

## Proceedings of the 141st Semon Club, 2 June 2011, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Chairman: Miss Elfy B Chevetton, 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 Toby Moorhouse for 'Persistent dysphonia in a 35-year-old'.

### Professor Leslie Michaels retires from the Semon Club

A HALL

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

The Semon club had the benefit of Professor Leslie Michaels' expert pathology opinion for over 42 years, before his retirement from this role following the 140th Semon Club meeting in November 2010.

Professor Leslie Michaels has had a long and distinguished career as a Professor in Head and Neck Pathology, continuing long past his official retirement in 1990. He was Professor of Pathology at the University College London Hospital Institute of Laryngology and Otology and at the Royal National Throat, Nose and Ear Hospital.

A celebration was held to honour Professor Michaels' service to the Semon Club, at the Royal Society of Medicine after the 142nd Meeting in November 2011.

### Otology and skull base section

Chairman: Professor Michael Gleeson

#### A rare skull base tumour: benign angiomyxomatous lesion of the central nervous system presenting in the temporal bone

E Yeung, C Butler, S Saeed

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

This is a case of a very rare skull base tumour. A 45-year-old woman presented with a facial nerve palsy and was diagnosed with an extensive skull base tumour. A ventriculoperitoneal shunt was inserted due to rising intracranial pressure causing seizures. The lesion was found to be protruding through the left ear canal, and the patient was referred by the neurosurgeons for a diagnostic biopsy.

A magnetic resonance imaging scan showed a large, solitary, hypervascular skull base mass with its centre in the destroyed left petrous temporal bone, causing midline shift to the right. Differential diagnoses included haemangiopericytoma, chondrosarcoma and malignant meningioma.

Results of the initial histological examination were consistent with a cellular, myxoid lesion with no cellular atypia or necrosis and a low proliferation rate, consistent with a benign lesion. However, a histological review concluded this case to be a low grade angiomyxomatous lesion with similar features to a benign angiomyxomatous lesion of the central nervous system. The lesion did not fall into any currently defined sarcoma category.

The case was discussed at the head and neck multidisciplinary team meeting. Primary surgical resection was considered; however, given the anatomical location, tumour extent and histological features, radiotherapy was deemed to be the optimum treatment option.

This case provides an example of a very rare, unclassifiable skull base tumour that proved to be a radiological and histological diagnostic challenge. At the meeting, Professor Gleeson suggested that embolisation might be a treatment option in these vascular tumours to avoid the risk of radiation to the brainstem. Given the rarity of such tumours, treatment options remain controversial and long-term results are largely unknown.

#### Bilateral, sudden sensorineural hearing loss associated with urticarial vasculitis

A C Hall, A C Leong, A Fitzgerald-O'Connor

From Guy's and St Thomas' Hospital, London

#### Introduction

Following a severe respiratory tract infection, a 53-year-old farm labourer presented with vertigo and bilateral, severe, sudden-onset sensorineural hearing loss. This did not improve with a course of oral steroids.

#### Case report

Adult-onset Muckle–Wells syndrome was eventually suspected due to the combination of the patient's otological symptoms together with his purpuric and urticarial rash of the lower limbs, recurrent joint pains, anaemia, and raised C-reactive protein level.

Genetic analysis of the *NLRP3* gene demonstrated a mutation indicative of the syndrome, despite the fact that the patient had 14 siblings without symptoms. Seven years

later, he was still suffering with bilateral, profound sensorineural hearing loss along with disequilibrium and tinnitus.

#### *Histological findings*

Biopsies of the urticarial rash demonstrated features of cutaneous small vessel leukocytoclastic vasculitis. This is a type III hypersensitivity reaction with antigen–antibody complexes deposited in the vascular lumen.

#### *Radiological findings*

Computed tomography and magnetic resonance imaging scans demonstrate severe, right-sided labyrinthine ossification. On the left, there was sparing of the basal turn of the cochlea, with slightly less marked ossification of the semicircular canals.

#### *Management*

The patient underwent left-sided cochlear implantation due to his inability to function with conventional hearing aids. This was completed without complication, despite the highly unusual nature of this case.

#### *Discussion*

Professor Gleeson discussed the need for genetic testing in the offspring of both the patient and his unaffected siblings. This is especially important given a recent case report showing reversal of sensorineural hearing loss through the use of interleukin 1 $\beta$  inhibitor (anakinra) in a patient with early Muckle–Wells syndrome.

### **Visual loss following acute otitis media**

A Al-Rikabi, J Addams-Williams, S Berry  
From the Royal Glamorgan Hospital, Llantrisant

#### *Introduction*

Intracranial complications following acute otitis media are rare. Otitic hydrocephalus involves increased intracranial pressure without radiological features of hydrocephalus.

#### *Case report*

An 11-year-old boy presented with right-sided otalgia and discharge following an upper respiratory tract infection. He was treated with intravenous co-amoxiclav and topical ciprofloxacin drops for two days. Six days after discharge from hospital, he was readmitted with dizziness and frontal headache. Examination revealed a right aural polyp.

#### *Radiology*

A computed tomography (CT) scan showed a right middle-ear and mastoid infection and thrombosis of the right internal jugular vein extending into the sigmoid sinus, with no signs of hydrocephalus.

#### *Management*

A cortical mastoidectomy was undertaken, the sigmoid sinus was opened and free blood noted.

Two days later, the patient experienced blurred vision. An ophthalmology review confirmed the presence of papilloedema. A CT scan of the head showed no signs of raised intracranial pressure. The patient was started on acetazolamide and heparin. An initial lumbar puncture drained off 50 ml of cerebrospinal fluid (leaving a closing pressure of 40 cm

H<sub>2</sub>O). Repeated lumbar puncture 3 days later, performed due to deterioration of vision, revealed an opening pressure of 75 cm H<sub>2</sub>O.

A ventriculoperitoneal shunt was placed by the neurosurgeons, and the patient was discharged on Clexane<sup>®</sup>.

The patient required two ventriculoperitoneal shunt revisions due to increasing headache and vomiting. Eventually, a lumbar peritoneal shunt was inserted. On discharge, there was significant and irreversible visual loss.

#### *Conclusion*

It is accepted that otitic hydrocephalus is a syndrome of decreased cerebrospinal fluid absorption associated with sinus thrombosis. There is agreement on treatment with antibiotics, but the role of surgery and anticoagulation are controversial. Professor Gleeson stated that, as antibiotics and anticoagulants are used in the treatment of cavernous sinus thrombosis, the same should apply to treatment of lateral and transverse or sigmoid sinus thrombosis.

### **A perplexing papilloma**

A D Dragan, A C Leong, D Jiang  
From Guy's Hospital, London

#### *Introduction*

Squamous papilloma is a benign, exophytic proliferation of squamous epithelium which rarely occurs in the ear. To our knowledge, this is only the second reported case of malignant transformation of an aural squamous papilloma.

#### *Case report*

A 56-year-old, immunocompetent woman presented with a six-month history of recurrent, left-sided otorrhoea. An excision biopsy of a growth in her left ear canal indicated a benign squamous papilloma. The ear discharge persisted, with the development six months later of ipsilateral, complete facial palsy and profound sensorineural hearing loss. Examination under general anaesthesia revealed extensive papillomatosis of the ear canal; multiple biopsies showed chronic inflammation, and deeper biopsies were recommended.

#### *Radiological findings*

An initial computed tomography scan was normal, but repeated scanning six months later detected extensive bony erosion of the mastoid. A magnetic resonance imaging scan showed a mass extending from the outer and middle ear into the parapharyngeal space and foramen ovale.

#### *Histological findings*

Debulked tissue from the ear showed irregular islands and strands at the tumour interface, and a well differentiated squamous cell carcinoma was diagnosed.

#### *Management*

After discussion in the multidisciplinary team meeting, the tumour was deemed unresectable, and the patient underwent radical radiotherapy alone, with marked clinical improvement and resolution of the facial palsy.

#### *Discussion*

Dr Sandison emphasised the fact that squamous cell carcinoma of the ear canal is under-recognised by pathologists;

thus, whenever the index of clinical suspicion is high, repeated, deep biopsies should be performed. Professor Gleeson and Mr Oakley stated that the patient's human papilloma virus (HPV) status should be established to aid prognosis, and also to assist cost-benefit analysis of the use of the quadrivalent HPV vaccine. Professor Gleeson commented that the lesson here was to biopsy papillomas of the ear canal early.

### **Progressive sensorineural hearing loss in a patient with Hajdu–Cheney syndrome**

L A Jablenska, H R F Powell, S R Saeed

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

#### *Introduction*

Case report and discussion of progressive, unilateral sensorineural hearing loss in a patient with Hajdu–Cheney syndrome.

#### *Case report*

A 13-year-old girl, who had been diagnosed with Hajdu–Cheney syndrome at the age of nine years, was referred for specialist otolaryngology opinion following development of progressive, left-sided hearing loss. Other symptoms included left-sided tinnitus, facial and limb paraesthesia, nausea, and blurred vision. Examination revealed bilateral dull tympanic membranes and oblique external auditory canals. The rest of the ENT examination was unremarkable. Pure tone audiometry revealed profound, left-sided sensorineural hearing loss.

#### *Radiological findings*

The computed tomography and magnetic resonance imaging scans showed some of the recognised radiological features associated with Hajdu–Cheney syndrome. Due to marked basilar invagination, the petrous pyramids were obliquely angled and upward-sloping, leading to an unusual, stretched course of the VIIth and VIIIth cranial nerves.

#### *Conclusion*

We hypothesised that stretching of the VIIIth cranial nerve on the left side was the cause of the patient's progressive, profound sensorineural hearing loss. Having carried out a comprehensive literature review, we believe that this case represents the first report of unilateral sensorineural hearing loss and associated stretching of the ipsilateral VIIIth cranial nerve in a patient with Hajdu–Cheney syndrome.

### **Head and neck section**

Charman: Mr Richard Oakley

### **Destructive subglottic lesion presenting with stridor**

S Malakouti, B Fu, A Balfour

From the William Harvey Hospital, Ashford

#### *Introduction*

Diffuse large B-cell lymphoma classically affects older individuals and is linked with autoimmune conditions such as Sjögren's disease. We report an unusual case of lymphoma

presenting with worsening stridor in the absence of any lymphadenopathy.

#### *Case report*

A 68-year-old woman presented with a three-week history of increasing dyspnoea, dysphonia, stridor, halitosis and productive cough. She had been diagnosed with Sjögren's disease three years previously and was being investigated for 'Sjögren's related interstitial lung disease'. Flexible nasendoscopy showed marked subglottic oedema. A computed tomography (CT) scan and subsequent microlaryngoscopy, biopsy and tracheostomy were performed.

#### *Radiology*

The neck CT scan showed a bulky right vocal fold and subglottis, with destruction of the trachea below this level. The appearances were non-specific and the differential diagnosis included vasculitic, granulomatous and neoplastic lesions. Dr Connor agreed with the findings.

#### *Histopathology*

Dr Sandison commented that the subglottic lesion showed an abnormal large lymphoid cell infiltrate with large nucleoli that stained positive for B cell markers. Dr Sandison agreed with the diagnosis of a high-grade B-cell non-Hodgkin's lymphoma.

#### *Management*

The case was discussed within the multidisciplinary team meeting. The patient was diagnosed with stage IV diffuse large B-cell non-Hodgkin's lymphoma, and underwent the 'R-CHOP' (rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine (Oncovin) and prednisolone) chemotherapy regime.

#### *Conclusion*

Lymphoid malignancy is associated with autoimmune conditions. Our case demonstrated an unusual presentation of lymphoma with stridor and a destructive subglottic lesion in the absence of any cervical lymphadenopathy and 'B'-type symptoms. Mr Oakley suggested a possible viral aetiology.

### **An unusual mandibular lesion**

K Amin, B Fu, C Barbaccia

From the Medway Maritime Hospital, Gillingham

#### *Background*

Parathyroid adenoma is the commonest cause of primary hypercalcaemia and usually presents with symptoms and signs of hypercalcaemia ('bones, stones, abdominal groans and psychic moans'). This case report highlights an unusual presentation.

#### *Case report*

A 27-year-old woman presented with a painful left mandibular swelling, suspicious of neoplasia. A computed tomography (CT) guided biopsy was performed. Based on the histology result, the serum calcium concentration was assessed, confirming the presence of hypercalcaemia (3.16 mmol/l). Ultrasonography and a Sestamibi scan of the neck suggested a left inferior parathyroid adenoma, subsequently removed via selective parathyroidectomy.

### *Radiology*

A CT scan of the mandible showed extensive erosive lesions at the left second and third inferior molar roots, with extension to adjacent soft tissues. Ultrasonography demonstrated a hypoechoic lesion at the left inferior thyroid pole. Sestamibi scanning showed a focus of uptake and delayed retention at the same place, suggestive of abnormal parathyroid tissue.

### *Histopathology*

Dr Sandison commented that the mandibular biopsy showed a giant cell lesion which was expansile (as opposed to destructive) and showed no malignant features. The differential diagnosis included a giant cell tumour of the bone or a 'Brown tumour' of hyperparathyroidism.

The parathyroid specimen revealed a 2 cm, nodular, circumscribed tumour composed of nests and islands of chief cells, composed of connective tissue and packed with multinucleate giant cells, consistent with a parathyroid adenoma.

### *Conclusion*

Clinicians should bear in mind the varied presentation of parathyroid adenomas. Dr Sandison commented that Brown tumours of bone are not so commonly seen nowadays due to early detection of parathyroid adenomas in patients with hypercalcaemia.

### **Persistent dysphonia in a 35-year-old**

T E Moorhouse, S Berry, P H Brumwell

From the Royal Glamorgan Hospital, Llantrisant

### *Background*

Published data are scanty on the management of pleomorphic sarcoma of the vocal fold.

### *Case report*

A 35-year-old man presented with a six-week history of dysphonia; there was no odynophagia, dysphagia, otalgia or weight loss. He was asthmatic and an ex-smoker. Flexible nasendoscopy revealed a large, granular-looking polyp of the middle third of the right vocal fold; both vocal folds were mobile. He underwent microlaryngoscopy and excisional biopsy.

### *Radiological findings*

A computed tomography (CT) neck scan showed a well-defined, strongly enhancing lesion involving the anterior half of the right vocal fold (11 × 4.5 × 8 mm) with no extension into para-glottic fat. An ultrasound scan of the neck and CT scan of the chest and abdomen were normal.

### *Histological findings*

Histological examination revealed a polypoid tumour composed of large, pleomorphic, epithelioid and stellate cells, occasional multinucleate giant cells, scanty mitoses (some atypical), and focal hypercellularity. Our pathologists provided a diagnosis of superficial malignant fibrous histiocytoma (also known as pleomorphic sarcoma). Dr Sandison asked for the opinion of Professor Fisher, pathologist at the Royal Marsden sarcoma unit. He postulated that the lesion may be a sarcomatous squamous cell neoplasm. Dr Sandison suggested that the patient should be monitored for cervical lymphadenopathy in case the lesion was a

squamous cell malignancy (sarcomatous lesions being more likely to metastasise to the lungs).

### *Management*

Recurrence occurred within one month. Following discussion within the head, neck and sarcoma multidisciplinary team, the patient underwent right total cordectomy with CO<sub>2</sub> laser. A microlaryngoscopy performed six weeks later showed no further recurrence. The patient was reviewed regularly, and at the time of writing had a stable voice, was eating and drinking normally, and had returned to work.

### *Conclusion*

The management of head and neck sarcoma is unusual due to the anatomical limitations of excision. Such sarcomas should be treated specifically as they do not behave like squamous cell carcinoma, nor do they behave in the same way as sarcomas occurring elsewhere. Mr Oakley agreed that superficial malignant fibrous histiocytomas have unpredictable malignant potential, and that wide excision is recommended. Mr Oakley also felt that neo-adjuvant chemo-radiotherapy can be considered in unresectable cases, or if the resection margins are close. He also noted the importance of centralising services for rare tumours. Dr Connor agreed, and added that six-monthly CT scanning was appropriate for follow up.

### **A unique case of a submandibular tumour**

C Theokli, M Wisniewska, A Balfour

From the William Harvey Hospital, Ashford

### *Introduction*

Nuchal fibroma is a rare, benign, fibrous tumour most commonly described in the nuchal and interscapular areas. It typically affects individuals between the third and sixth decade of life, and there is a close association with diabetes and Gardner's syndrome. Originally described in 1988, this is the first documented report of nuchal fibroma affecting the submandibular gland.

### *Case report*

A 69-year-old man presented originally with a six-week history of having the sensation of food sticking in his throat, and subsequently developing a painless right submandibular swelling. He had type two diabetes and was an ex-smoker. Examination revealed a hard, mobile, 1.5 cm mass in the right submandibular gland. Fine needle aspiration cytology was performed but the sample was non-representative.

A computed tomography scan revealed a well defined, nodular, enhancing, mass lesion in the inferior pole of the right submandibular gland, measuring 18 × 15 mm. The patient subsequently underwent a selective level I and II neck dissection.

### *Histology*

Sections of the submandibular salivary gland contained a well circumscribed but unencapsulated nodule composed of hypocellular fibrous tissue containing small blood vessels and scattered adipose tissue. Immunohistochemical staining showed the lesion to be positive for beta-catenin and cluster of differentiation 34 protein, with no atypia or signs of malignancy; the appearance was suggestive of a nuchal-type fibroma.

### Discussion

In patients with such a diagnosis, it is important to exclude the rare but important association with Gardner's syndrome. Our patient underwent an oesophago-gastroscopy and was found to have adenocarcinoma of the oesophagus, which was subsequently treated. It was agreed that only with a sound knowledge base can such associations be recognised and effectively managed.

### Rhinology section

Chairman: Mr James Earnshaw

### Skull base aspergilloma following chemotherapy for acute lymphoid leukaemia

R Harris, A Toma, G I Smith

From St George's Hospital, London

### Introduction

Aspergilloma is a mass caused by fungal infection from spores that contaminate the upper respiratory tract. It can grow slowly and asymptotically in any body cavity, and patients tend to present late. Isolated sphenoid sinus aspergilloma is exceptionally rare; a small percentage of such cases present with symptoms caused by local destruction.

### Case report

A 57-year-old woman was treated with chemotherapy for acute lymphoid leukaemia. Three days later, she presented to the ENT clinic with a right lateral rectus palsy and altered olfaction. Rigid nasendoscopy revealed a firm, white mass in the post-nasal space.

### Radiological findings

Magnetic resonance imaging and computed tomography scans of the sinuses showed opacification of the right sphenoid sinus, destruction of the skull base and occlusion of the internal carotid artery.

### Histological findings

Histological examination confirmed the lesion to be an aspergilloma.

### Management

The patient was treated by an open resection of the skull base via an Altemir lateral facial swing approach. A necrotic section of sphenoid bone was excised and an intracranial abscess was irrigated. The skull base defect was reconstructed with a radial forearm flap, providing excellent functional and cosmetic results. This was a joint procedure performed by the maxillofacial and ENT teams.

### Non-healing ulcer on the nasal dorsum: a rare side effect?

F Salim, A Joshi, C Hopkins

From Guy's Hospital, London

### Background

Mucous membrane pemphigoid is a rare disease characterised by ulcerative and blistering lesions on the mucous membranes and skin.

### Case report

A 64-year-old man, known to have mucous membrane pemphigoid and multiple medical problems, presented in our ENT out-patient clinic with a 10-week history of a painful, non-healing ulcer on the dorsum of the nose.

### Management

Multiple biopsies of the nasal ulcer excluded a malignant aetiology. The presence of pemphigoid in other regions initially masked the diagnosis, but this aetiology was essentially ruled out following histopathological examination.

Eventually, it was noted that the patient was taking nicorandil, and this was thought to be the cause of the ulcer. Chronic, non-healing ulcers of the gastrointestinal tract and genitals are well recognised complications of nicorandil, a potassium channel blocker commonly used to treat angina. Significant improvement was noted following withdrawal of nicorandil.

### Histopathology

Histological examination of multiple biopsies was reported as showing non-specific inflammation and no signs of malignancy.

### Conclusion

This is the first documented case of nicorandil affecting the dorsum of the nose. This case highlights the importance of not overlooking therapeutic drugs as an aetiological factor.

### Recurrent epistaxis in a Jehovah's Witness

D Walker, I Pai, L Pitkin

From the Royal Surrey Hospital, Guildford

### Introduction

Mucosal melanoma presents a significant therapeutic challenge to the ENT surgeon. It behaves far more aggressively than its cutaneous counterpart and carries a very poor prognosis, with many patients presenting with established metastatic disease.

### Case

A 72-year-old woman was referred to the emergency ENT clinic with a two-month history of intermittent epistaxis. She had no history of sinonasal problems and no sinister symptoms. She was a Jehovah's Witness. On examination, a black, friable polyp was seen in the right nasal cavity. The examination was otherwise normal.

### Radiology

A magnetic resonance imaging scan of the head showed mucosal thickening in the right nasal cavity with obstruction of the osteomeatal complex. Dr Siddiqui confirmed that there were no features of frank invasion.

### Histopathology

Histological examination of a biopsy showed malignant melanoma.

### Management

The patient underwent a septectomy, partial medial maxillectomy and modified radical neck dissection (type III). The tumour was fully resected (closest margin 6 mm), and all 24 lymph nodes removed were negative. Post-operatively, the multidisciplinary team decided to opt for close observation with no adjuvant therapy. Unfortunately, two months after surgery the patient developed local recurrence, and underwent debridement and post-operative radiotherapy.

### Conclusion

Mr Earnshaw suggested from the radiological findings that the tumour would have been suitable for endoscopic resection; however, he accepted that an open approach was the safer option regarding haemostasis, given the patient's religious objection to blood transfusion. Mr Earnshaw mentioned the possibility of sentinel node biopsy as a precursor to neck dissection. We felt that, given our experience with over 50 cases of mucosal melanoma, with 12 per cent metastasising to the ipsilateral lymph nodes, neck dissection was indicated. Mr Earnshaw agreed that local control was the key to reducing recurrence, and that patients should receive post-operative radiotherapy however adequate the resection margin.

### Complete nasopharyngeal stenosis after radiotherapy for squamous carcinoma of the oropharynx

C Igwe, C Hopkins, D Roberts

From Guy's Hospital, London

### Introduction

The development of acquired choanal atresia following radiotherapy for oropharyngeal cancer is a rare and poorly described condition. Consequently, its management remains contentious.

### Case report

A 69-year-old man presented with an unknown primary, later staged as tumour-node-metastasis (TNM) stage T<sub>x</sub> N<sub>2</sub> M<sub>0</sub>. The tumour was successfully treated with wide local excision of a right neck lump and panmucosal radiotherapy.

Six years later, he was diagnosed with a second primary cancer of the right tonsillar fossa, staged as T<sub>2</sub> N<sub>0</sub> M<sub>0</sub>. On this occasion, he was treated with transoral laser resection. Following treatment, he complained of bilateral nasal obstruction and anosmia. Complete choanal atresia was confirmed by flexible nasendoscopy.

Endoscopic surgery was performed to open the stenosis, and a modified endotracheal tube was used to stent the opening for nine days. After the procedure, the patient had excellent improvement in nasal airflow. However, three months later he was noted to have partial restenosis of his choanae.

### Radiological findings

A computed tomography scan confirmed the presence of a soft tissue stenosis.

### Histological findings

Histological examination of a biopsy of the nasopharyngeal stenosis was reported as showing fibrosis with focal elastosis and a light inflammatory infiltrate.

### Management

An oesophageal balloon catheter was placed within the restenosed area and inflated to a pressure of 3 atmospheres. This method successfully restored nasopharyngeal patency.

### Conclusion and lessons learnt

Management of this condition is an ongoing challenge. However, meeting attendees agreed that balloon catheter dilatation is effective and is a valuable alternative to repeated surgery. This technique can be easily performed in the outpatient department, and can be repeated as many times as necessary to achieve the desired effect. Some colleagues commented that they would implement this technique in future.

### Sudden increase in headache in a patient with a ventriculoperitoneal shunt

D Bangaru-Raju, C Hopkins

From Guy's Hospital, London

### Introduction

Delayed, symptomatic pneumocephalus following ventriculoperitoneal shunt placement is very rare. The only three reported cases were all secondary to a frontal sinus fistula with cerebrospinal fluid (CSF) rhinorrhoea. In most cases, the air entry is via a skull base defect (i.e. postsurgical communication, trauma or thinning crania). In our patient, massive pneumocephalus occurred following ventriculoperitoneal shunt insertion for hydrocephalus. Bone erosion of the posterior wall of the frontal sinus was likely to have resulted from a period of untreated intracranial hypertension.

### Case report

A 19-year-old woman presented with increasing frontal headache, fever, photophobia, nausea and vomiting. There was no CSF rhinorrhoea. On examination, there was diplopia on extreme lateral gaze. A lumbar puncture excluded meningitis, and there was no clinical indication of sinusitis. Notably, at the age of 14 years the patient had had a ventriculoperitoneal shunt placed to treat hydrocephalus following an intracranial haemorrhage; at the age of 16 years, this shunt had needed revision due to persistent hydrocephalus.

### Radiological findings

A computed tomography scan demonstrated an intracranial, extra-axial gas collection measuring 13 mm deep, with a wide frontal recess and a well pneumatized frontal sinus. Most of the postero-superior wall and roof of the frontal sinus was eroded. There was no evidence of chronic sinusitis.

### Management

The immediate management was conservative. Plans were made for elective surgery.

### Discussion

The optimum management of such cases remains contentious. In patients without CSF leakage, the treatment is limited to primary repair of bony and dural defects with craniotomy to plate the defect posteriorly, together with obliteration or cranialisation of the frontal sinus. In the presented patient, it was planned to undertake a bicoronal flap and frontal sinus cranialisation.

## Paediatric section

Chairman: Mr Ian Hore

### Pre- and post-natal management of a fetal neck mass

L Guthrie, N Eze, I Hore

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

#### Background

Cervical teratomas are rare, congenital tumours derived from all three embryonic germ cell layers. They are associated with a high mortality rate due to compression or distortion of the airway, which can result in airway obstruction at birth.

#### Case report

A 30-year-old woman presented in the second trimester of pregnancy with a large fetal neck mass detected on a routine antenatal ultrasound scan. A fetal magnetic resonance imaging (MRI) scan identified a large cystic component to the mass. This was aspirated in utero to decompress the mass prior to delivery. A caesarean section was planned with the involvement of a multidisciplinary team.

Due to early rupture of membranes and premature labour, an emergency caesarean section was performed. The infant developed upper airway obstruction minutes after delivery. Subglottic resistance prevented full intubation. Immediate aspiration of the cystic component sufficiently decompressed the mass to allow successful intubation. An MRI was performed soon after birth, followed by excision of the mass 3 days post-natally.

#### Radiology

The MRI demonstrated a complex, multilocular, solid and cystic mass within the left side of the neck, displacing the aerodigestive tract across the midline and the carotid sheath posteriorly.

#### Histology

Histopathological assessment showed a completely excised, immature cervical teratoma.

#### Management

This case was managed as an emergency by a multidisciplinary team comprising otolaryngologists, obstetricians, paediatricians, paediatric surgeons, a neonatologist and a paediatric anaesthetist.

#### Conclusion

Such cases would ideally be managed with an ex utero intra-partum treatment procedure at a specialist centre. However, this was prevented in this case by the emergency presentation. Early involvement of a multidisciplinary team was fundamental to this patient's successful outcome.

### A rare cause of paediatric airway obstruction

J M Bernstein, I A Bruce, M P Rothera

From the Royal Manchester Children's Hospital

#### Introduction

We present a case of paediatric airway obstruction with an extremely rare cause.

#### Case report

A 14-month-old girl presented to the Royal Manchester Children's Hospital with a 7-day history of progressive, biphasic stridor. Direct laryngo-tracheo-bronchoscopy revealed a tumour obstructing 70 per cent of the upper tracheal lumen. The oesophagus was free of tumour, with mobile mucosa over the lesion. The tumour was debulked and sent fresh for histology. Post-operatively, the child was managed in the intensive care unit. She was extubated after 4 days and weaned off steroids after two weeks.

#### Radiological findings

A computed tomography scan confirmed the presence of a tumour located between the trachea and the oesophagus.

#### Histological findings

Histological and immunohistochemical analysis revealed an embryonal rhabdomyosarcoma.

#### Management

Nine cycles of induction chemotherapy with ifosfamide, vincristine and actinomycin were given. After the first three cycles, tumour reduction was demonstrated on a magnetic resonance imaging scan. Residual tumour was debulked via an external approach. Following this, proton beam radiotherapy with concurrent chemotherapy was administered in Florida, USA.

Proton beam therapy involves the release of ionisation over a few millimetres (the 'Bragg peak'), with minimal distal ionisation, which helps to reduce side effects. There are currently plans to develop a proton beam therapy centre in the UK.

#### Conclusion

Three other cases of tracheal rhabdomyosarcoma have been reported in the literature; however, ours is the first reported case to have been treated with proton beam therapy. Approximately 35 per cent of cases of rhabdomyosarcoma occur in the head and neck.

### Tuberculosis: the great mimic, again? A case of tuberculosis with combined cholesteatoma

R Cetto, S Haikel, S Abramovich

From St Mary's Hospital, London

#### Introduction

Primary tuberculosis (TB) of the middle ear is a rare condition.

#### Case report

A three-year-old boy presented with a one-day history of a right facial nerve palsy and a two-year history of intermittent right otalgia and otorrhoea. He was treated with intravenous acyclovir for suspected Ramsey-Hunt syndrome, despite a lack of vesicles. Both his mother and father were known to be infected with human immunodeficiency virus (HIV). The child was negative for HIV and had no known TB contact. Three months prior, he had developed a right

facial nerve palsy which had resolved with oral steroids. Microscopic ear examination revealed an attic defect with squamous debris consistent with cholesteatoma. Audiometry revealed a 50 dB threshold in the right ear; the left ear was normal.

#### *Radiological findings*

A computed tomography scan showed opacification of the right petromastoid air cells and the middle-ear cleft, with bony erosion, consistent with cholesteatoma.

#### *Histological findings*

Following tympanomastoidectomy, a specimen macroscopically consistent with cholesteatoma was sent for histological analysis, microscopy and culture. Histological examination showed keratinising squamous epithelium and acid-fast bacilli. *Mycobacterium tuberculosis* was subsequently cultured.

#### *Management*

Anti-tuberculous therapy was commenced. The patient recovered his facial nerve function and his hearing levels improved.

#### *Conclusion*

This patient's clinical, radiological and histological findings were consistent with cholesteatoma, and he had no symptoms of pulmonary or extra-pulmonary TB, usually a preceding condition. Miss Pitkin enquired as to why a specimen had been sent for microscopy and culture, and Mr Haikel explained that, at operation, an excess of granulation tissue had been noted in the mastoid air cells. This case highlights the need for a high index of clinical suspicion of TB as a cause of a chronically discharging ear, especially in our ethnically diverse inner city area. If recognised and treated early, complications such as facial nerve paralysis or permanent hearing loss may be avoided.

### **An usual cause of neonatal stridor with associated cyanosis**

T S Ahmed, B Okoye, H Daya  
From St George's Hospital, London

Stridor in infants is a common presentation in paediatric otolaryngology. Cyanosis can be associated with stridor and may indicate tracheal pathology. Bronchogenic cysts are congenital lesions of the primitive foregut and are among the most common primary cysts of the mediastinum.

We report a three-week-old infant who had intermittent stridor from birth, which subsequently became constant and was associated with cyanotic episodes on feeding. The infant presented acutely to hospital with respiratory distress requiring intubation and transfer to the paediatric intensive care unit.

Micro-laryngoscopy and bronchoscopy the following day showed indentation of the posterior tracheal wall with partial luminal collapse. Oesophagoscopy showed compression of the anterior wall of the cervical oesophagus. Contrast-enhanced computed tomography imaging showed a large, partially septated, fluid-filled cyst in the right side of the neck, passing into the superior mediastinum between the trachea and oesophagus and extending inferiorly to the right main bronchus.

The lesion was excised via a transverse neck incision, delivering the thoracic component into the neck. The child was subsequently successfully extubated, with resolution of his stridor. He had a hoarse cry as a result of right recurrent laryngeal nerve damage.

Histological examination showed a cyst lined with respiratory epithelium and including cartilaginous, smooth muscle and mucous glandular components, in keeping with a bronchogenic cyst.

This case illustrates the need to consider this diagnosis in infants presenting with stridor and cyanotic episodes. Surgical excision is recommended and should be curative.

### **Submandibular mass in an infant**

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#### *Background*

Sialoblastomas are rare congenital tumours originating from epithelial cells. They can be locally invasive and may metastasise. Although they predominantly arise within the parotid gland, they have also been described in the submandibular gland. Surgical excision is the treatment of choice. Local recurrence has been described in five of the 24 cases reported in the English language literature. Radiotherapy and chemotherapy are reserved for incompletely excised or unresectable tumours.

#### *Case report*

A five-month-old boy presented with a three-month history of a neck mass. He was otherwise well. Examination revealed a firm, mobile mass within the right submandibular triangle. The remainder of the ENT examination was unremarkable. Blood tests and viral screening tests were normal.

#### *Radiological findings*

A magnetic resonance imaging scan showed a 3 cm, homogeneous, enhancing mass arising from the submandibular gland and invaginating the tail of the parotid.

#### *Histopathological findings*

Fine needle aspiration cytology showed a cellular aspirate with features of a primary salivary gland neoplasm.

Histopathological examination suggested a completely excised sialoblastoma. There appeared to be some perineural association but no nerve invasion. Therefore, Dr Sandison felt that perineural spread would be unlikely in this case; however, there was a risk of local or regional recurrence, and she suggested monitoring the patient with ultrasound scans.

#### *Management*

The child underwent excision of the right submandibular gland and surrounding level two lymph nodes. Four months post-operatively, the child was well with no evidence of recurrence.

#### *Conclusion*

It was agreed that a repeated ultrasound scan six months post-operatively would be the most appropriate way to monitor for recurrence.