolism, he subsequently underwent bilateral adrenalectomy. In 2004, repeated investigations suggested the presence of a residual pituitary lesion. The patient underwent further trans-sphenoidal surgery, without evidence of adenoma. In 2006, he underwent pituitary irradiation. While taking hormone replacement therapy from 2006 to 2010, he became gradually more pigmented. In 2010, repeated investigations again suggested ACTH secretion of pituitary origin. A review of all imaging in 2010 suggested the presence of a mass in the olfactory region which retrospectively appeared to have increased in size over time. An olfactory, ACTH-secreting aesthesioblastoma was suspected. The patient underwent complete resection of the mass, with normalisation of his ACTH levels. Histology was typical of an ACTH-secreting adenoma, with no suggestion of olfactory neuroblastoma.

Discussion

This case demonstrates that an ectopic pituitary adenoma can present the same hormonal regulation and inferior petrosal sinus venous sample results as a pituitary source of ACTH. Inferior petrosal sinus venous sampling is considered the 'gold standard' for the diagnosis of pituitary adenomas. However, any ectopic adenoma with venous drainage via the inferior petrosal sinus will present a diagnostic challenge; hence, any lesions found on imaging these regions should be carefully considered.

Rare neoplasm of the frontal sinus posing a diagnostic and reconstructive challenge

R Exley, A Markey, R K Bhalla
From the Manchester Royal Infirmary

Introduction

Forehead swelling may pose a diagnostic and management dilemma for the ENT surgeon.

Case report

A 31-year-old man with a history of chronic frontal sinusitis presented with a midline forehead swelling of 11 years' duration, unsuccessfully managed in Nigeria. The current presentation included an infero-lateral displacement of his left globe and resultant diplopia, with gross cosmetic deformity.

Radiological findings

Computed tomography scanning revealed bifrontal and left orbital bony abnormalities, suspicious for cranial fibrous dysplasia or osteoma.

Management

Following multidisciplinary discussion between the ENT, neurosurgical and oculoplastic surgeons, a bifrontal craniotomy approach was used to excise the bony abnormality, with immediate reconstruction using a novel, custom-made orbital roof prosthesis and bifrontal titanium cranioplasty.

Histological findings

Initial histology revealed fibrous and osseous components. Margins were clear and no evidence of malignancy was

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present. Appearances were suggestive of fibrous dysplasia. Histology was reviewed by an expert pathologist at the University of Manchester, who diagnosed multiple frontal osteomas.

Conclusion and lesson learnt

Unlike fibrous dysplasia, obliterative paranasal sinus osteomas uncommonly occur with recurrent sinusitis. The natural history of each is distinct. In our patient, the surgical risks were grave and reconstruction warranted a novel approach. Histological diagnosis can be difficult. At the Semon Club, Dr Connor agreed that the scans were equivocal. However, Dr Sandison felt that different areas of the specimen demonstrated appearances consistent with both osteoma and fibrous dysplasia. She recommended the specimen undergo *GNAS1* mutation testing. Positivity would confirm fibrous dysplasia.

Head and neck section

Mr Ricard Simo

An unusual cause of dysphonia

S Sharma, I Kwame, J Almeyda

From the West Middlesex University Hospital, London

Introduction

Syphilis is caused by *Treponeum pallidum* and seldom presents in the larynx. We describe an unusual case of advanced-stage syphilis presenting with dysphonia. A literature review indicated only one previous reported case of laryngeal syphilis in the Western World in the past two decades.

Case report

A 51-year-old Somalian, male ex-smoker presented with a 4-year history of dysphonia. Notably, he recalled being affected by a sexually transmitted disease many years ago. Flexible nasendoscopy demonstrated oedematous and erythematous vocal folds with irregular edges and a very granular appearance throughout the supraglottis and larynx. Blood investigations demonstrated neutrophilia and positive syphilis serology results.

Radiological findings

A chest X-ray was normal.

Histological findings

Biopsy specimens from the glottis and supraglottis were obtained under direct microscopic laryngoscopy. The histopathology report described mucosa with mild to moderate acanthosis, spongiosis with exocytosis and prominent dyskeratosis, suggestive of laryngeal secondary syphilis.

Management

The patient was referred to the sexual health clinic for antibiotic treatment. The response to treatment was dramatic, with marked improvement in the quality of voice and significant reduction in the glottic oedema.

Discussion

Dr Sandison felt that the histology showed an abundance of plasma cells, suggestive of an infective process, which, along with the clinical picture, was consistent with laryngeal secondary syphilis. Advanced-stage syphilis is rare in the United Kingdom; however, otolaryngologists should be aware of the many faces of this complex disease, the 'great imitator', especially as an accurate diagnosis can often lead to complete remission.

Extensive intravascular invasion of poorly differentiated thyroid carcinoma in a 36-year-old: management and treatment options

H Edmond, N Gibbins, R Simo

From Guy's Hospital, London

Introduction

Poorly differentiated thyroid cancer is rare, especially in young patients. Optimal treatment remains controversial. Surgery followed by radio-iodine ablation, and external beam radiotherapy are considered options.

Case report

We present a 36-year-old individual with poorly differentiated thyroid cancer and extensive intravascular involvement. The patient had a multinodular goitre which had enlarged over the previous five years. Fine needle aspiration (FNA) of the left hemi-thyroid returned a Thy3a result.

Radiology

Magnetic resonance imaging, computed tomography and magnetic resonance angiography demonstrated a 4 cm mass in the left hemi-thyroid. The tumour invaded the internal jugular and brachiocephalic veins. Intraluminal tumour extended from the skull base into the left brachiocephalic and subclavian veins.

Histology

Histopathological analysis indicated a staging of $pT_3\ N_0\ M_x$. The thyroid tissue demonstrated poorly differentiated thyroid cancer with prominent intravascular invasion. No lymph node or thymus metastases were present. Sections of the internal jugular vein demonstrated carcinoma attached to the luminal aspect and also present within smaller vessels of the wall.

Management

Total thyroidectomy, left modified radical neck dissection and removal of intramural tumour via an endarterectomy approach were performed in conjunction with the thoracic surgical team. The internal jugular vein was resected. Postoperative radio-iodine ablation was given. External beam radiotherapy was recommended as adjuvant therapy.

Discussion and lessons from the meeting

Mr Simo cited discussion with European colleagues who had managed a comparable case. Despite lack of official consensus, management was similar. The disparity between FNA and histopathology results was raised. Dr Sandison emphasised that FNA reporting is probability-based and should be interpreted in the context of other findings.

'Take home message'

Poorly differentiated thyroid cancer with extensive intravascular invasion is rare. Such cases should be discussed in a multidisciplinary setting in order to optimise treatment.

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A rare cause of subglottic stenosis presenting with cough and fluctuation in voice

R Hone, O Wakelam, S Baer
From the Conquest Hospital, Hastings

Case report

A 60-year-old call centre worker was referred to the ENT service with a 5-month history of a dry, barking cough, fluctuation in voice, and occasional throat pain. She had a past medical history of asthma controlled with salbutamol, and had never smoked. The ENT examination included flexible nasendoscopy and was initially thought to be normal, with bilateral vocal fold movement. The patient was diagnosed with functional dysphonia and referred to the speech and language therapy service, which advised voice exercises.

Six months later, the respiratory team reviewed the patient regarding her cough. Spirometry revealed an obstructive defect, and bronchoscopy now showed paradoxical vocal fold movement but no endobronchial abnormalities. The speech therapists were unhappy with the patient's progress and referred her back to ENT. She re-presented with stridor and over 6 kg of unintentional weight loss in the past 6 months. Neck examination revealed a right sternoclavicular joint swelling and tenderness, and flexible nasendoscopy showed subglottic stenosis. She was referred urgently for microlaryngoscopy, biopsy and laser debulking.

Histology

Histology showed a plasmacytoma/malignant myeloma. Positron emission tomography revealed multiple lytic bone lesions (in the right sternoclavicular joint, the fourth and fifth thoracic vertebrae, the left and right scapula, and the left seventh rib).

Management

Following discussion at the head and neck multidisciplinary team meeting, urgent treatment with cyclophosphamide, thalidomide and dexamethasone was started. The patient was considered for autologous stem cell transplantation but this was not given as she made a good recovery, with improvement in her voice and a clear airway.

Conclusion

The soft tissue of the head and neck is the second most common site for multiple myeloma, after bone, and should be considered in the differential diagnosis of malignancies. Dr Sandison commented that it is not known why malignant lymphoid cells settle in any particular location, and that multidisciplinary team management and tissue banking will assist with further research in this area.

A rare source of catastrophic bleeding from a tracheostome following total laryngopharyngectomy, oesophagectomy and stomach 'pull-up'

N Gibbins, R Oakley
From Guy's Hospital, London

Introduction

Catastrophic bleeding following major resections for neoplastic disease is potentially fatal. The patient may exsanguinate or drown. Management must be swift and should involve a multidisciplinary team. We present a rare case of major haemorrhage from a tracheo-innominate artery fistula following surgery.

Case report

A 56-year-old woman presented with a thyroid mass. Computed tomography (CT) and biopsy confirmed subglottic squamous cell carcinoma (SCC) invading the thyroid. The patient underwent successful total laryngopharyngectomy, oesophagectomy and a stomach 'pull-up' procedure. Three weeks post-operatively, she had a major haemorrhage from her tracheostome.

Radiology

Contrast CT and angiography revealed a fistula between the trachea and a cervical innominate artery. Intraluminal stents were placed to control the bleeding.

Histology

Histopathological analysis confirmed SCC of the subglottis, staged as $pT_4\ N_0\ M_0$.

Management

The bleeding was controlled by direct pressure from the endotracheal tube cuff and stabilisation in the operating theatre. Computed tomography angiography confirmed the origin, and endovascular stents were placed as a temporising measure. Sternotomy was performed and the irreparable innominate artery was transected. Two days later, an omental flap was used to cover the innominate artery stump and the trachea. This case highlights the need for an urgent, multidisciplinary approach to tracheo-arterial fistulae.

Discussion

Primary tissue coverage was discussed. Professor Howard suggested that primary resection of the clavicular heads with partial manubriectomy could have been performed to reduce the tension on the stoma, and commented that sternomastoid or pectoralis major flap coverage for the exposed vessels may have helped in this case. The role of covered endovascular stents was discussed, although there is currently no evidence concerning their efficacy in tracheo-innominate artery haemorrhage.

'Take-home message'

Prevention is better than cure. If vascular abnormalities are noted pre-operatively, the surgical team should plan accordingly.

Acute airway obstruction in papillary thyroid carcinoma

L Savage, A Alaani, R Simo
From Guy's Hospital, London

Introduction

We describe a case of neglected papillary thyroid carcinoma presenting acutely with airway obstruction.

Case report

A 69-year-old, Caucasian woman presented acutely to the emergency department with stridor, tachypnoea and airway compromise. She had a huge mass arising from the anterior aspect of the neck and causing tracheal compression.

Radiological findings

Computed tomography of the neck showed a large, mixed cystic and solid goitre with retrosternal and cranial extension, and thrombosis of the right internal jugular vein. Bilateral pleural effusions, multiple lung parenchymal lesions and airspace opacification were noted. Overall

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appearances were consistent with malignant transformation within a multinodular goitre, with right jugular vein thrombosis and lung metastases.

Histological findings

Histological analysis confirmed a columnar cell variant of invasive papillary thyroid carcinoma, staged as T_4 N_{1b} M_1.

Management

Acute management followed Advanced Life Support guidelines, with oxygen and nebulised bronchodilators. Definitive airway management was required, with awake fibre-optic intubation. The patient was subsequently admitted to the intensive care unit for sedation and ventilation, followed six days later by a scheduled tracheostomy. Following histological confirmation of the tumour, a total thyroidectomy and bilateral selective neck dissection were performed, with adjuvant thyroid-stimulating hormone suppression and radioiodine treatment.

Discussion

The chairman and meeting discussed the factors affecting the reasons for late presentation in such an extreme case. Of note, the patient was socially isolated, with no close friends or family contacts, although she was educated, articulate and had not previously consulted for the same problem. The chairman noted that such late presentations are more frequently seen in the South London catchment area.

Paediatric section

Mr Ian Hore

An enlarging cystic parotid gland lesion in a child

T S Ahmed, I Beegun, H Daya
From St George's Hospital, London

Case report

Paediatric salivary gland tumours are uncommon. A high index of suspicion for underlying malignancy is needed for any enlarging neck mass in this age group. We report a three-year-old girl who presented with a three-month history of a progressive, non-tender, enlarging swelling in the left parotid region. Clinical examination revealed a 3-cm diameter, firm, intraparotid lesion, but there was a history of recent size regression. Non-tuberculous mycobacterial infection was suspected, and magnetic resonance imaging (MRI) with fine needle aspiration cytology was performed. This revealed a predominantly cystic lesion in the superficial parotid lobe with some internal, solid, murally-based, frond-like papillary elements. Cytology was suggestive of low-grade mucoepidermoid carcinoma.

Total parotidectomy was performed. The facial nerve was preserved apart from two minor branches which were encased by tumour. Formal histology showed a partly cystic lesion with the appearance of an acinic cell carcinoma, and additional features of its rarer papillary-cystic variant. No vascular or perineural invasion was seen. Adjuvant treatment is controversial and was not recommended following multidisciplinary discussion. At the time of writing, the child was being closely monitored with clinical assessment and serial MRI scans.

Cystic head and neck lesions in children are often benign; however, this case illustrates the need to be wary of making this assumption particularly when imaging has demonstrated unusual solid elements in a lesion. Although not routinely advised in children, fine needle aspiration cytology may be helpful in these circumstances. Surgical excision should be recommended in all cases in which diagnostic uncertainty exists.

Professor Howard advised that, in children, facial nerve dissection must be performed under a microscope to avoid sacrificing the facial nerve branches, and agreed that there was no advantage to adjuvant radiotherapy. Mr Simo commented that parotid lumps in children should be considered malignant until proven otherwise.

Unexplained neonatal respiratory distress

G Narasimhan, A Kinshuck, R Clarke
From Alder Hey Children's NHS Foundation Trust, Liverpool

Introduction

Congenital tumours in neonates are rare but should be considered as a possible cause of airway obstruction.

Case report

A five-day-old girl presented with partial airway obstruction and oxygen desaturation since birth. Choanal atresia was excluded. Magnetic resonance imaging of the brain, performed to exclude intracranial pathology, detected a large nasopharyngeal mass.

Radiological findings

Magnetic resonance scans revealed a nasopharyngeal mass with high signal on T1-weighted images, suggestive of a teratoma; scanning also confirmed the absence of any intracranial connection. Ischaemic changes were noted in both cerebral hemispheres.

Management

An examination of the nasopharynx under anaesthesia was performed with a 120° rigid endoscope transorally. The mass was found to be attached to the posterior septum, and was carefully removed in toto with blunt dissection. The procedure was uneventful. The neonate was discharged home the following day. She was reviewed after six weeks, and was making excellent progress.

Histology

Complete removal was confirmed. The mass included the presence of ectoderm, mesoderm and endoderm components, without immature or malignant elements, consistent with a benign congenital teratoma.

Discussion

Professor Howard commented that although these tumours are benign, they can grow to a very large size and should always be operated upon as they have a well defined capsule which facilitates surgical excision. In every neonate with unexplained respiratory distress, the possibility of a nasopharyngeal mass should be considered.

From purpura to a nasal mass

K Davies, J McPartland, S De
From Alder Hey Children's NHS Foundation Trust, Liverpool

Introduction

Rosai–Dorfman disease is a rare disorder characterised by increased production of histiocytes. The most common accumulation is in the nodes of the neck; however, extranodal disease has been reported and is most prevalent in the upper respiratory tract and sinuses.

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Case report

We report the case of a three-year-old girl who presented to her local hospital with Henoch–Schonlein purpura. Over the next six weeks, she deteriorated, with left nasal discharge and an expanding nasal mass. She underwent computed tomography and was referred to our service. We carried out a biopsy, from which the diagnosis of Rosai–Dorfman disease was made.

Radiology

Magnetic resonance imaging showed a mass occupying the left nasal space, which extended into the medial wall of the maxillary antrum and the lamina papyracea and which remained intact. It lay against the nasal septum causing deviation to the right. The left inferior turbinate had been destroyed.

Histology

There was a diffuse infiltrate of large, histiocytic cells. Many cells were multi-nucleate, and occasional emperipolesis was seen of both lymphocytes and granulocytes. Immunohistochemistry revealed the histiocytes to be positive for S100 and cluster of differentiation 68 protein. These findings were consistent with a diagnosis of Rosai–Dorfman disease.

Management

On discussion with the radiology department, we organised an ultrasound scan to exclude any other lymphadenopathy. We then performed debridement of the nasal mass to improve the child's airway and symptoms. Rosai–Dorfman disease has been reported to have links with other autoimmune conditions, but an association with Henoch–Schonlein purpura has not previously been documented. Professor Howard agreed that our patient needed to be monitored regularly for any evidence of recurrence.

Difficult management of an unusual inflammatory paediatric neck condition

A V Kasbekar, G Kokai, A J Donne

From Alder Hey Children's NHS Foundation Trust, Liverpool

Introduction

Angiolymphoid hyperplasia with eosinophilia disease is a rare and idiopathic vascular disorder preferentially affecting the head and neck subcutaneous tissues of middle-aged women of far-eastern extraction. It is rare in children.

Case report

A three-month-old girl presented with inflammatory right neck lymph nodes that had developed into an abscess requiring drainage. The wound was slow to heal and required primary closure four months later, including biopsy of a protruding wound granuloma. Investigations were negative for tuberculosis. Serum immunoglobulin E levels were raised but there was no eosinophilia. At the time of writing, the child remained clinically well with a fluctuating nodal mass beneath the now healed scar.

Radiology

The original ultrasound of the lesion demonstrated a nodal, $4 \times 4 \times 3$ cm mass lying anterior to the subclavian vein, which eventually became necrotic. A more recent magnetic resonance imaging scan, taken after primary wound closure, displayed persistent cervical lymphadenopathy beneath the surgical scar. The thorax, abdomen and pelvis were unremarkable.

Histology

Initial specimens displayed the appearance of a chronic abscess wall. Biopsy of the wound granuloma revealed characteristics of angiolymphoid hyperplasia with eosinophilia disease, including prominent 'hobnail'-type, 'histiocytoid' endothelial cells within the vessel walls, along with numerous eosinophilic granulocytes. Dr Saunders felt the florid inflammatory response could represent a fungal infection, and suggested the possibility that a non-representative biopsy may have been taken.

Management

Professor Howard agreed with our policy of conservative management, i.e. observation until the mass becomes sufficiently symptomatic. He also recommended pathology panel discussion for such cases. The possibility of renal involvement, a reported association, was excluded.

Discussion

In view of the unusual diagnosis and presentation, it was suggested that the case be sent for expert review to Dr Tim Helliwell, a specialist head and neck pathologist in Liverpool. He confirmed the benign diagnosis.

A rare case of a penetrating, life-threatening injury to the oropharynx

A Crockett, S Shah, J Pickles

From the Luton and Dunstable Hospital, Luton

Case report

A six-year-old child presented to the accident and emergency department after he had been found in the street apnoeic and cyanosed with a broken pencil embedded in his mouth and a Glasgow Coma Score of 3. In the accident and emergency department, the child's Glasgow Coma Score fluctuated between 8 and 13. A penetrating injury of the soft palate was noted, and a provisional diagnosis of penetrating intracranial injury was made. Antibiotics were given and the child was intubated and ventilated. Computed tomography (CT) showed no evidence of brain or spinal cord injury. Examination under anaesthesia revealed a small penetrating wound in the nasopharynx with cerebrospinal fluid (CSF) leakage. The child was transferred, intubated, to Great Ormond Street Hospital. Attempted extubation after 72 hours failed because of a poor gag reflex and aspiration. Magnetic resonance imaging (MRI) and magnetic resonance angiography showed an ill-defined lesion in the right side of the medulla and upper spinal cord. Antibiotics were continued. After 9 days, a tracheostomy was fashioned. The child's level of consciousness subsequently improved, but there was evidence of a partial bulbar palsy together with left facial and upper and lower limb weakness. The CSF leak ceased spontaneously within 72 hours. Further management consisted of nasogastric tube feeding and rehabilitation, including swallowing therapy and physiotherapy. After six months, there was still a mild left-sided limb weakness.

Conclusion

This unusual penetrating injury of the brain stem was diagnosed on clinical grounds and correct management instituted despite a normal CT scan. Suspicions were subsequently confirmed by MRI scanning, together with the development of focal neurological signs which became apparent following extubation.

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