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126th Meeting

OTOLARYNGOLOGY DEPARTMENT, GUY'S HOSPITAL, LONDON

Chairman: Ms E. B. Chevretton, Consultant Otolaryngologist, Guy's & St. Thomas' Hospitals
Pathologists: Professor L. Michaels, Consultant Pathologist, University College London; Dr A. Sandison, Consultant Pathologist, Charing Cross Hospital, London
Radiologist: Dr G. Rottenberg, Consultant Radiologist, Guy's & St. Thomas' Hospitals
Minutes: Mr A. Bibas, Specialist Registrar in Otolaryngology, Guy's & St. Thomas' Hospitals

Case 1: An unusual cause of stridor
W. Kennedy, S. Ashraff, G. Rowlands (Whipps Cross Hospital, London)

The case of a 25-year-old Somali woman with a four-day history of stridor preceded by hoarseness and dyspnoea was presented. There was no associated trauma or URTI. She had been in the UK for five years. Examination revealed a small post-nasal space mass and a subglottic mass causing Grade III stenosis. Microlaryngoscopy and tracheostomy with debulking of the subglottic lesion were perfomed and Augmentin and dexamethasone were commenced. Histology taken at admission and at three weeks showed chronic inflammation only. All other relevant investigations were normal. The patient improved on steroids and was decannulated. Over the next few months the patient complained of increasing nasal obstruction. The PNS mass was removed and Dr R. Owen (Consultant histopathologist) reported that Mikulicz cells were evident. The relevant organisms were best seen with Warthin Starrey silver stains. The diagnosis of rhinoscleroma was made and the patient commenced on long-term ciprofloxacin.
Discussion: This case demonstrates that rhinoscleroma should be considered in the differential diagnosis of laryngeal lesions causing stridor in patients from endemic areas. Professor Michaels held the opinion that had immunohistochemical staining been done earlier, we would have been able to confirm the presence of *Klebsiella rhinoscleromatis* sooner.
Acknowledgements
Mr G. Kenyon, Mr R. Dasgupta

Case 2: Vernet's syndrome following blunt head injury
R. Hornigold, N. Donnelly, M. L. Harries (Brighton and Sussex University Hospitals Trust)

A 55-year-old gentleman presented to the out-patient department, complaining of a five-month history of a hoarse voice. He had a history of alcohol abuse and sustained a fall directly onto his head whilst attempting to climb in through a caravan window. He denied loss of consciousness and was confident that he immediately developed a hoarse voice, loss of power in his right shoulder and problems swallowing following the injury. He had noticed no improvement in symptoms in the months preceding presentation. On examination, he was found to have marked wasting of the right trapezius muscle and an absent gag reflex. Tongue movement was normal, as were pupillary reflexes. His voice was poor, and on examination of his larynx he was found to have a paralysed, atrophic right vocal fold. These findings were consistent with Vernet's syndrome, a jugular foramen syndrome consisting of palsies of cranial nerves IX, X and XI. Computed tomography (CT) scans of the brain to diaphragm, including neck and skull base revealed no fracture or lesion of the jugular foramen, no intracranial haemorrhage and no other abnormality. He did not tolerate magnetic resonance (MR) imaging. He underwent medialization of his right vocal fold (thyroplasty) in October and has had an excellent improvement in voice.
Discussion: Cranial nerve palsy to nerves IX, X, XI and XII have been reported in basal fractures of the skull, especially those involving occipital condyle fractures. There have been two reports in the literature of Vernet's syndrome secondary to blunt head injury, both of which had obvious skull base fractures on CT. There was no evidence of any haematoma, fracture or other abnormality in this patient. Temporary jugular foramen syndrome secondary to subdural haematoma encroachment after head injury has been seen by members of our department; however, this patient has made no recovery six months post-injury. It was suggested that he undergo further imaging, for example positron emission tomography (PET) or open MR.

Case 3: An unusual parapharyngeal mass
D. E. J. Whitehead, A. Sandison, P. Clarke (Charing Cross Hospital, London)

A 34-year-old man had presented two years ago with an enlarged right tonsil and an enlarged lymph node in the right neck. Histology of the tonsil showed reactive lymphoid hyperplasia. The neck node showed a necrotizing xanthogranulomatous inflammation of uncertain aetiology. Four months later he returned with dysphonia and a right-sided vocal fold palsy. Review of CT imaging revealed a soft tissue mass in the parapharyngeal area. Further exploration and biopsy of a lymph node showed the same xanthogranulomatous inflammation. On developing a IX, X and partial XII nerve palsy he was referred to Charing Cross Hospital for further exploration. The infra-temporal fossa was explored and an infiltrating mass encircling the internal carotid, IX, X, and XII cranial nerves was debulked. Frozen section histology was suggestive of a malignant nerve sheath tumour. However, immunohistochemistry on fixed material showed the tumour was a high grade diffuse large B-cell non-Hodgkin's lymphoma arising in soft tissue.
Discussion: The tonsil enlargement that was initially reported may well have been caused by the parapharyngeal mass medializing the tonsil. The two lymphoid biopsies (tonsil and lymph node) did not assist in the diagnosis, as the lymphoma was extra nodal. This is an unusual presentation in soft tissue and raised the possibility of retroviral infection, which later proved negative. The xanthogranulomatous inflammation still remains unexplained. The biopsy of the mass encasing the carotid through a far lateral approach was important in providing a representative histological sample. The use of immunohistochemistry was crucial in providing the diagnosis.

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Case 4: An unusual cause of conductive hearing loss
A. Joshi, S. Habashi, A. Al Ayoubi (Chase Farm Hospital, London)

A 41-year-old lady presented with a two-year history of right-sided hearing loss. The otoscopic appearance resembled otitis externa with a degree of meatal stenosis. There was a soft cystic-looking mass arising from the posterior meatal wall occupying the entire external auditory meatus (EAM). A CT scan confirmed this and was reported as showing a chronic inflammatory process. Incision was attempted under anaesthetic but merely resulted in bleeding. Subsequent exploration was undertaken via a post-auricular approach. The mass was filling the entire EAM, involving most of the tympanic membrane (TM), epitympanum and additus. The bony wall of the EAM was partially eroded. The entire mass was resected with preservation of the ossicular chain. The posterior wall defect was reconstructed with conchal cartilage and the TM and EAM grafted with temporal fascia. Histology was reported as meningiothelial meningioma.
Discussion: Professor Michaels agreed that this was an extremely rare case of extra-cranial temporal bone meningioma. Such tumours constitute just one to two per cent of all temporal bone tumours. As recently reported by Thompson *et al*., 2003, they are commoner in females (2:1) with a mean age at presentation of 50 years and a mean symptom duration of two years. Deafness, otalgia and dizziness are the chief symptoms. Complete surgical excision is the treatment of choice. It was agreed during the meeting that we should re-explore our patient with a view to more radical clearance, including the removal of the incus and malleus. To conclude, meningiomas in the ear are very rare and can mimic an inflammatory process. When complete surgical clearance is possible prognosis is excellent.
References
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Case 5: Worsening shortness of breath associated with stridor and voice change due to an idiopathic subglottic stenosis
H. R. F. Powell, M. J. Gleeson (Guy's Hospital, London)

A 54-year-old lady presented in September 2001 with progressive dyspnoea that was attributed to asthma but failed to improve significantly with inhaled steroids. A subglottic narrowing was seen at bronchoscopy that prompted referral for laryngoscopy and biopsy in February 2003. Multiple biopsies failed to identify the cause and as her condition continued to deteriorate she was transferred to Guy's Hospital under the joint care of the Thoracic and ENT surgeons. Further endoscopy and biopsies failed to advance the diagnosis. The subglottic pathological tissues seemed to contain a polyclonal lymphocytic infiltrate. No evidence of a lymphomatous process could be found. She was treated empirically with azathioprine 50 mg o.d. and prednisolone 20 mg o.d. and appeared to improve for a short period of time. By May 2003 her airway obstruction had increased and this was now associated with slight huskiness of voice and the globus sensation. Fibre-optic nasolaryngoscopy showed a posterior, Cotton's grade I subglottic stenosis with the airway about 75 per cent patent. CT and PET scanning were both normal, autoantibody titres were repeatedly normal; the only positive test was a mildly raised LDH level (non-specific marker of possible lymphoma). Bone marrow trephine biopsy was also inconclusive, but lymphoma could not be excluded. Her condition continued to deteriorate and on 25 September she was admitted for elective tracheostomy with simultaneous microlaryngoscopy and biopsy. The biopsies again showed an intense lymphocytic infiltrate. The results of PCR are awaited.
Discussion: Acquired subglottic stenosis in the adult population always provides a management challenge. When the cause is obscure, it becomes even more difficult to treat. It is our contention that this patient has a localized form of lymphoma, probably of the MALT type. We hope PCR will provide a molecular diagnosis before deciding on further treatment options for this woman.

Case 6: A necrotic middle turbinate?
P. Chatrath, S. Duvvi, A. Dasgupta, C. R. Chowdhury (Harold Wood Hospital, Essex & Southend General Hospital, Southend)

A 67-year-old man presented to the ENT out-patient department at Harold Wood Hospital with a history of facial pains and headaches. He was known to suffer with myeloid dysplasia for which he was taking prednisolone 5 mg o.d. Endoscopic examination showed evidence of mild chronic rhinosinusitis only and he was treated accordingly. However, the patient returned to the department two days later complaining of increasing headache. Repeat rigid nasendoscopy revealed a prominent intensely black lesion that appeared to be entirely replacing the right middle turbinate. Differential diagnosis at that stage included a thromboembolic event secondary to his myeloid dysplasia, a haemangiomatous lesion, bleeding secondary to trauma or possibly a malignant melanoma. A CT scan was arranged which showed a soft tissue density in the region of the right middle turbinate but with no bony destruction. He was admitted for EUA of the nasal cavity and excision biopsy. Histopathology revealed degenerating and necrotic bony tissue containing non-caseating granulomatous inflammation with multinucleated giant cells, features suggestive of Wegener's granulomatosis. Subsequently, the ANCA was negative and renal function was found to be normal. Unfortunately, the patient is known to have died approximately one month ago, from ARDS and cardiorespiratory failure.
Discussion: Professor Michaels reviewed the histopathology. He reported that the features were highly suggestive of mucormycosis, an aggressive fungal infection with a poor prognosis leading to death usually within days. There was no evidence to suggest Wegener's granulomatosis as previously reported. Interestingly, the patient had the nasal biopsy over a year before he died, a period of time which is unheard of with this condition. He had in fact been treated aggressively by the medical team with steroids and high dose intravenous antifungal medication in view of his relative immunosuppression and haematological compromise secondary to his underlying myelodysplasia. These treatments must therefore have saved his life in the time period that followed the biopsy. The post-operative endoscopic photographs appeared to show a good wide excision of the necrotic mass with no residual disease. His eventual decline is not thought to have been related to mucormycosis although a formal post-mortem examination is awaited.

Case 7: Atypical presentation of squamous cell carcinoma of the pharynx
A. Qayyum, R. Pratap, A. Narula (St. Mary's Hospital, London)

A 43-year-old patient presented with a 36-hour history of infected right neck mass and intermittent dysphagia. Within 12 hours of admission he developed progressive cellulitis and skin necrosis. A diagnosis of necrotizing fasciitis of the neck was made. Neck debridement and tracheostomy was recommended, which he declined. Conservative treatment with high dose intravenous antibiotics was initiated. Empirical treatment began with benzylpenicillin, flucloxacillin and metronidazole and later changed to ceftriaxone, clindamycin and metronidazole once specificities were known. He improved clinically following cutaneous discharge of the abscess. Endoscopic examination of the laryngopharynx was performed which revealed an ulcer on the posterior pharyngeal wall from the level of the soft palate to the cricopharyngeus. Biopsies from this lesion showed a squamous cell carcinoma with basaloid differentiation. He was referred to the oncologists for further management as he had declined surgery. Only one case is mentioned in the literature[1] in which the patient developed necrotizing fasciitis following oesophageal cancer and his diagnosis was finally confirmed on post-mortem examination.
References
1 Francque SM, Van Laer C, Struyf N, Vermeulen P, Corthouts B, Jorens PG. Perforating oesophageal cancer presenting as necrotizing fascitis of the neck. *Eur J Gastroentrol Hepatol* 2001;**13**:1261–4

Case 8: Osteosarcoma of the lateral nasal wall: An unusual outcome
L. Kapoor, R. Natt, T. Tatla (Whipps Cross University Hospital, London)

A 78-year-old female presented with six-week history of recurrent epistaxis, nasal obstruction and unilateral discharge. Examination revealed a grey/white frond-like mass within the right nasal cavity. Twelve years previously she had had a chondroma excised from

the right collumellar septum. Computed tomography (CT) scan confirmed a mass occupying the right nostril, maxillary antrum, anterior ethmoids and nasopharynx, without bony erosion. Histopathology of biopsy under anaesthesia revealed malignant chondrosarcoma. Post-biopsy magnetic resonance imaging (MRI) scans suggested residual tissue in the right maxillary antrum and consensus opinion from multidisciplinary head and neck meeting, was to proceed to right lateral rhinotomy and medial maxillectomy. Subsequent histology showed absence of neoplasia in the excised tissue. Further specialist pathology opinions were sought from the Royal London Hospital and Harvard University, USA. Final biopsy report was high-grade pleomorphic spindle-cell sarcoma.

Discussion: Dr Rottenberg suggested that post-biopsy scans showed no residual tumour. Dr Sandison, highly experienced in sarcomas, clearly demonstrated malignant osteoid tissue in biopsy slides, with absence of cartilage components, consistent with an osteosarcoma. Its rarity at this age; especially in absence of previous bone disorders (i.e. Paget's disease) was highlighted. Sarcomas rarely present as nasal tumours, often with non-specific clinical features, requiring high degree of clinical suspicion. Initial biopsy, if doubtful, warrants consulting an experienced pathologist conversant with sarcomas, so that combination treatment may be optimized.

Case 9: Selective embolization of the right superior thyroid artery in the treatment of an adult laryngeal haemangioma

D. Bray, C. Giddings, S. Mady (St George's Hospital, London)

A 57-year-old woman presented with a one-year history of dysphagia, dysphonia and recurrent sore throats. At panendoscopy, a large thick walled mass was seen adherent to the right post-cricoid and arytenoid continuous with the right pyriform fossa. Biopsy caused significant bleeding. Histology showed abnormal thin-walled cavernous blood vessels consistent with haemangioma or arteriovenous malformation. A subsequent MRI scan was concordant with haemangioma. She was subsequently admitted for angiography and embolization of the right superior thyroid artery as an adjunct to subsequent formal excision. Post-embolization with polyvinyl alcohol, no other feeding vessels were noted, and the mass had decreased in size significantly. Post-embolization laryngoscopy revealed a significantly shrunken mass and no further management at the time was deemed necessary. At two-month follow up, the patient remained asymptomatic and flexible nasendoscopy revealed no further growth of the lesion. Unfortunately, at three-month follow up the lesion had recurred and the decision was made for the patient to undergo repeat embolization and excision of the laryngeal haemangioma.

Discussion: This case demonstrates a previously undescribed, safe, adjunctive treatment for adult laryngeal haemangioma.

Case 10: An interesting case involving skull base fungal infection

K. Amonoo-Kuofi, P. Tostevin, J. F. Knight (St George's Hospital, London)

A 19-year-old female with systemic lupus erythematosus (SLE) presented with a two-week history of left otalgia and hearing loss in association with a preceding upper respiratory tract infection. Current medication included prednisolone and azathioprine for control of SLE. On examination, there was mild inflammation of the left external auditory canal extending to the pars flaccida. The rest of her ENT examination including tuning fork tests was normal. A diagnosis suggestive of acute otitis media plus mild otitis externa was made and she was commenced on a course of oral and topical antibiotics (co-amoxiclav + gentisone HC). She presented a week later with worsening symptoms. On examination, her left tympanic membrane was hyperaemic and thickened with evidence of an effusion. Tuning fork tests were consistent with a left conductive hearing loss confirmed by a 40–50 dB hearing loss on pure tone audiometry with a type B tympanometry trace. She was admitted and treated with intravenous antibiotics, topical nasal decongestants and topical steroid nose drops. A CT scan showed opacification of her left mastoid air cell system and middle ear but no evidence of an intracranial collection. A left grommet was inserted under local anaesthetic. She continued to have aural discharge, which on repeated culture grew *Aspergillus fumigatus*. An MRI scan revealed fluid and an enhancing soft tissue involving the left mastoid air cells, in keeping with mastoiditis.

A wide cortical mastoidectomy with opening and clearing of the mastoid antrum was performed. Large amounts of granulation tissue were identified and sent for histological examination and cell culture. This confirmed the presence of invasive *Aspergillus* infection. She was successfully treated with systemic antifungals (voriconazole), initially intravenously then orally, for a total of six weeks. She was pain-free at follow up with no evidence of infection.

Discussion: What is already known on this topic: Invasive temporal bone mycoses are rare, predominantly associated with host immunodeficiency and usually difficult to diagnose. Cases usually have a fatal outcome. Treatment consists of antifungal chemotherapy, surgical debridement and attempts to control the underlying immunological condition. Professor Michaels held the opinion that the patient was most likely suffering from *Aspergillus* fungal infection as a result of relative immunosuppression following medical treatment for SLE.

Case 11: A unique bloody cough

P. M. Patel, R. J. Oakley, R. M. Sudderick (Royal Surrey County Hospital, Guildford)

We report a unique case of metastatic papillary thyroid carcinoma of the left piriform fossa. Commonly, metastases are localized in the lung, bone and less frequently the brain, liver and skin. A fifty-nine-year old gentleman presented with a six-month history of haemoptysis. Examination revealed a diffuse non-tender goitre, and flexible nasendoscopy found prominent vessels at the base of his tongue. An ultrasound scan showed a 2 cm single nodule in a dominant left multi-nodular lobe.

Direct pharyngolaryngoscopy revealed a small punctated lesion oozing fresh blood from the lateral wall of the left piriform fossa. S100 positivity on histological examination suggested a benign papillary sialadenoma arising from a minor salivary gland, however, epithelial cytomorphology along with diffuse vimentin positivity of glandular epithelium and cytoplasmic positivity for thyroglobulin suggested a metastatic deposit of papillary carcinoma of the thyroid. Subsequent STIR sequence MRI demonstrated a part cystic/necrotic mass in the left lobe of the thyroid gland extending posteriorly with a solitary right-sided neck lymph node. FNA cytology of the left lobe nodule yielded cells consistent with papillary carcinoma of the thyroid. At surgery for thyroidectomy and selective neck dissection, the mass within the thyroid gland was proved to be independent of a separate mass closely related to the posterior aspect of the thyroid and totally enclosed in a blind sac of pharyngeal mucosa. Subsequent management was completed with radioiodine and thyroxine suppression without complication. This unusual case highlights one of the myriad of ways in which metastatic papillary carcinoma of the thyroid can present.

Case 12: Hemi-laryngopharyngectomy for giant cell tumour of the larynx

Y. Gupta, J. Hamann, J. P. Jeannon, R. Ng (Guy's & St Thomas' Hospitals, London)

We present a case of a 51-year-old gentleman with a six-month history of hoarseness and discomfort on swallowing. Palpation revealed a hard, fixed mass in the left thyroid lobe. Laryngoscopic examination detected a sub-mucosal mass in the left ventricle and false fold. CT confirmed a mass in the left lobe of the thyroid gland, invading the larynx with thyroid cartilage destruction. The appearance of the FNAC was suggestive of papillary carcinoma of the thyroid. A total thyroidectomy, hemilaryngectomy, partial pharyngectomy and modified radical neck dissection was performed. Reconstruction was achieved using a rectus myo-cutaneous free-tissue transfer. Subsequent histological examination demonstrated giant cell tumour of the larynx. Twelve months on he is tolerating a normal diet and has excellent voice function and is disease-free.

Discussion: Giant cell tumours are benign tumours usually found in the ends of long bones. Presentation in the larynx is exceptionally rare, only 21 cases having been reported to date. Professor Michaels confirmed our diagnosis, attesting that histological diagnosis can often prove difficult. This case merits peer group discussion as it is an extremely rare, benign condition

that mimics a locally advanced thyroid malignancy. The complex reconstruction of such a large laryngeal and pharyngeal defect is also discussed.

Case 13: Unusual middle-ear tumours
N. Choudhury, P. Trojanowski, M. Gleeson (Guy's Hospital, London)

We presented two cases that were diagnosed with middle-ear adenoma. The first case was a 47-year-old male who presented with a two-year history of unilateral conductive hearing loss and pulsatile tinnitus. On examination, there was a red fullness behind an intact tympanic membrane, the appearance of which was suggestive of a glomus tumour. Angiography, however, did not demonstrate any abnormal circulation within the mass. The tumour was completely excised and histology confirmed a middle-ear adenoma. Our second case was a 52-year-old man who was referred with a four-year history of unilateral conductive hearing loss and otalgia. He also had a background history of a parathyoid adenoma that had been excised. On examination, there was a large exophytic mass occupying the external ear canal. A CT scan identified that the mass was originating from the middle ear. The mass was surgically excised via an endaural approach. We discussed the histology, which had been reported as a glandular tumour with neuroendocrine differentiation, consistent with a middle-ear adenoma.
Discussion: Professor Michaels, who originally described this histopathological entity in 1976, reviewed our histology. He held the opinion that variable degrees of neuroendocrine differentiation are a normal feature of all middle-ear adenomas. However, he said that our histology was not that of a middle-ear adenoma. He identified features of a malignant change within this tumour and suggested it may be metastatic from a possible medullary thyroid carcinoma. This will now undergo further immunohistochemical staining and histological analysis. Professor Michaels concluded that this case highlighted the diagnostic difficulties with this entity. He thought this was due to its rarity and unfamiliarity amongst clinicians and histopathologists, with this lesion.

Case 14: The management of residual endolymphatic sac tumour associated with von Hippel-Landau disease
R. J. D. Hewitt, R. Millard, M. J. Wareing (St Bartholomew's and The Royal London Hospitals, London)

A 19-year-old male, with von Hippel-Landau disease, presented with a recent history of left sensorineural hearing loss. Otoneurological examination was otherwise normal. MRI showed a large complex mass, consistent in this clinical scenario, with an endolymphatic sac tumour. The tumour was exposed and resected via a translabyrinthine approach. Histopathology confirmed the diagnosis. Post-operative MR scanning suggests a residual cyst adjacent to the jugular bulb and the panel was asked to comment on this and its management.
Discussion: Professor Michaels noted the tumour's resemblance to papillary carcinoma, its potential to invade bone and its vascular nature. Interestingly, he recalled a similar case in a lady with otitis media, with papillae and epithelial lining but not involving the endolymphatic sac. In that case the tumour was confined to the middle ear and the semantics of the tumour were debated. It was suggested that the tumour originated from the middle ear, and perhaps, should not be referred to as endolymphatic in origin. Dr Rottenberg proposed that fat-suppressing MR scanning (STIR) would be helpful in diagnosing and observing residual/ recurrent disease. Overall, the panel recommended serial MR scanning to monitor any residual disease and to screen for a contra-lateral lesion. No comment was made on the potential role of radio-surgery for these tumours.

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