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Proceedings of the 145th Semon Club, 23 May 2013, 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, Charing Cross Hospital, Imperial Healthcare NHS Trust

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

Professor Leslie Michaels prize for the best presentation of the meeting was awarded to Mr Peter Radford for 'Refractory otitis externa: when malignant means malignant'.

The chairman and secretary 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.

Otology and skull base section

Chairman: Mr Rupert Obholzer

One kidney, two ears

S Roy, S N Ifeacho, N Patel

From the Whipps Cross Hospital, Barts Health NHS Trust, London

Introduction

Metastatic renal cell carcinoma in the head and neck region is rare. To our knowledge, metastatic papillary renal cell carcinoma in the external auditory canal has not been previously reported.

Case report

A 70-year-old gentleman presented with a 2-month history of right-sided offensive otorrhoea that was unresponsive to topical antibiotic therapy. His past medical history was remarkable: he had previously been treated for chronic myeloid leukaemia, renal cell carcinoma and transitional cell carcinoma.

Clinically, friable soft tissue filled the external auditory canals. In addition, the left side demonstrated areas of bony erosion and exposure.

Radiological findings

Temporal bone computed tomography scans demonstrated an expansile soft tissue mass filling the right external auditory canal, and extending into the middle ear and attic. A smaller left-sided mass extended to the middle ear, with erosion of the bony part of the external auditory canal.

Histological findings

Tissue sections were infiltrated by malignant neoplasm comprising nests and trabeculae. No features of keratinisation were seen. Immunohistochemical staining was negative for squamous cell markers, but strongly positive for alpha-methyl-acyl-CoA racemase ('AMACR'). In light of the patient's history of renal cell carcinoma, the histological findings were suggestive of metastatic papillary renal cell carcinoma.

Management

This gentleman's care was discussed at both the head and neck, and the urology multidisciplinary meetings. A decision was made to commence with chemoradiotherapy.

Conclusion

Common diseases are common; however, in the presence of advanced clinical signs and treatment failure, the list of differential diagnoses must be expanded and investigated further. Tissue biopsy is very useful in this respect.

An unexplained conductive hearing loss

G Reddy-Kolanu, S J Broomfield

From the Royal Blackburn Hospital

Introduction

Middle-ear disease with conductive hearing loss is most commonly due to otitis media with effusion. Rarer causes such as tumours and systemic inflammatory diseases must also be considered.

Case report

A 56-year-old lady presented with conductive hearing loss and a bulging intact tympanic membrane. Past history included nasal polyposis and adult-onset asthma. There were no other systemic symptoms. Haematological investigations only showed mild eosinophilia.

Radiological findings

Computed tomography and magnetic resonance imaging revealed soft tissue filling the left middle-ear space.

Histological findings

Middle-ear exploration revealed inflammatory tissue filling the middle ear. Histology showed granulation tissue with increased numbers of eosinophils. There was no evidence of vasculitis or granuloma formation.

Management

A trial of systemic corticosteroids was administered, with symptomatic improvement. A rheumatological opinion was sought, and this suggested a clinical diagnosis of upper

airway vasculitis, based on the history, eosinophilia and steroid responsiveness. The patient was subsequently treated with methotrexate.

Conclusion and lessons learned

In the presence of normal haematological and histological findings, upper airway vasculitis can be diagnosed clinically. We would advise management in conjunction with rheumatology.

Sequential destruction of both temporal bones in a 48-year-old female over 7 years

T Jacques, S Sharma, G Watters

From the Southend University Hospital, Westcliff-on-Sea

Introduction

Whilst temporal bone involvement in Langerhans' cell histiocytosis is well reported, bilateral involvement is extremely rare. Bilateral temporal bone destruction associated with Langerhans' cell histiocytosis in an adult has been described in only three case reports. We present an adult patient who experienced a wide array of otological symptoms due to Langerhans' cell histiocytosis over a prolonged period.

Case report

A 48-year-old female presented with a painful left-sided facial palsy, which was self-limiting. She subsequently developed an enlarging, fluctuant swelling above the left pinna. Seven years later, the patient presented with mild conductive hearing loss and pressure symptoms affecting the right ear, followed by complete sensorineural hearing loss on this side, accompanied by bloody otorrhoea.

Radiological findings

Computed tomography revealed significant destruction of the left temporal bone, followed seven years later by destruction of the right side. In both episodes, the lesion was osteolytic, involving the mastoid antrum and air cells, and extending to the squamous temporal bone and external auditory canal.

Histological findings

Surgical biopsies revealed histiocytic cells positive for cluster of differentiation 1a and langerin, confirming Langerhans' cell histiocytosis.

Management

The first episode was controlled with low-dose radiotherapy. The second episode was treated as a systemic relapse with cytotoxic chemotherapy (prednisolone, vinblastine and mercaptopurine). The patient unfortunately suffered neutropenia complicated by pneumonia, from which she died.

Discussion

Mr Obholzer stated that whilst temporal bone Langerhans' cell histiocytosis would certainly be considered in such a presentation in a child, it is extremely unusual in an adult. Because of the recalcitrant nature of the disease, it is important to monitor patients with Langerhans' cell histiocytosis long term, using serial scans. Dr Sandison warned that a diagnosis of Langerhans' cell histiocytosis in adults should be made with caution, as a reactive histiocytosis can be caused by other pathology, such as malignancy.

Soft tissue swelling of the external auditory canal

N Amin, N DeZoysa, S Lew-Gor

From the Worthing Hospital, Western Sussex NHS Trust

Introduction

Angioleiomyoma is a vascular subtype of leiomyoma. It is a benign tumour of smooth muscle and vascular endothelium. Only 10 per cent are found in the head and neck. We present the fifth recorded case of an angioleiomyoma in the external auditory canal.

Case report

A 92-year-old lady presented with a 1 cm, painless swelling at the entrance of her right external auditory canal, which was increasing the difficulty of hearing aid insertion. The rest of the external auditory canal and tympanic membrane were normal, with no associated head and neck lymphadenopathy.

There was a past medical history of a surgically resected colonic tumour.

Management

Surgical excision biopsy under local anaesthetic was undertaken using an endaural approach. At follow up, there were no post-operative complications or recurrence.

Histological findings

Macroscopically, there was a $15 \times 12 \times 9$ mm, irregular, pale and haemorrhagic lesion within the right external auditory canal.

Microscopically, there was evidence of mature smooth muscle tissue surrounding irregular, dilated vascular spaces. No cellular atypia or necrosis was identified.

Conclusion

There have only been a handful of recorded cases of angioleiomyoma in the external auditory canal. The aetiology is poorly understood. It can represent a challenging diagnosis. Treatment should involve examination of the external auditory canal under anaesthesia and total surgical excision.

Refractory otitis externa: when malignant means malignant

P Radford, N Foden, J Ramsden

From the John Radcliffe Hospital, Oxford

Introduction

Deeply painful otitis externa in the immunocompromised patient often indicates necrotising (malignant) otitis externa; however, a non-infective aetiology should also be considered. We report a patient with refractory symptoms, despite treatment. Biopsies confirmed adenocarcinoma.

Case report

An 81-year-old man who was being treated for otitis externa developed worsening otalgia, which caused him to wake at night. Necrotising otitis externa was confirmed on computed tomography (CT). Biopsies showed scanty epithelial cells. Intravenous antibiotics led to full improvement and the patient was discharged on oral therapy. Two weeks later, his otalgia returned with a new onset of facial nerve palsy. Following a repeat CT scan, he underwent a mastoidectomy.

Radiological findings

The second CT scan demonstrated necrotising otitis externa and mastoiditis, with erosion of the ear canal wall. Following

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mastoidectomy, magnetic resonance imaging demonstrated local neoplastic infiltration. Positron emission tomography (PET) (fluorodeoxyglucose) imaging demonstrated a residual local tumour with bony metastases. No alternative primary was found.

Histological findings

The deep biopsies revealed poorly differentiated adenocarcinoma with widespread lymphovascular and perineural invasion. Immunohistochemistry showed strong positivity for cytokeratin 7. A diagnosis of primary ceruminous adenocarcinoma was suggested.

Management

Given the bony metastases, the multidisciplinary team decided to forego further surgery, opting instead for palliative radiotherapy.

Discussion

Dr Sandison stated that although primary ear canal adenocarcinoma is rare, immunohistochemistry markers excluded metastases from a gastrointestinal malignancy. Dr Siddiqui analysed the PET images showing local lymphadenopathy with no other primary sites. Dr Sandison commented that the endolymphatic sac can be a rare source for temporal bone adenocarcinoma, but this was radiologically clear.

Conclusion

Rare ear canal malignancy must be suspected when a patient with necrotising otitis externa fails to improve.

Head and neck section

Chairman: Mr Richard Oakley

A suspicious right-sided, deep neck space lesion – is organic farming responsible?

S Hadjisymeou, P Loizou, P Kothari

From the Luton and Dunstable NHS Foundation Trust

Introduction

A 66-year-old female patient was admitted with a right-sided, deep neck space lesion; malignancy was suspected.

Case report

The patient described a six-week history of a sudden, initially painful, neck swelling. On examination, a fixed level II and III mass was noted on the right side of the neck, measuring 5–6 cm.

Radiology

Ultrasound and magnetic resonance imaging revealed a large necrotic mass, with evidence of extra-capsular spread and involvement of the sternocleidomastoid muscle. The right carotid artery was medialised and encased by the necrotic mass that extended to the skull base.

Histology

Histological analysis of the tissue revealed acutely inflamed granulation tissue with foamy macrophages. There was no evidence of malignancy or lymphoid tissue.

Management

Lactococcus lactis cremoris infection was diagnosed, and the patient was treated with antibiotics and weekly dressings. The patient recovered rapidly, and a repeat ultrasound scan conducted four weeks later revealed complete resolution of the neck abscess. A likely reason for the infection was the intake of unpasteurised milk and cheese acquired from a farmer's market.

Conclusion

The case shows that atypical organisms should always be considered in the working diagnosis of an atypical neck abscess, especially in light of the rise in popularity of organic farming. Mr Oakley mentioned that there are studies in progress utilising genetically modified *L l cremoris* as the vector for delivering interleukin-10 and tumour necrosis factor antibodies to various tissues in the body. This technique can be used to treat conditions such as Crohn's disease.

Spontaneous regression of a rare mass in the temporomandibular joint

H Zhang, G Lloyd, T Odutoye

From the St George's Hospital, London

Introduction

Tenosynovial giant cell tumour is a benign proliferative disorder of the synovium characterised by the destructive invasion of synovial-like mononuclear cells. It is extremely rare in the head and neck.

Case report

A 43-year-old man presented with a painless, persistent, left-sided pre-auricular mass. He had no symptoms of infection, and suffered no weight loss or night sweats. He had no significant past medical history. On examination, a 5 cm, smooth, non-tender, bony mass was palpated. The mass was fixed to the temporomandibular joint (TMJ), with no associated lymphadenopathy. Facial nerve function was intact.

Radiological findings

Magnetic resonance imaging showed a 5×3 cm bony lesion associated with the left TMJ. The tumour was of uniform low signal on all sequences.

Cytology findings

Fine needle aspiration cytology revealed numerous mononuclear dendritic cells (lesional), with small lymphocytes and large multinucleate osteoclast-like giant cells (non-lesional), consistent with the diagnosis.

Management

After discussion of its zero malignant potential and the risks of surgery, the patient opted for watchful observation of his tumour. He attended annual review with no progression of symptoms.

Discussion

At the meeting, the images from 9 and 10 years after initial presentation were reviewed, which showed radiological evidence of regression. Mr Oakley suggested that the regression of this benign lesion could have been mediated by an immune response, or the tumour could have been inhibited by cytokines or growth factors. Mr Oakley also mentioned that the gene translocation causing this tumour had been identified as a gene producing growth-stimulating factor 1, and thus it was the local reaction to this that caused tissue damage in this case.

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Conclusion

This is the first case that describes the spontaneous regression of tenosynovial giant cell tumour in the head and neck. The findings support a conservative approach, which eliminates the risk of surgery and specifically facial nerve damage.

A recently described salivary gland tumour in a patient with a rare syndrome

N Foden, C Burgess, S Winter

From the John Radcliffe Hospital, Oxford

Introduction

Rubinstein–Taybi syndrome is a rare condition characterised by multiple congenital abnormalities and an increased risk of developing cancer. Mammary analogue secretory carcinoma is a recently described salivary gland tumour that morphologically resembles secretory carcinoma of the breast with the same t(12;15) (p13;q25) chromosome translocation.

Case report

A 35-year-old gentleman with Rubinstein–Taybi syndrome presented with a 1-month history of a rapidly enlarging right parotid mass. A 5×4 cm mass was found, involving the overlying skin. The facial nerve was intact.

Radiological findings

Ultrasound revealed a hypoechoic, homogeneous mass in the right parotid gland. Computed tomography confirmed the presence of a tumour, with no evidence of bony invasion or chest metastases.

Histological findings

Fine needle aspirate and core biopsies showed features consistent with a salivary gland neoplasm, but further subtyping was not possible. Post-surgery histology revealed the morphological and immunohistochemical features of a mammary analogue secretory carcinoma with a pathological tumour stage of T4a. Immunohistochemistry showed that the tumour cells were diffusely and strongly positive for S100, vimentin, and cytokeratins 7 and 19.

Management

Following a multidisciplinary team meeting, the patient underwent total parotidectomy with facial reanimation, as the facial nerve had to be sacrificed.

Discussion and lessons learned

Dr Sandison talked about the recent discovery of mammary analogue secretory carcinoma, and commented that retrospective analyses may uncover new cases that were previously labelled as something else, most likely acinic cell carcinoma. Genetic testing may unearth a link between mammary analogue secretory carcinoma and Rubinstein–Taybi syndrome in the patient.

Conclusion

Previous diagnoses may be reconsidered in light of newly recognised diseases.

An extremely rare pathology mimicking metastatic head and neck cancer

S Davey, M Wickstead

From the Norfolk and Norwich University Hospital

Introduction

Castleman's disease is a rare lymphoproliferative disease of unknown aetiology. We present a case of the hyaline vascular subtype manifesting in the neck. Extrathoracic presentations of this disease are very rare.

Case report

A 57-year-old female presented clinically with a large right supraclavicular mass that had increased in size over the preceding 2 months. She had been referred to the thyroid lump clinic as there was a background of diffuse thyroid goitre and hyperthyroidism. During the same period, she lost 8 kg and had developed a poor appetite. A 30-year history of heavy smoking was revealed. There were no other clinical findings.

Radiology

An ultrasound scan revealed diffuse thyroid enlargement and a pathological right supraclavicular node sized 2.7×1.6 cm. Microcalcification within the mass suggested a thyroid origin. A computed tomography scan 9 days later revealed the mass had grown.

Histopathology

Initial fine needle aspiration cytology and core biopsies were unhelpful, and definitive diagnosis via excision biopsy was recommended. This revealed little normal lymph node architecture, which had been replaced by a granulomatous inflammatory process. Extensive perivascular hyalinisation was noted, along with areas of necrosis, microcalcification and fibrosis. There were also foreign body type giant cells.

Management

Localised (unicentric) Castleman's disease of the hyaline vascular form can be cured following complete surgical excision.

Discussion and lessons learned

Although rare, this condition is an important differential diagnosis to metastatic head and neck cancer, and lymphoma. Originally, concomitant sarcoidosis was also considered; however, the initial report from our histopathology department was revised. Dr Sandison agreed that this was an extremely complex case because of the patient's other immunological conditions.

Lingual nerve ancient schwannoma – removal with nerve preservation

J Baldy, V Prasad, M Dilkes

From the St Bartholomew's Hospital, London

Introduction

Schwannomas are infrequent encapsulated neoplasms derived from neural crest cells. Ancient schwannomas are a variant of these which present with degenerative changes and nuclear atypia. These histopathological features may lead to a diagnosis of malignant mesenchymal neoplasm.

Case report

A 50-year-old Caucasian man presented with a 6-week history of left submandibular swelling. There was neither a relationship to eating nor 'B' type symptoms of lymphoma. A mobile, hard, level 1b nodal mass was found, possibly indicative of lymph node metastasis.

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Radiology

Ultrasonography revealed a 2.8 cm mass in the area of the left submandibular salivary gland. Fine needle aspiration cytology (FNAC) was performed. A magnetic resonance imaging scan showed a vascular mass, but it was not possible to determine if the lesion was deep or superficial to the mylohyoid muscle (sublingual or submandibular respectively). A positron emission tomography (fluorodeoxyglucose) scan was performed in view of the cytological findings and was negative.

Histopathology

Initial FNAC was inconclusive. Post-operative histology revealed spindle cells arranged in Verocay bodies (Antoni A), with no evidence of malignancy. Immunostaining of the specimen was strongly positive for S100 protein and negative for cytokeratin.

Management

Intra-operatively, a mass was found deep to the submandibular gland, lying directly on the lingual nerve. When dissection was attempted, the mass separated relatively easily from the nerve, preserving its form. Post-operatively, ipsilateral tongue sensation and coarse taste testing were normal.

Conclusion

The initial impression of a lingual nerve schwannoma was inconsistent with the intra-operative findings, as the excision of schwannomas rarely allows preservation of the nerve of origin. Taking into account the histopathology, it was agreed during the meeting that this mass likely originated either from the preganglionic or postganglionic fibres. Dr Siddiqui suggested the origin may have been from the mylohyoid nerve. Mr Oakley commented that if the origin had been the submandibular ganglion, reduced post-operative gland function would be expected.

Rhinology section

Chairman: Mr Ravinder Natt

Schhh…! It's not an inflammatory nasal polyp!

N Hariri, H A S Choudhury

From the Charing Cross Hospital, Imperial College Healthcare NHS Trust, London

Introduction

A schwannoma is a benign neurogenic tumour of nerve sheath origin and is rare within the sinonasal tract. They are difficult to diagnose clinically, radiologically and histologically. Treatment is primarily surgical, with most cases managed via an open approach; however, endoscopic resection offers the advantage of adequate clearance with good cosmetic results.

Case report

A 25-year-old woman was referred to our tertiary centre for surgical management. She initially presented with unilateral, left-sided nasal obstruction, persistent frontal headaches and preceding left-sided epistaxis. Biopsies at the referring centre were diagnostic. She was offered open surgical resection but expressed concern regarding facial scarring and was referred for assessment and endoscopic resection.

Radiological findings

Magnetic resonance imaging revealed a large, unilateral, heterogeneous, lobulated mass filling the nasal roof, with extensive erosion of the anterior skull base and nasal septum with deviation. There was involvement and remodelling of the middle and inferior turbinates.

Histological findings

Biopsy and resection specimens confirmed a benign schwannoma with degenerative changes.

Management

The patient underwent complete endoscopic resection of the tumour and repair of the skull base defect using septal cartilage.

Discussion and lessons learned

Dr Siddiqui emphasised the importance of excluding intraorbital and intracranial extension, and described how this may affect management. Miss Chevretton felt that the patient should have initially been offered endoscopic resection rather than open surgery; this view was echoed by all those present at the meeting.

Conclusion

Rare tumours should be considered in young patients with non-specific unilateral nasal signs and symptoms. Endoscopic surgery is the preferred approach when the multidisciplinary team diagnosis is secure and the operative skill is available.

Acute-on-chronic painful nasal swelling in a 43-year-old female

T Sammut, N Ryan, S Gillett

From the Royal United Hospital Bath

Introduction

Rosai–Dorfman disease, or sinus histiocytosis with massive lymphadenopathy, is a rare clinicopathological entity. Our patient presented with painful extranodal involvement without lymphadenopathy, and her condition mimicked a nasoseptal abscess.

Case report

A 42-year-old Asian female presented with chronic nasal swelling, nasal congestion and rhinalgia. There were no systemic features and no rhinorrhoea. There was deformity of the root of the nose externally, extending to the midpoint and mostly focused on the right lateral aspect. The lesion was large and erythematous. It was not fluctuant. Anterior rhinoscopy showed inflamed rhinitic mucosa.

Radiological findings

Magnetic resonance imaging showed soft tissue swelling of the base of the nose, with small focal collections of lobulated fluid in its most anterior aspect. Pneumatisation of the middle left turbinate was noted.

Histological findings

Sections showed emperipolesis, indicative of sinus histiocytosis with massive lymphadenopathy. Immunohistochemistry showed that the large histiocytes were positive for S100 and cluster of differentiation 68 (markers of histiocytes).

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Management

Endoscopic incision and drainage was performed. Operative notes commented on the appearance of caseous necrosis.

Discussion

The expert panel agreed that sinus histiocytosis with massive lymphadenopathy should be considered as a differential diagnosis for an atypical abscess. Histological confirmation of sinus histiocytosis with massive lymphadenopathy is required for diagnosis and subsequent treatment. The panel concluded that all atypical abscesses should undergo histological examination.

Left eye proptosis: a cavernous issue?

A Carter, K Kapoor, C Hopkins

From the Guy's Hospital, London

Introduction

Eduardo Tolosa and William Hunt independently described a rare disease involving non-specific inflammation within the cavernous sinus or superior orbital fissure, causing constant hemicranial or periorbital pain.

Case report

A 31-year-old female was admitted to a district general hospital with suspected periorbital cellulitis following 3 days of severe left eye pain, proptosis and diplopia.

Fifteen years prior, she had undergone subdural empyema drainage thought to be secondary to sinusitis, and a Lynch–Howarth incision for suspected periorbital cellulitis (with no pus).

Radiological findings

Magnetic resonance imaging (MRI) showed an abnormally enhancing, bulky left medial rectus muscle and left lateral cavernous sinus. The MRI scan after symptom resolution was unremarkable.

Management

Despite antibiotic treatment, there was no improvement of the patient's symptoms. Lumbar puncture was normal. A diagnosis of orbital pseudotumour or Tolosa–Hunt syndrome was made. Treatment with high-dose steroids produced an excellent response.

Discussion

Both orbital pseudotumour and Tolosa–Hunt syndrome are on the spectrum of 'idiopathic orbital inflammatory disease'. Both share the clinical features of periorbital pain and cranial nerve palsies, and both show an excellent response to high-dose corticosteroids. Tolosa–Hunt syndrome, however, specifically describes cavernous sinus or superior orbital fissure involvement.

Within the Semon Club meeting, both diagnoses were considered likely to be related to the same pathological process; however, Tolosa–Hunt syndrome describes a specific location.

Conclusion

Orbital pseudotumour and Tolosa–Hunt syndrome show an excellent response to high-dose steroids, and are likely to be related to the same pathological process. Recurrence can occur in up to 40 per cent of patients.

Epistaxis in a patient with von Hippel–Lindau syndrome

A Lee, J Emmett, D Roberts

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

Introduction

The von Hippel–Lindau syndrome is an autosomal dominant condition, which predisposes an individual to developing both benign and malignant tumours.

Case report

A 40-year-old male patient with von Hippel–Lindau syndrome presented with a 6-month history of epistaxis. The patient had been previously treated for multiple tumours, including grade 1a renal cell carcinoma. Examination at presentation revealed a polypoidal lesion within the right nostril.

Endoscopic biopsy of a right nasal mass revealed very friable tissue and superior palatine artery involvement, and resulted in an estimated 3 litres of blood loss.

Radiological findings

Magnetic resonance imaging and computed tomography (CT) scans showed a 45 mm nasal tumour, centred on the upper nasal septum just beneath the cribriform plate. The tumour involved the ethmoid sinuses and middle turbinate with local expansion. There was no bony destruction or intracranial extension. Abdominal CT scans showed no evidence of recurrent renal cell carcinoma or metastatic disease.

Histological findings

Histological examination showed extensive carcinoma of the ducts, tubules and small cell cysts, with a prominent clear cell component. However, the immunostaining results were not consistent with those of his previously excised renal cell carcinoma, and the possibility of a microcystic adenoma was raised by the initial reporting histologist.

Semon Club discussion

Dr Sanderson and Prof Eddy Odell (Guy's Hospital) felt that the histological findings were consistent with renal cell carcinoma, despite the controversy in immunostaining. Dr Siddiqui suggested a positron emission tomography CT scan to look for evidence of other metastases from the renal cell carcinoma.

Unilateral epiphora and amastigote-like cells – a diagnostic dilemma

C Yiannakis, D Walker, V Ward

From the Mid Yorkshire Hospitals NHS Trust

A 40-year-old gentleman was referred to ENT by ophthalmology with a 6-week history of left periorbital swelling and unilateral epiphora. In 2000, the patient underwent a dacryocystorhinostomy. In 2003, he presented to ENT with a unilateral neck mass. He was diagnosed with peripheral T-cell lymphoma, and successfully treated with chemoradiotherapy and a stem cell transplant.

Clinical examination revealed diffuse swelling of the left eyelid, and a firm, nodular swelling over the left ethmoid bone. Nasal endoscopy showed generalised enlargement of the lymphoid tissue in the right post-nasal space, multiple polyps (worse on the left) and mucopurulent discharge.

Computed tomography (CT) revealed diffuse involvement of the sinuses, particularly on the left, but no explanation for the periorbital oedema. Multiple biopsies of the nasolacrimal apparatus and post-nasal space were taken. The initial

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histology report confirmed chronic inflammation, but there was no evidence of lymphoma. This was later amended to describe the presence of intra-cytoplasmic particles that resembled the amastigotes of leishmania species within the macrophages.

The patient was reviewed at the regional tropical medicine centre; polymerase chain reaction and serology findings were negative for leishmania species. The patient's condition deteriorated, with the swelling extending over the nasal bones and involving the contralateral eye. Endoscopic examination confirmed the presence of tumour-like material in both nasal cavities. Repeat CT and biopsy of the nasal cavity and post-nasal space showed recurrent peripheral T-cell lymphoma.

The patient was treated with salvage chemoradiotherapy, but continued to deteriorate. Review of the initial radiological and histological findings at this meeting confirmed that a diagnosis of lymphoma could not have been made at initial presentation. Leishmaniasis was considered a rare but possible differential.

Paediatric section

Chairman: Mr Ian Hore

Refractory periorbital cellulitis in a five-year-old girl

S G Derbyshire, R Clarke, S De

From the Alder Hey Hospital, Liverpool

Introduction

Acute lymphoblastic leukaemia is the most common malignancy diagnosed in children. Patients usually present with symptoms secondary to bone marrow failure. Head and neck manifestations of this disease are rarely reported.

Case report

We present the case of a five-year-old girl who initially presented to a peripheral hospital with left-sided periorbital cellulitis. Initial management with antibiotics resulted in symptom resolution. Recurrence of periorbital swelling and erythema occurred within two weeks of discharge. A further episode was described prior to additional investigation.

Radiological findings

Computed tomography and magnetic resonance imaging showed a lesion arising from the left zygoma, breaching the cortex. Findings suggested a diagnosis of osteomyelitis, but could not rule out a malignant process. Subsequent biopsy of the lesion was performed.

Histological findings

These indicated precursor B-cell lymphoblastic leukaemia or lymphoma.

Management

The patient was referred for chemotherapy.

Discussion

The panel felt that cases of refractory periorbital cellulitis should be imaged to exclude more complex diagnoses. The images supported a malignant process over osteomyelitis.

Conclusion

A high degree of suspicion of rare aetiology should be considered in refractory periorbital cellulitis.

Permanent inability to phonate since childhood

M Raja, C Theokli, A Aymat

From the University Hospital Lewisham, London

Introduction

Paralysed vocal folds can be in the abducted position, mid-position or adducted position, depending on the level of nerve injury. This is the first case report describing bilateral vocal fold paralysis with folds fixed in full abduction.

Case report

A 12-year-old boy presented with a history of poor vocalisation, and a breathy hoarse voice with a lack of volume since learning to speak. As a baby, he had a poor cry. There was no history of swallowing or breathing difficulties.

Investigations

Videostrobolaryngoscopy demonstrated bilateral fixed vocal folds held in abduction, with flickering movement of the arytenoids only. Electromyography suggested a possible brainstem abnormality; however, no definite cause was identified. Microlaryngoscopy demonstrated bilateral, immobile abducted vocal folds, with no movement on palpation of the cricoarytenoid joints. Neck computed tomography (CT) revealed no cause for the fixed folds.

Management

Despite extensive investigation, a clear cause has not been found. Literature searches and discussion with colleagues nationally and internationally identified no cases of vocal folds fixed in abduction. A congenital cause has been considered because of the longstanding history. The patient is currently awaiting bilateral endoscopic medialisation of the anterior third of the vocal folds, to allow apposition of the folds and phonation.

Discussion

Discussions focused on the need to ensure that potential reversible causes for this unusual presentation were ruled out (with suggestions of brain magnetic resonance imaging and high-resolution CT of the arytenoids) before specialist referral to Prof Birchall for electromyography and assessment of suitability for re-innervation. Miss Chevretton felt that if short-term treatment was to be trialled, temporary fillers should be considered before more permanent procedures were contemplated. She also commented that (based on the video shown), assessment of adduction of the vocal folds would be difficult with the endotracheal tube in situ.

An unusual cause of recurrent ear infections in a child

B Attlmayr, G Kokai, S De

From the Alder Hey Children's NHS Foundation Trust, Liverpool

Introduction

Cases of recurrent ear infection are commonly referred to the out-patient paediatric ENT clinic. Rarely, symptoms may originate from non-infective aetiology.

Case report

A four-year-old boy with a six-month history of recurrent left ear infections presented to the ENT clinic. Otoscopy demonstrated a bony lesion occluding the external auditory canal. Pure tone audiogram showed an associated 40–50 dB conductive hearing loss.

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Radiological findings

Computed tomography (CT) revealed a $33 \times 32 \times 35$ mm expansile bony lesion arising from the petrous temporal bone, protruding into the middle cranial fossa. The differential diagnosis included aneurysmal bone cyst or giant cell tumour. Subsequent magnetic resonance imaging (MRI) revealed a heterogeneous, multi-loculated mass (with post-gadolinium enhancement). No definite radiological diagnosis could be made.

Histological findings

A biopsy was taken via a small endaural incision. This demonstrated a blood-filled cystic space surrounded by haemosiderin-laden macrophages, plasma cells and lymphocytes. This was in keeping with either organising haematoma or aneurysmal bone cyst.

Management

Following multidisciplinary team discussion, excision was attempted. The uneventful surgery was performed by paediatric ENT and skull base surgeons. Because of the close proximity of the facial nerve, the lesion could not be completely excised. At the four-month follow up, the patient was asymptomatic, and no residual disease could be demonstrated on CT or MRI.

Final histological analysis revealed a benign fibro-osseous lesion, most probably an ossifying fibroma.

Discussion

Radiological diagnosis or diagnosis from a limited biopsy can be challenging. Even histological diagnosis following subtotal excision may not be definitive.

Conclusion

A multidisciplinary approach to rare conditions such as ossifying fibroma is required to reach a conclusive diagnosis.

Nasal blockage in an 11-year-old girl leading to an unusual histological report

J G Barr, H Al-Reefy, I Hore

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

Introduction

Juvenile nasopharyngeal angiofibroma is a tumour only reported in adolescent males.

Case report

An 11-year-old girl presented to the ENT clinic with 6-month history of right-sided nasal blockage and snoring. On examination, there was a polyp filling the right nasal cavity.

At endoscopic sinus surgery, the polyp was found to originate from the right ethmoid sinuses. Removal proceeded as expected for a simple nasal polyp. Post-operatively, the patient's symptoms completely resolved.

Radiological findings

Pre-operative computed tomography (CT) showed a large, unilateral, polypoidal soft tissue mass filling the right side of the nasal cavity and extending into the nasopharynx. The right ostiomeatal complex was obstructed. The right maxillary sinus was aerated. Dr Siddiqui also noted lytic areas in the surrounding bone.

Histological findings

The excised polyp was covered by ciliated epithelium. The stroma was largely composed of irregular fascicles of spindle cells containing a mixture of thin-walled and muscular ectatic vessels. These findings were reported as being in keeping with juvenile nasopharyngeal angiofibroma.

Management

Following the unexpected histology report, we brought the case to the Semon Club for further discussion.

Discussion

The CT scan showed that the polyp originated from the ethmoid sinuses, rather than the sphenopalatine foramen where a juvenile nasopharyngeal angiofibroma would normally originate. Dr Sandison suggested the findings could represent florid reactive changes in an inflammatory polyp. She also suggested that androgen receptor status be determined on the specimen, a positive result being in keeping with a juvenile nasopharyngeal angiofibroma. It was also suggested during the meeting that hormone and chromosome testing of the patient be performed to exclude testicular feminisation syndrome.

Conclusion

The imaging and histology were not suggestive of juvenile nasopharyngeal angiofibroma; it was decided that genetic testing would not be performed unless the diagnosis was confirmed. The patient will be reviewed following androgen receptor testing.

Torticollis in a child – don't sleep on it

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Introduction

With the rising incidence of tuberculosis, this case serves as a timely reminder of its presentation in the head and neck, and the consequences of delayed diagnosis.

Case report

An 11-year-old girl presented with a 1-week history of sore throat, on a background of 3 months of weight loss, night sweats and worsening neck pain. She was previously well, having immigrated to the UK from North Africa seven years earlier. Restricted neck movements were noted on clinical examination. She had a soft posterior neck swelling and a retropharyngeal mass.

Radiological findings

Computerised tomography and magnetic resonance imaging scans demonstrated a 7 cm retropharyngeal abscess extending from the skull base down to the sixth cervical vertebra. There was evidence of osteomyelitis of the first cervical vertebra and extradural involvement around the second cervical vertebra.

Microbiological findings

Pus specimens were negative for acid-fast bacilli staining; however, 15 days later, *Mycobacterium tuberculosis* was grown from cultures.

Management

A multidisciplinary management plan initially included intra-oral aspiration of the collection. Subsequently, quadruple anti-tuberculous therapy was commenced. The cervical

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bony destruction was deemed unstable, and required open reduction and internal fixation of the first two vertebrae.

Conclusion and lessons learned

Children presenting with a stiff neck warrant full clinical examination that includes the oropharynx. Tuberculosis should be considered in the differential diagnoses, especially when there is no clear diagnosis. Mr Hore suggested that in cases with spinal involvement, surgical drainage in the Trendelenburg position avoids neck extension and may reduce the risk of spinal injury due to vertebral instability. Miss Chevretton added that neck extension should be avoided in all cases of retropharyngeal abscess, and that radiologically guided drainage was an alternative option.

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