

Online Only Abstract Selection

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Proceedings of the 153rd Semon Club, 13 November 2017, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Chairpersons: Miss Elfy B Chevretton and Miss Irume Pai, Guy's and St Thomas' NHS Foundation Trust

Secretary: Mr Sherif Haikel, Royal National Throat, Nose and Ear Hospital, University College London Hospitals NHS Trust

Invited panel for pathology: Dr Ann Sandison, Charing Cross Hospital, Imperial College Healthcare NHS Trust

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 Laura Leach for 'A rare case of a lingual mass in a neonate'.

The chairperson 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.

Rhinology section

Mr Ahmed Shahzada, Chairperson

Like a hole in the head? An unusual case of atraumatic skull base defect

SP Hudson-Phillips, A Walker and C Hopkins

From the Guy's and St Thomas' Hospital, London

Introduction

Granulomatous polyangiitis is a rare necrotising granulomatous condition, the aetiology of which is still unclear. Granulomatous polyangiitis is typically diagnosed by the presence of cytoplasmic anti-neutrophilic cytoplasmic antibodies (ANCA), and a histological triad of fibrinoid vasculitis, necrosis and granuloma. All three only feature in 15–25 per cent of cases.

Case report

A previously fit and well 63-year-old female presented acutely with suspected cytoplasmic ANCA negative localised granulomatous polyangiitis, which had resulted in extensive destruction of the osteocartilaginous structures of the nasal cavity and skull base. Flexible nasendoscopy revealed complete erosion of the nasal septum (Figure 1).

Radiological findings

Computed tomography imaging demonstrated pneumocephalus secondary to a large (2.5 cm) bony defect within the midline of the anterior cranial fossa, with complete destruction of the cribriform plate. Magnetic resonance imaging showed almost complete destruction of the cribriform plate and a small amount of herniation of the left gyrus rectus (Figure 2).

Histological findings

Histology showed diffuse chronic inflammation of the superficial mucosa, several hyalinised fibrous nodules with surrounding inflammation, and a scanty number of immunoglobulin G4 plasma cells. There were no convincing features of vasculitis, necrosis or granulomatous inflammation.

Management

Debridement of the nasal cavity and repair of the anterior skull base defect with a Biodesign® graft was performed.

Discussion

At the 153rd Semon Club, the panel were surprised by the lack of nasal crusting in this patient. As the findings were negative for cytoplasmic ANCA, Mr Shahzada Ahmed suggested re-naming the diagnosis as a fibrosing inflammatory process until myeloperoxidase and proteinase 3 were checked. These are additional blood markers that confirm a diagnosis of granulomatous polyangiitis. Dr Sandison agreed that there was insufficient evidence for a diagnosis of granulomatous polyangiitis. Miss Chevretton noted that some patients become

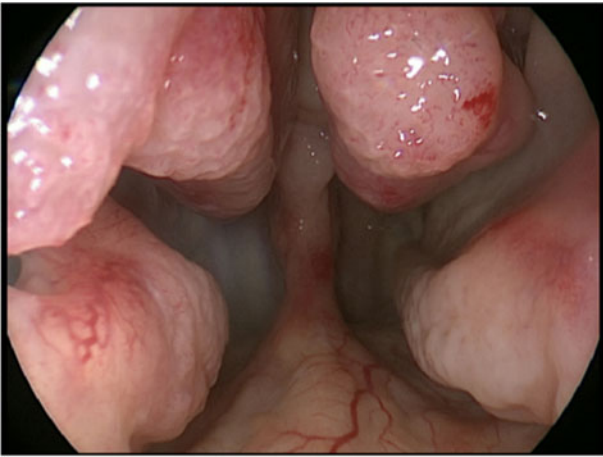


Fig. 1. Complete destruction of nasal septum on nasendoscopy with a striking lack of granulation tissue.



Fig. 2. Sagittal magnetic resonance imaging scan of the head showing almost complete destruction of the cribriform plate and a small amount of herniation of the left gyrus rectus.

cytoplasmic ANCA positive later in the course of the disease. Mr Rothera (Manchester) enquired if the patient had received treatment prior to presentation, but as far as we know this was not the case. Dr Connor pointed out that there were no other radiological features of granulomatous polyangiitis on the available imaging, and stressed the importance of taking adequate biopsies as early as possible in such cases.

A rare complication of nasogastric tube insertion

G Aldous and S Ahmed

From the Queen Elizabeth Hospital Birmingham

Introduction

Inadvertent extra-luminal insertion of enteral feeding tubes during placement is associated with significant morbidity and mortality.

Case report

A 72-year-old woman had had a nasogastric (NG) tube inserted while sedated on the intensive care ward post-

operatively, and her condition had then deteriorated. She had undergone computed tomography (CT) contrast studies of her feeding tube, which showed extensive contrast in the peritoneal cavity. She was referred to us in the operating theatre when the tip of the tube could not be found.

Radiological findings

Dr Connor showed how the CT scan with contrast injection into the feeding tube demonstrated that the tube punctured through mucosa into the retropharyngeal space at the level of the nasopharynx, descended through the mediastinum and past the diaphragm, with the tip in the peritoneum.

Management

The patient underwent exploratory laparotomy to wash out the feed. We performed pharyngoscopy and rigid nasal endoscopy of the nasopharynx, which demonstrated the perforation site to be in the post-nasal space. The tube was removed and a feeding tube was placed into the stomach under direct vision.

Discussion

To prevent future similar cases, Miss Pai suggested that a radiologist should report all NG tube chest radiographs. Miss Chevetton advised that senior clinicians should be involved in cases of NG tube insertion identified as difficult, with a low threshold for referral to ENT.

Conclusion

We report a rare complication of a common procedure. Junior clinical staff should continue to perform the procedure, while being aware of the conditions that increase the risk of such a complication occurring.

When Bactroban bites back

A Bernic, I Proctor and C Hopkins

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

Introduction

Topical antibiotics are frequently used in endoscopic sinus surgery, although the value of coating absorbable packs with topical antibiotics is uncertain.

Case report

A 30-year-old patient with aspirin-exacerbated respiratory disease and severe allergic rhinitis was referred with severe nasal obstruction. The clinical findings were grossly hypertrophied turbinates. The patient reported a history of prior nasal surgery. Eosinophilic granulomatosis with polyangiitis was considered the likely diagnosis, but antineutrophil cytoplasmic antibody test results were negative, and erythrocyte sedimentation rate and C-reactive protein were only slightly elevated. Eosinophil levels were mildly elevated.

Radiological findings

Computed tomography imaging confirmed turbinate hypertrophy, with unusual osteitic changes in both the middle and inferior turbinate bones, but relative sparing of the paranasal sinuses.

Histological findings

Histopathology showed no evidence of vasculitis, but instead demonstrated a picture of histiocytes and multinuclear giant

cells, thought likely to be a reaction to a lipophilic foreign body.

Management

Surgical excision of the middle and inferior turbinates was undertaken, with further division of adhesions. The original operation notes were requested. Following bilateral turbinate reduction, absorbable packs had been placed, which were covered with Bactroban® ointment. This contains polyethylene glycol, which is reported to cause a foreign body reaction when injected.

Discussion

The use of antibiotic-coated packing material post-operatively may allow penetration into turbinate submucosal tissue, the same as when injected. The panel agreed that this could be a cause of the intense foreign body reaction seen.

Conclusion

Water-based cream may be preferable to ointment formulations. Possible unwanted reactions need to be considered.

Lifelong nasal obstruction presenting with apnoeic episodes

A Jain, A Hariri and S Pal

From the London North West Healthcare NHS Trust

Introduction

We describe a rare case of increasing unilateral nasal obstruction caused by a slow-growing, large unilateral mass.

Case report

A 49-year-old African gentleman presented with a 30-year history of increasing unilateral (right) nasal obstruction. He was experiencing choking episodes at night, preventing sleep. Flexible nasoendoscopy revealed a large, firm mass that completely occluded the right posterior nasopharynx and extended into the choana. It was visible from the left side.

Radiological findings

A computed tomography scan showed a large calcified lesion within the posterior nasal cavity, originating from the right middle turbinate.

Management

Urgent excision biopsy was performed. A 4.5 cm solid mass was completely removed via the oropharynx, as it was too large to remove through the nose.

Histological findings

Histology revealed the mass to be a benign hamartoma. Nasal hamartoma is considered an infantile condition. Since its first description in 1974, few paediatric and even fewer adult cases have been reported. Given the atypical presentation, size, location and histology, a second histological opinion was sought, which confirmed the diagnosis.

Outcome and follow up

At follow up, complete relief of symptoms was reported. Examination revealed clear nasal passages bilaterally. The patient remains disease-free, and will be followed up once more before discharge.

Discussion

Dr Sandison commented on how, histologically, hamartomas appear to contain tissue constituents but in a disordered fashion. She advised that if such lesions are biopsied instead of excised, it would be easy to misdiagnose this as an inflammatory polyp. She also mentioned the importance of considering low-grade sinonasal adenocarcinoma as a differential diagnosis.

Conclusion

All unilateral nasal masses should be investigated with urgent excision biopsy. Benign nasal hamartoma is an important differential diagnosis.

When the clinical picture changes, question the underlying diagnosis: transformation on a background of vasculitis

A Bernic, R Hettige and C Hopkins

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

Introduction

Localised vasculitis is not uncommon, but is almost certainly under-diagnosed and easily overlooked. Nasolacrimal tumours are very rare, but diagnosis is often delayed and the condition mistaken for an infective process.

Case report

A 57-year-old male had received a histologically confirmed diagnosis of granulomatous polyangiitis 10 years earlier. He re-presented under a different team with a presumed flare-up of disease. He was found to have nasal crusting, peri-orbital swelling and extensive tissue destruction, with a large septal perforation. A biopsy of the perforation showed persistent granulomatous polyangiitis. Six months of increased topical and systemic treatment, including oral prednisolone and rituximab, failed to improve the clinical picture. Multiple courses of antibiotics were prescribed for progressive orbital cellulitis. The patient underwent regular debridement in clinic. After referral back to the original team, expansion of the nasolacrimal duct and sac were noted, and he was scheduled for an urgent biopsy.

Radiological findings

Dr Connor demonstrated diffuse mucosal thickening, a septal defect and amputation of the left inferior turbinate on initial imaging. Subsequent imaging showed an increase in the size of the septal defect and bony erosion of the left lateral nasal wall. There was new soft tissue now extending into the extraconal orbit. All these changes are notably consistent with a diagnosis of granulomatous polyangiitis.

Histological findings

Dr Sandison demonstrated areas of dense cellularity with necrosis, new blood vessel formation, and a rim of inflammatory cells, consistent with a diagnosis of nasal-type extranodal natural killer/T-cell lymphoma.

Management

The patient was referred to the haematology department for further treatment.

Discussion

A retrospective review of the biopsy taken six months earlier confirmed features consistent with granulomatous polyangiitis;

however, it also showed evidence of lymphoma. Dr Sandison felt that this was a very challenging case, and the panel agreed that asking the right questions is the key to finding the right answers.

Conclusion

When there is a clear change in presenting features, it is important to consider alternative diagnoses.

Otology section

Miss Irumee Pai, Chairperson

Progressive unilateral facial palsy

G E Hogg, R A Steven and P M Spielmann

From the Ninewells Hospital, Dundee

Introduction

Mixed benign pleomorphic adenomas are associated with a risk of recurrence and the ability to transform into metastatic tumours. Although commonly found in the salivary gland, rare cases have been found in the mastoid air cells. We describe an unusual presentation of an isolated benign pleomorphic adenoma in the facial canal.

Case report

A 55-year-old man presented with a 4-month progressive history of right facial paresis of House–Brackmann grade VI. He was a non-smoker, with a past medical history of right near total parotidectomy nine years previously and submandibular gland excision six years previously.

Radiological findings

A T2-weighted magnetic resonance imaging scan revealed an irregular, solid mass, sized 1.4 cm (craniocaudal) × 0.9 cm (transverse), centred on the descending portion of the right facial nerve, proximal to the right stylomastoid foramen (Figure 3).

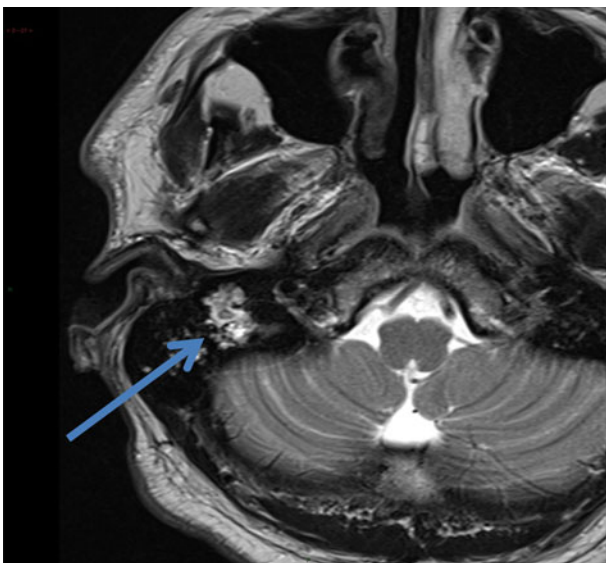


Fig. 3. T2-weighted axial magnetic resonance imaging revealed an irregular, solid mass, sized 1.4 cm (craniocaudal) × 0.9 cm (transverse), centred on the descending portion of the right facial nerve, proximal to the right stylomastoid foramen (arrow).

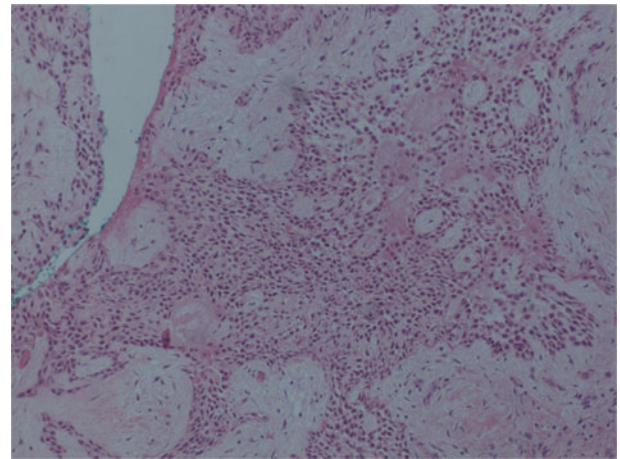


Fig. 4. Histological examination of the tumour revealed chondromyxoid and epithelial components, but no cellular atypia, indicating a mixed benign tumour of parotid origin. (H&E stain at magnification of × 200)

Histological findings

Dr Sandison demonstrated a tumour with chondromyxoid and epithelial components, but no cellular atypia, indicating a mixed benign tumour of parotid origin (Figure 4).

Management

The patient was managed with a VIIth cranial nerve excision, and upper neck and mastoid dissection, with an immediate cable graft of the great auricular nerve. Adjuvant therapy was declined, but he is currently undergoing dynamic facial re-animation with long-term otolaryngology follow up.

Discussion

Possible explanations included ectopic embryological parotid tissue and incomplete surgical excision of the primary lesion. Miss Pai held the opinion that it is likely to be a result of incomplete excision or seeding from the surgery performed nine years earlier. Miss Pai noted the risks of side effects or sarcomatous change if the patient is offered adjuvant radiotherapy. Dr Sandison suggested follow up with regular imaging as an alternative to adjuvant radiotherapy.

A rare syndromic cause of unilateral hearing loss

K Shanthakunalan and H R F Powell

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

Case report

In 2014, a 17-year-old female presented to primary care with fluctuating left-sided hearing. Community audiograms showed a unilateral sensorineural hearing loss (SNHL); however, on review by otolaryngology, her hearing had recovered and repeat audiometry was within normal limits. In 2017, she re-presented to otolaryngology with a sudden unilateral severe (70–80 dB) SNHL. Magnetic resonance imaging (MRI) of the internal auditory meatus (IAM) was requested. Contemporaneous recognition of proteinuria by her general practitioner prompted referrals to the nephrology and clinical genetics departments.

Radiological findings

The MRI showed subacute haemorrhage into the left vestibule and endolymphatic sac; hence, a contrast-enhanced MRI of the IAM was arranged. Gadolinium enhancement of the left vestibule was suggestive of an endolymphatic sac tumour

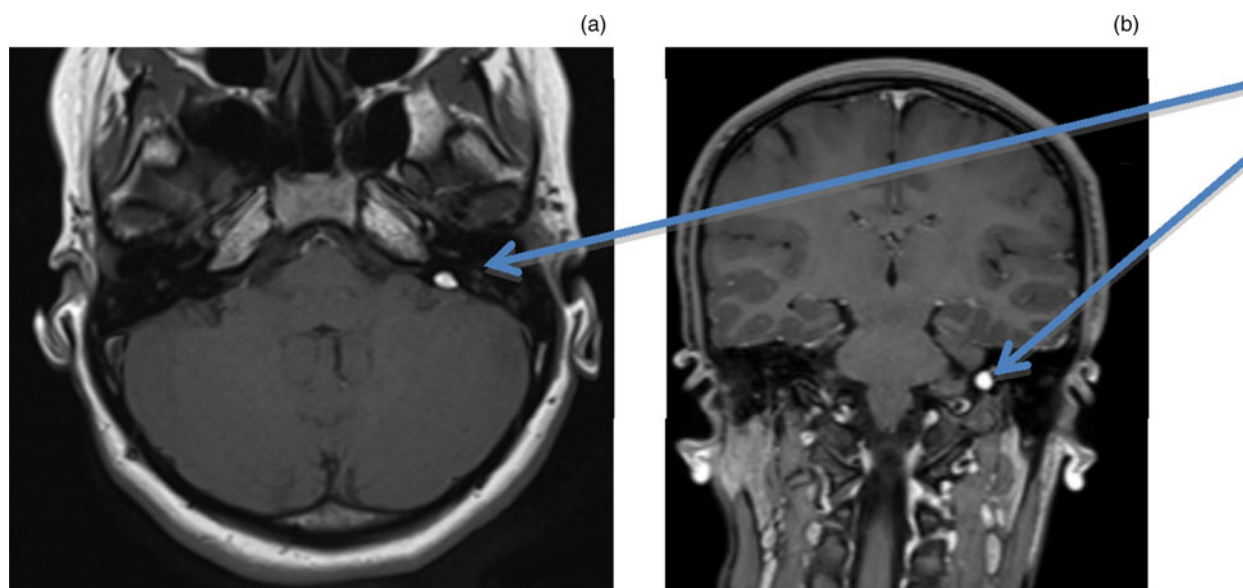


Fig. 5. (a) Axial and (b) coronal T1-weighted magnetic resonance imaging scans with gadolinium enhancement of the endolymphatic sac tumour (arrows).

(Figure 5), and, in light of the proteinuria, there was a suspicion of von Hippel–Lindau disease.

Management

Von Hippel–Lindau disease is an autosomal dominant condition characterised by the presence of haemangioblastomas. Patients may also develop visceral cysts in the kidneys, pancreas and genital tract, as well as tumours, including renal carcinoma and, in approximately 10 per cent, endolymphatic sac tumour. Investigations for other von Hippel–Lindau disease related tumours, ophthalmology review for retinal angiomas, and audiology review and hearing aid provision were arranged. Clinical genetics confirmed the VHL gene mutation.

Discussion

A discussion ensued as to whether surgical excision of the endolymphatic sac tumour would be appropriate. Given the stable aidable hearing and a lack of vestibular symptoms, Miss Pai advised against surgical intervention in the absence of a growing tumour.

Conclusion

Endolymphatic sac tumours are rare tumours often associated with von Hippel–Lindau disease. Rigorous multidisciplinary follow up is paramount to enable early diagnosis and management of von Hippel–Lindau disease associated tumours.

Tinnitus following lumbar puncture and its usual cure

W Jia and B Alomari

From the James Paget University Hospital NHS Trust, Great Yarmouth

Introduction

Audiometric disturbances, such as tinnitus or hearing loss, are recognised complications after dura puncturing procedures. We report the case of a patient with severe bilateral tinnitus following lumbar puncture, who was treated with an epidural blood patch. The mechanism for tinnitus is unknown. It could result from a negative intracranial pressure caused by a cerebrospinal fluid leak during these procedures, which can

cause an intra-labyrinthine pressure disturbance via the cochlear aqueduct. It could also result from the negative pressure causing traction on the vestibulocochlear nerve crossing the posterior cranial fossa, leading to a neurapraxia. A blood patch seeks to alleviate this negative pressure.

Case report

A 40-year-old lady presented with ongoing bilateral severe tinnitus and headache for 6 days after a lumbar puncture. She received a blood patch from the anaesthetic team.

Management

Eighteen millilitres of venous blood was injected into the epidural space. The patient scored 84 (grade 5) on the Tinnitus Handicap Inventory before the blood patch; 3 hours post-procedure her score improved to 16 (grade 1), with complete resolution by 24 hours.

Discussion

Dr S Connor said that audiovestibular symptoms are not uncommon in patients with spontaneous hydrocephalus.

Take home message

One should enquire about epidural puncture procedures in patients presenting with tinnitus. An epidural blood patch can be considered in these cases.

Acute unilateral ‘muffled’ hearing and tinnitus after exercise

E Casselden, W Ahmed and C Skilbeck

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

Introduction

Skull base chondrosarcomas account for 5 per cent of chondrosarcomas. Most occur on anterior and middle cranial fossae sphenoid chondrosarcomas; it is rare on the lateral skull base.

Case report

A 53-year-old male presented with left-sided muffled hearing and tinnitus. He denied discharge, vertigo and otalgia.

Otological examination findings were normal. There was no cranial nerve palsy and no post-nasal space mass. An audiogram showed mild sensorineural hearing loss at 0.5, 4 and 8 kHz on the right side.

Radiological findings

Dr Connor demonstrated a heterogeneous lesion on magnetic resonance imaging with gadolinium. The lesion was not centred on the jugular foramen, raising the possibility of petrous bone pathology or an exophytic lesion. A transmastoid biopsy was recommended at the skull base multidisciplinary meeting.

Histological findings

Dr Sandison demonstrated a high-grade tumour with gelatinous and myxoid components. She explained that, given the lack of bone in the specimen, it was not possible to confirm invasion. The combination of strong S-100, but no epithelial membrane antigen expression or brachyury, made this a chondrosarcoma. Genetic testing showed no nuclear receptor subfamily 4 group A rearrangement, making it a conventional subtype.

Post-operatively, the patient has intact facial nerve and inner-ear function. He will undergo surveillance imaging.

Discussion

This condition is rare, meaning there is limited consensus regarding management. Proton beam therapy is increasingly advised. This is not without risk (sarcomatous transformation and damage to surrounding structures). Conversely, local invasion and spinal drop metastases are reported in the literature.

Conclusion

The Semon Club panel agreed on conservative management for this case.

Unilateral middle-ear effusion in an adult: a rare presentation of an intracranial lesion

V Twigg, BL Wong and D Grant

From the Queens Medical Centre, Nottingham

Introduction

Here we present a case of delayed diagnosis of an intraosseous skull base meningioma causing unilateral middle-ear effusion.

Case history

A 51-year-old, fit and well female presented to the otolaryngology clinic with a right-sided hearing loss, nasal obstruction and intermittent pulsatile tinnitus. Clinical examination revealed unilateral otitis media with effusion and a mass in the nasopharynx. Urgent ventilation tube insertion was undertaken, and post-nasal space biopsy was performed showing only reactive lymphoid hyperplasia. Three weeks later, the patient re-presented to the ophthalmology department with deterioration in right-sided vision. Further examination showed right-sided optic neuropathy with nasal visual field defect.

Radiological findings

A computed tomography scan revealed a large skull base meningioma with a significant intraosseous component involving the optic canal. There was significant bony sclerosis of the temporal and sphenoid bone. The differential diagnoses include meningioma en plaque, metastatic deposits and other primary causes of neoplasia.

Management

After consultation with the neurosurgical team, a watch and wait policy has been adopted, with no current plans for surgical intervention.

Discussion

The case described here illustrates an unusual case of an intracranial lesion. While sensorineural hearing loss is not an uncommon presentation for an intracranial lesion, this case highlights that conductive hearing loss should be approached with caution. Attendees at the Semon Club commented that there was an argument to be made for early imaging in all adults with unilateral conductive hearing loss, and more so for patients with associated unilateral pulsatile tinnitus.

Destructive lesion of the temporal bone

G Wong, T Mawby and R Obholzer

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

Introduction

Temporal bone lesions are uncommon; aggressively destructive lesions are even rarer. Differentials include cholesteatoma, aggressive cholesterol granuloma and malignancy.

Case report

A 46-year-old man with chronic ear problems presented with a 1-year history of symptom progression. A rapidly enlarging right-sided post-auricular swelling ensued, culminating in a discharging post-aural sinus. Otoscopy demonstrated unilateral right-sided middle-ear effusion, with a mild ipsilateral conductive hearing loss on pure tone audiogram. There was a healed right-sided post-auricular cutaneous defect. There was no significant past medical history. Excisional biopsy ensued following magnetic resonance imaging (MRI) and computed tomography (CT). Intra-operatively, a destructive bony lesion was identified between the posterior and middle fossa dura over the transverse sinus. Macroscopic clearance of the lesion was achieved, with no symptom recurrence post-operatively.

Radiological findings

On the CT scan, Dr Connor demonstrated a well demarcated destructive lesion centred on the right posterior mastoid and sigmoid plate, with bony erosion of the skull and thickening of soft tissue. The MRI demonstrated the lesion to be isointense on T1-weighted imaging and mixed on T2-weighted imaging, with avid enhancement comparable with venous sinus structures. Appearances were suggestive of an infective or inflammatory process, with no features of cholesteatoma.

Histological findings

Dr Sandison demonstrated lots of fibrous tissue, mixed cells and histiocytes, all of which suggest a fibro-inflammatory lesion without malignancy. There was immature bone formation with a cortical reaction and normal architecture, indicating chronic inflammation.

Discussion

This is a rare lesion with no definitive diagnosis. Dr Sandison opines that it is important to preserve the histological specimen, because a future diagnosis may be attainable as we learn more about fibro-inflammatory lesions.

Paediatric section

Mr Christopher Pepper, Chairperson

Neck lump with associated Horner's syndrome in a four-month-old baby

L Leach and N Jonas

From the Addenbrooke's Hospital, Cambridge

Introduction

Extrarenal malignant rhabdoid tumour is a very rare and aggressive paediatric tumour with an extremely poor prognosis. Diagnosis is largely based on histopathology; however, this can be difficult because of the tumour's varied morphological features.

Case report

A four-month-old baby presented with an acute-onset, right-sided neck swelling, and was admitted for intravenous antibiotics administration. After an initial reduction in size, the mass persisted and the baby was reported to have right-sided Horner's syndrome.

Radiological findings

Initial ultrasound and magnetic resonance imaging scans revealed a lobulated cervical mass extending from level three down to the thoracic inlet. This is likely to represent abnormally enlarged cervical lymph nodes, although Dr Connor noted that there was no other lymphadenopathy. Repeat ultrasound showed enlargement, with concerning sonographic appearances. Staging computed tomography showed new retrosternal extension and tracheal displacement (Figure 6).

Histological findings

Histology revealed sheets of pleomorphic tumour cells, with some showing an eccentric eosinophilic collection within the cytoplasm, and others a large open nucleus with a prominent eosinophilic nucleolus. Immunostains showed a complete lack of INI-1 expression.

Management

In view of airway compromise, the baby was admitted immediately for chemotherapy, as the tumour was not surgically resectable. Following a good initial response, severe diarrhoea delayed chemotherapy for 10 days, during which there was rapid tumour regrowth. The baby was repatriated abroad for early phase trials.

Discussion

Dr Sandison highlighted the histological morphology of eosinophilic cytoplasm with globular pink cytoplasmic inclusions comprising whorls of intermediate filaments. Mr Pepper enquired if fine needle aspiration cytology would allow diagnosis. Dr Sandison responded that it might be possible, but only if the sample taken was representative and of adequate size to allow immunohistological characterisation.

Conclusion

A high index of suspicion and early biopsy is vital in the management of these aggressive tumours; more than 80 per cent of children die within one year and early diagnosis aids the chances of complete surgical excision.



Fig. 6. (a) Coronal and (b) sagittal computed tomography images of a right-sided heterogeneous cervical mass containing small enhancing blood vessels, displacing the trachea and pharynx to the left (a) and the right common carotid anteriorly (b).

The travelling pin: a swallowed foreign body

V Selimi, L Telesia and A Majithia

From the Northwick Park Hospital, London

Introduction

Upper airway foreign bodies are a common ENT emergency. We present the case of a swallowed pin migrating from the trachea to the gastrointestinal tract, thus changing management. To our knowledge, only one other case in the literature describes such migration.

Case report

A 14-year-old female, born at term with hay-fever and asthma, presented overnight after accidentally inhaling a 35 mm head-scarf pin. Initial chest and lateral neck radiographs revealed its location in the trachea. Rigid bronchoscopy was postponed until the morning as the patient was stable and there were logistical difficulties obtaining consent. The following morning, the patient reported coughing overnight. Fibre-optic nasoendoscopy to confirm the pin's position pre-procedure was not tolerated. Chest radiograph, requested instead, showed no pin. A subsequent abdominal radiograph revealed its



Fig. 7. Lateral neck radiograph showing the pin in the trachea. L = left



Fig. 8. Abdominal radiograph demonstrating the pin in the hepatic flexure. R = right

location in the hepatic flexure. The general surgeons advised that as there were no signs of perforation and uneventful passage through the large bowel could be expected, the patient could be discharged.

Radiological findings

A lateral neck radiograph showed the pin in the trachea (Figure 7) and an abdominal radiograph demonstrated the pin in the hepatic flexure (Figure 8).

Discussion

This case highlights the importance of visualising foreign bodies pre-procedure in the immediate period before extraction and after any significant events such as coughing, as in this case. Furthermore, clinicians should consider the gastrointestinal tract as a potential migratory site of displaced airway foreign bodies.

A paediatric patient with haemoptysis, stridor and an unusual diagnosis

I Rothera, M Sadacharam and M Rothera

From the Royal Manchester Children's Hospital

Introduction

Behçet's disease is a rare form of relapsing chronic vasculitis not normally associated with laryngeal involvement and airway obstruction.

Case report

A seven-month-old child presented with a rash to her face, transient haemorrhagic blisters to the nostrils, haemoptysis and haematemesis. Oesophagoscopy and colonoscopy detected no abnormalities. Later, she developed biphasic stridor. Direct laryngotracheobronchoscopy showed a swollen larynx, ventricular mucosal prolapse and very unusual appearances. Viral, vasculitic and autoimmune markers were negative. Further direct laryngotracheobronchoscopy showed continuing supraglottic and glottic well-delineated erosions and stenosis, with a normal appearance of the lower airways. Biopsies showed non-specific acute on chronic inflammation. She was commenced on high-dose prednisolone, but then developed dysphagia. In addition, she had worsening episodes of haemoptysis and occasional oropharyngeal ulceration (Figure 9).

Histological findings

Eventually, a lip ulcer biopsy sample was sent to Leeds. The histological features were compatible with a vasculitic process, characterised by the predominantly neutrophilic cell infiltration of small blood vessels. No acantholysis, epithelial spongiosis, blister formation or granulomas were observed.



Fig. 9. Image of the patient's larynx prior to commencing treatment and following a trial of prednisolone.

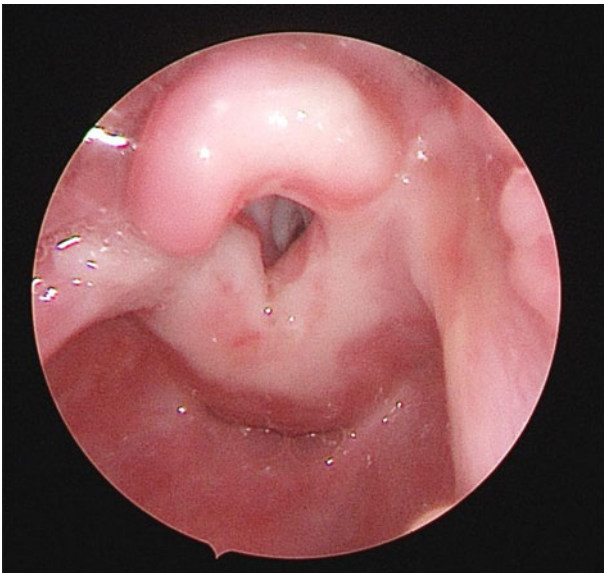


Fig. 10. Image of the patient's larynx following a trial of colchicine and azathioprine treatment.

Management

Prior to a diagnosis, the patient required a tracheostomy and gastrostomy. Once the diagnosis of Behçet's disease was made, colchicine and azathioprine were commenced. The patient is clinically improving on this treatment (Figure 10).

Discussion

Although the case did not exactly match Behçet's disease diagnosis criteria, it was agreed as reasonable, based on the pathological and clinical findings, to trial treatment for Behçet's and to titrate up the dose of azathioprine until an adequate treatment response is achieved.

A rare case of a lingual mass in a neonate

L Leach and N Jonas

From the Addenbrooke's Hospital, Cambridge

Introduction

Lingual thyroglossal duct cysts are a rare congenital malformation that can cause respiratory distress or even airway obstruction in a newborn. A rare variant of thyroglossal duct cysts, these cysts can be potentially difficult to manage. Normally located in the tongue base, we present an even rarer variant, that of an anterior lingual thyroglossal duct cyst.

Case report

A large oral cyst was noted on the 28-week antenatal ultrasound scan, which grew in proportion to the fetus (Figure 11). An elective caesarean section was performed at 39 weeks in view of airway obstruction concerns. At birth, a large lingual cyst was noted, and the neonate required oxygen to maintain saturations. At age one month, there were significant feeding difficulties secondary to rapid cyst enlargement, causing the tongue to be pushed superiorly against the hard palate (Figure 12).

Histological findings

Histology revealed a $48 \times 20 \times 5$ mm cyst lined by stratified squamous epithelium in some areas and bundles of smooth

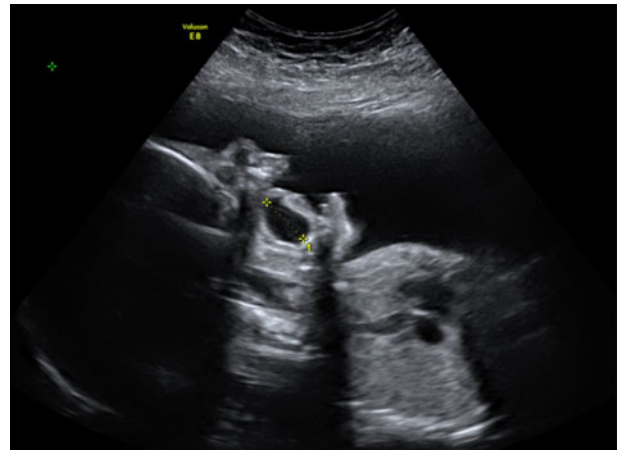


Fig. 11. Antenatal ultrasound at 28 weeks, demonstrating a large fluid-filled oral cyst.



Fig. 12. Pre-operative photograph of superior aspect of tongue demonstrating significant tongue swelling, resulting in the tongue protruding past the lips.

muscle in others, with small cystic structures lined by mucin-producing columnar cells. Dr Sandison explained that the lack of epidermoid structures or keratinisation indicated that this was a benign embryological cyst as opposed to a dermoid cyst.

Management

A total of 15 ml of dark brown fluid was aspirated from the cyst in clinic. This immediately improved feeding; however, this fluid had re-collected at age two months, filling the entire oral cavity. Further aspiration was performed and the patient underwent surgical excision of the cyst from the anterior tongue to the foramen caecum.

Discussion

It was suggested that most patients with lingual thyroglossal duct cysts should be treated as in-patients given the potential of airway obstruction. Mr Rothera (Manchester) advised early excision of such cysts before the patient is discharged from hospital.

Conclusion

As demonstrated with this case, aspiration of the cyst is useful for immediate symptom relief, but is often associated with recurrence. For definitive treatment, surgical management is required.

A rare, multifactorial presentation of hearing loss

K Fraser-Kirk, H Powel and I Pai

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

Introduction

Hajdu–Cheney syndrome is an extremely rare, autosomal dominant disorder of connective tissue. It is characterised by severe osteoporosis and excessive bone resorption (acroosteolysis). This results in the shortening of distal bones such as fingers and toes, and progressive deformation of axial bones including the skull base. Hearing loss in Hajdu–Cheney syndrome has been reported as multifactorial, due in part to remodelling of the middle ear and ossicular chain, and in part due to alterations of outer and inner hair-cell structure. This report lends further support for the role of VIIIth cranial nerve traction injury due to mechanical stretch between the brainstem and the internal auditory canal.

Case presentation

A 20-year-old female presented with mild mixed bilateral hearing loss in the setting of Hajdu–Cheney syndrome, with associated skull base instability. Audiometry revealed a minor right-sided conductive hearing loss with accompanying mild high-frequency sensorineural loss. Lower cranial nerve symptoms caused by progressive skull base flattening and brainstem invagination prompted neurosurgical intervention. The left-sided sensorineural thresholds underwent dramatic stepwise reduction, stabilisation, and deterioration related to the removal of a halo fitted for craniovertebral stabilisation.

Radiological findings

Radiology revealed features characteristic of Hajdu–Cheney syndrome, including severe platybasia, with marked intracranial protrusion of the vertebral column and distortion of the cranial nerves.

Discussion

Hearing rehabilitation in Hajdu–Cheney syndrome should be adapted to the degree and type of loss, with particular attention paid to anatomical constraints, middle-ear status, skull thickness, distribution of wormian bone and bone quality for osseointegration. The changes to the skull base and cervical spine resulted in cranial nerve stretching. Fixation of the halo improved hearing because of reduced tension on the VIIIth cranial nerve, and subsequent removal of the halo resulted in reduced hearing levels.

Head and neck section

Miss Lisa Pitkin, Chairperson

A rare thyroid tumour in a myasthenic patient

A Maity, A Salem and A Ali

From the Royal London Hospital

Introduction

Mature adipocyte-containing thyroid tumour, a rare entity, has been mainly divided into thyrolipoma and thyrolipomatosis types.

Case report

A 40-year-old man was referred by neurology colleagues following an incidental finding of a right thyroid lump on a



Fig. 13. Axial computed tomography scan of the chest, showing the heterogeneous appearance of the thyroid with enlargement of the right thyroid lobe (arrow).

computed tomography (CT) scan of the chest. He had recently been diagnosed as seropositive for myasthenia gravis and had stage 4 disease at presentation. He was on pyridostigmine and prednisolone 10 mg daily. Clinical examination revealed a soft lump in the right thyroid lobe.

Radiological findings

The CT scan demonstrated extreme widening of the mediastinum and pericardium, with obliteration of the lower half of the right lung (Figures 13 and 14). The right thyroid lesion was 3 cm in diameter. T1- and T2-weighted magnetic resonance imaging showed fatty tissue extending in continuity from the thyroid to the mediastinum and left hemithorax.

Management

Following discussion by the thyroid and lung multidisciplinary team, the patient was pre-operatively optimised with intravenous immunoglobulin. Total thyroidectomy and radical thymectomy were performed. A 2.4 kg mass was excised as a single specimen. He had complete resolution of myasthenic symptoms, and his prednisolone was gradually reduced to 6 mg daily.

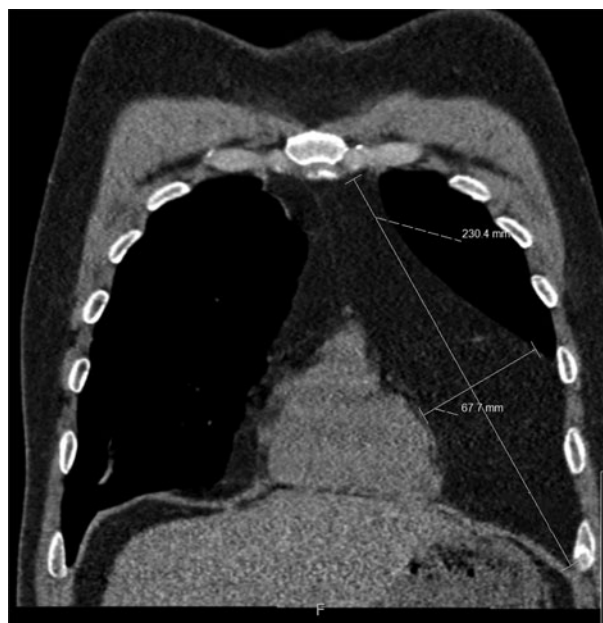


Fig. 14. Axial computed tomography scan of the chest, showing the tumour extending from the root of the neck to the left costophrenic angle, displacing the left lung.

Histological findings

Histology revealed: (1) diffuse effacement of the thyroid by intersecting lobules of mature adipocytes, which surrounded and engulfed residual follicles, imparting a nodular architecture, indicating thyrolipomatosis; and (2) a lipomatous tumour of the thymus, showing mature lobules of adipose tissue separated by fibrous septae, with intervening foci of mature thymic tissue, indicating thymolipoma.

Discussion

Dr Siddique explained that CT scans are good for the diagnosis of fatty tumours. Prof Sandison mentioned that histopathologists need to be careful, as foci of papillary thyroid carcinoma can be mixed in the tumour. The case represented a unique presentation of a rare benign tumour of the thyroid and thymus, of possible embryological origin.

Airway emergency in a young female with rare pathology

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

Introduction

Malignant peripheral nerve sheath tumours are rare neoplasms representing 5–10 per cent of all sarcomas. They commonly affect the head and neck. Triton tumours are malignant peripheral nerve sheath tumours with rhabdomyoblastic differentiation, which characteristically behave more aggressively than other malignant peripheral nerve sheath tumours.

Case report

A 26-year-old female presented to the emergency department with progressive dysphagia, difficulty breathing, and a 10-month history of a growing left-sided neck mass, previously investigated abroad with no diagnosis confirmed. She had undergone a tonsillectomy five weeks earlier in the UK privately. Flexible nasolaryngoscopy showed significant left-sided pharyngeal swelling obstructing her airway. She underwent emergency awake fibre-optic intubation to protect her airway.

Radiological findings

A computed tomography (CT) scan of the neck demonstrated a large, locally extensive, left-sided, deep cervical cystic and necrotic mass, causing medial displacement of the left pharyngeal wall and significant narrowing of the airway. A subsequent positron emission tomography CT image is shown in [Figure 15](#).

Histopathology findings

Biopsies confirmed a high grade 3 triton tumour, infiltrating fibrofatty tissue and entrapping neurovascular bundles ([Figure 16](#)).

Management

The patient was transferred to Stanmore Sarcoma Unit for neoadjuvant doxorubicin and ifosfamide chemotherapy. The mass has reduced in size during treatment; however, the prognosis remains poor.

Discussion

During the meeting, we discussed that the patient's chemotherapy management was appropriate, but had she presented sooner this would have been followed by surgery with curative intent.

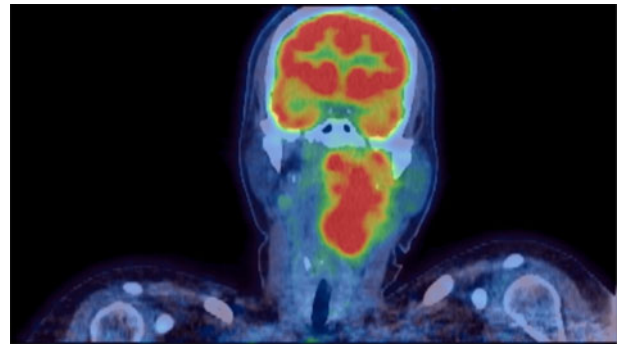


Fig. 15. Positron emission tomography computed tomography scan showing the metabolically active left neck mass extending to the skull base (approximately 9 cm × 6 cm), causing significant airway narrowing.

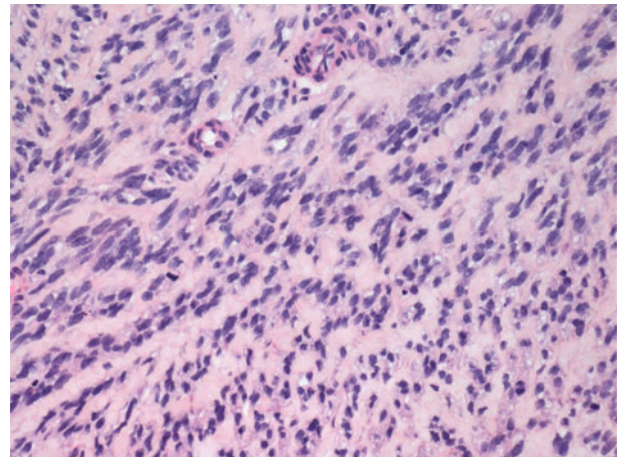


Fig. 16. Slide showing a high-grade tumour infiltrating fibrofatty tissue and entrapping neurovascular bundles. The tumour is composed of cellular fascicles of uniform spindle cells with wavy nuclei in a fibrous stroma. (H&E stain, magnification × 150)

Conclusion

The steps in emergency airway management are the same, regardless of the cause of obstruction. This case illustrates the importance of timely diagnosis, and how this affects potential treatment options and prognosis.

Longstanding progressive dysphagia and dysphonia: an unusual presentation of a soft palate tumour

C T Forde, S Ali and R Millard

From the Royal London Hospital

Introduction

Pleomorphic adenomas of the palate are the most common minor salivary gland tumour. The massive tumour in this case is both unusual in its size and in the surgical approach required to excise it.

Case report

A 53-year-old man presented with longstanding progressive dysphagia and a 'hot potato' voice. He was only managing a soft or liquid diet, and he slept with two pillows because of dyspnoea when lying flat. He also complained of significant weight loss. He reported that he had had a lump in his mouth for 30 years. He had previously presented to ENT services but did not attend follow up. Examination revealed a large oropharyngeal mass without lymphadenopathy.



Fig. 17. Sagittal T2-weighted magnetic resonance imaging scan demonstrating the size of the tumour in the craniocaudal dimension. P = posterior; F = feet

Radiological findings

Magnetic resonance imaging demonstrated a large tumour, which originated from the soft palate and extended to the larynx, measuring 9.9 cm in craniocaudal dimensions (Figure 17).

Histological findings

Examination revealed a macroscopically encapsulated and lobulated tumour measuring 9.5 × 7.5 × 3.9 cm. Microscopic findings were consistent with a pleomorphic adenoma.

Management

The patient underwent an awake tracheostomy, prior to a lip split mandibulotomy and mandibular swing with excision of the tumour. Following decannulation, his dyspnoea and speech improved, but he required extensive swallow rehabilitation.

Discussion

Dr Sandison demonstrated a myxoid tumour, with myoepithelial, tubular epithelial and chondromyxoid elements. Ms Pitkin agreed with the surgical approach taken in order to excise this tumour given its size.

Conclusion

Excisional biopsy is necessary to provide an accurate histopathological diagnosis. Careful pre-operative planning should be undertaken to determine the best surgical approach to ensure complete excision.

An unusual cause of sore throat in a 25-year-old patient

M Brimiouille, E Molena and L Pitkin

From the Frimley Park Hospital, Frimley

Case report

A 25-year-old Kenyan female presented with a year's history of unilateral throat pain and dysphagia. She denied a change in voice, otalgia, weight loss or anorexia. She had received several courses of antibiotics and nasal sprays from the general practitioner, with no improvement. She was an ex-smoker of three years. Oropharyngeal examination revealed a large mass on the left posterior tonsillar pillar, extending to the soft palate and the uvula. Flexible laryngoscopy showed involvement of the posterior pharyngeal wall, the left aspect of the epiglottis and left aryepiglottic fold, with superficial ulceration. A biopsy was taken in clinic.

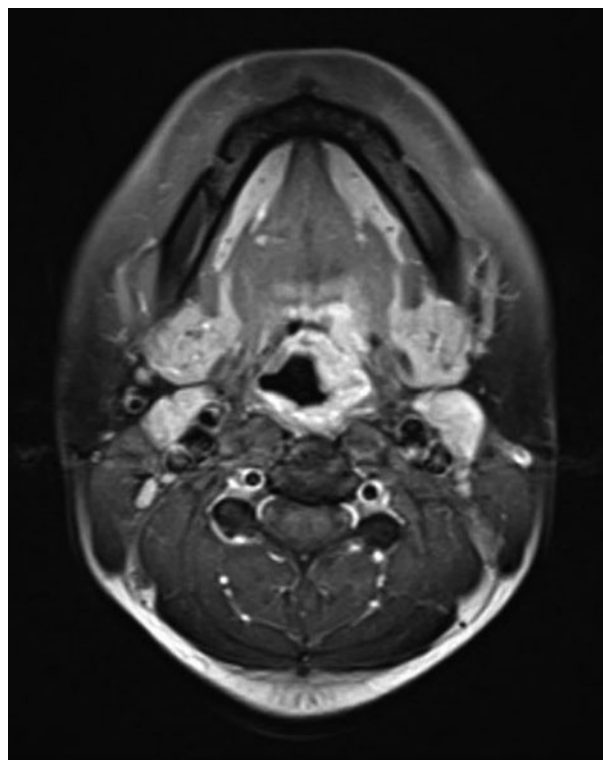


Fig. 18. Axial magnetic resonance imaging scan of the neck showing a large tumour of the oropharynx, crossing the midline and extending to involve the left aryepiglottic fold and the uvula, with bilateral cervical lymphadenopathy.

Radiological investigations

A magnetic resonance imaging scan of the neck was performed urgently. This revealed asymmetry of the mucosa of the posterolateral pharyngeal wall, extending down to the hypopharynx and larynx (Figure 18). A computed tomography scan of the chest excluded pulmonary disease.

Histological investigations

Histological analysis showed a fibro-inflammatory lesion with epithelioid granulomas, necrosis and multinucleate giant cells. There was no evidence of malignancy. Ziehl-Neelsen staining was negative.

Management

Tuberculosis was suspected, and a further biopsy was taken for microbiological analysis. The patient was referred to a respiratory physician and received empirical anti-tuberculous therapy.

Microbiological investigations

Staining for acid-fast bacilli was negative, but the culture was positive for mycobacterium tuberculosis after three weeks.

Outcome

At three months, the mass had disappeared and the symptoms had completely resolved.

Discussion

Laryngeal tuberculosis is not often considered when a patient presents with laryngeal symptoms. This case report aims to increase awareness of it, and to prompt the clinician to: investigate tuberculosis exposure, order appropriate investigations and initiate treatment without delay.