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Pathologist: Professor L. Michaels, University College London
Radiologist: Dr J. Bingham, Consultant Radiologist, Guy's & St. Thomas' Hospitals
Minutes: Mr A. Bibas, Specialist Registrar in Otolaryngology, Guy's & St. Thomas' Hospitals.

Case 1: Thyroid mass with bilateral cord palsy

P. Sardana, A. Singh, M.E. Papesch (Whipps Cross University Hospital, London)

A 92-year-old woman presented with stridor for 48 hours and a history of a thyroid lump for three years. On examination there was a large irregular thyroid mass with bilateral abductor cord palsy. Fine needle aspiration cytology was suggestive of a papillary or follicular carcinoma. Computerized tomography (CT) scan demonstrated an extensive invasion lesion of the left thyroid lobe. Given these findings coupled with increasing respiratory effort, surgical intervention was discussed and agreed by the patient and her family. She underwent a total thyroidectomy, left selective neck dissection and tracheostomy and made an uneventful recovery. Initial histology was reported as oncocytic follicular adenoma. This was not consistent with the clinical findings. A review of histology detected vascular invasion in some of the prepared slides leading to a revised diagnosis of follicular carcinoma.

Discussion: Thyroid tumours have overlapping cytomorphological features making histological differentiation between benign and malignant lesions difficult. Specific combinations of immunohistochemical markers may help in differentiating between these tumours. Discussion highlighted the fact that this case represents a difficult group to manage. The essence must be on obtaining fully informed consent prior to any surgical intervention. Professor Michaels illustrated vascular invasion on one slide, but reiterated that histologically, thyroid tumours continue to provide a challenge.

Case 2: Diagnostic difficulty of a neck lump in a child

A. Amirfeyz, N. Patel, G.S. Kenyon (Whipps Cross University Hospital, London)

A three-year-old girl presented to A & E with three-day history of a left level II neck node, erythema of overlying skin and fever. She was admitted for i.v. antibiotics, but failed to settle. After four days the mass became fluctuant and incision and drainage was performed. There was no growth from the pus and initial serology tests were negative. An infected sinus with granulation tissue formation developed at the drainage site. She subsequently underwent excision of the granulation tissue with wedge biopsy of the neck node. Again all the results were negative with no evidence of granuloma or malignancy. Anti-TB chemotherapy was started empirically and she responded well to this; however the TB culture remained negative. Serology tests were repeated and showed a two-fold rise in anti-Bartonella Henslea IgG and the diagnosis of cat scratch disease was made with discontinuation of anti-TB therapy.

Discussion: Professor Michaels confirmed that the biopsy was consistent with a reactive lymph node. Mr Kaddour suggested that it may have been better to perform ultrasound-guided fine-needle aspiration (FNA) in the first instance, to avoid the risk of a discharging sinus after incision and drainage, whilst waiting for serology tests. It was agreed that heigthened parental anxiety can influence early surgical intervention.

Case 3: Nasal congestion in a traveller

A. Norris, J. Kanagalingham, H. Grant (Royal National Throat Nose & Ear Hospital, London)

A 38-year-old gentleman presented with an 18-month history of nasal congestion and crusting rhinorrhoea following travel in South America 21 months previously. Clinical examination revealed the presence of multiple painless crusting lesions over the nasal tip in association with hypertrophied, granular nasal mucosa. Investigations included routine serological tests, CT scan and biopsy of the nasal mucosa the histology of which indicated non-specific granulomatous changes only. A diagnosis of leishmanial infection was made by polymerase chain reaction (PCR) tests on the biopsy material. The patient subsequently responded to treatment with 40 days of intravenous stibogluconate.

Discussion: Mucocutaneous leishmaniasis is a sequel of infection with the protozoan *Leishmania* of the subgenus viannia, which is native to South America. Presentation may occur years after initial inoculation by the sandfly. If untreated, it can be extremely locally destructive resulting in gross nasal deformation and erosion. Professor Michaels confirmed that the clinical features of leishmanial infection and histopathological appearances of biopsy material can closely resemble those of other granulomatous diseases such as tuberculosis, Wegener's and sarcoidosis, thereby confounding diagnosis. Unless a history of foreign travel is obtained, the clinician may not be moved to arrange the often discriminatory PCR test.

Case 4: Cystic hygroma: surgical strategy

D. Jiang, S.P.A. Blaney, M. Gleeson (Guy's Hospital, London)

A 10-month-old boy presented with a swelling on the left side of his face since birth. No abnormalities had been noted on antenatal scan. Clinical examination showed a large soft fluctuate swelling over the left parotid and submandibular areas, that extended to the mastoid and the occipital area. An MRI scan showed a multicystic lesion involving the left parotid gland. It extended superiorly into the infratemporal fossa and posteriorly into the occipital area. A surgical management option was adapted as the tumour had increased significantly in size over a 10-month period. The tumour was removed by conservative total

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parotidectomy. All main branches of facial nerve were preserved, but the patient developed a post-operative severe facial nerve palsy, which significantly recovered one month after the operation. Histology confirmed the diagnosis of cystic hygroma.

Discussion: The complete surgical excision of cystic hygroma was the mainstay of treatment before 1990s. The reported 15–40 per cent spontaneous regression and development of safer sclerosing agents has persuaded some clinicians to adopt a 'wait and see' policy. We believe that, for fast growing and debilitating cystic hygromas, early surgical excision is the treatment of choice. Although complete excision may not be achieved we believe that preservation of function takes priority. Subsequent recurrence can be managed with other treatment modalities with less morbidity.

Case 5: A rare case of primary neuroendocrine small cell carcinoma of parotid gland

H.M.B. Khalil, A. D'Souza, M. Macnamara (Birmingham Heartlands Hospital, Birmingham)

A 75-year-old man presented with a painless swelling of the right parotid region, of five months duration, which was rapidly increasing in size. Examination of the neck revealed ipsilateral enlargement of level I and II lymph nodes. Facial nerve examination was normal. Fine needle aspiration cytology revealed the presence of malignant cells. CT scanning showed a solid mass occupying mainly the superficial lobe of the parotid gland as well as the presence of cervical lymphadenopathy. A right partial parotidectomy with facial nerve preservation was performed. Intra-operative frozen section of the adjacent lymph nodes showed malignant cells. Ipsilateral modified radical neck dissection was subsequently performed.

Discussion: The histopathological diagnosis revealed a small cell neuroendocrine carcinoma of the parotid gland with clear resection margins and regional metastasis in adjacent lymph nodes. A whole body CT scan was performed to exclude other possible primary sites. A radical course of radiotherapy was given to the parotid and ipsilateral neck. The patient sadly died of distant metastasis in the lung within two years of diagnosis Professor Michaels confirmed the histological diagnosis and described the rarity of neuroendocrine carcinomas in salivary glands.

Case 6: Disseminated *Fusobacterium* infection secondary to poor oral hygiene and chronic disability

R.J. Oakley, G. Tsiropoulos, G.S. Kanegaonkar

A 39-year-old gentleman with dystrophica myotonica and systemic lupus erythematosus presented with a left sided parotid swelling. Intra-oral examination revealed a grossly carious dentition and poor oral hygiene. Ultrasound examination suggested a neoplastic mass in the left parotid gland and a superficial parotidectomy was performed. Histological examination suggested chronic sialadenitis. The wound healed completely only to spontaneously dehisce later. Abdominal ultrasound and CT examination suggested a neoplastic mass in the upper pole of his right kidney. A right radical nephrectomy was performed and histological examination revealed a partly necrotic inflammatory lesion similar in parts to the parotid specimen. A CT chest revealed an abscess in the right lung apex an aspirate of which cultured *Fusobacterium nucleatum*. Nine months after his initial presentation the patient died from pneumonia due to disseminated fusobacterial infection.

Discussion: *Fusobacterium nucleatum* is one of the main constituents of dental plaque but can spread by the haematogenous route or aspiration to produce disseminated abscess formation. Professor Michaels reviewed the renal histology. Professors Gleeson and Cheeseman commented on the importance of treating dental neglect as part of the management of complex ENT cases.

Case 7: The Mystery of Lac Thien: aetiology and investigation of childhood sensorineural hearing loss

C. Hopkins. (Royal Sussex County Hospital, Brighton)

The case of a congenitally deaf Vietnamese chef was presented, along with his family tree, which included six affected siblings but no other family members. The aetiology and investigation of childhood hearing loss was discussed, both in regard to the case presentation and in the clinical setting.

Discussion: The prevalence of bilateral childhood deafness is estimated between 0.5–1.2/1000 live births. The importance of early identification and management has been demonstrated; those children diagnosed before six months of age have significantly higher vocabulary, language and comprehension scores than those identified at a later age. A recent publication from GOS suggests investigation of such children should include screening blood tests, ophthalmology review, ECG, renal U/S, CT of the temporal bones and genetic linkage studies where indicated. Recent developments were discussed; including OAE screening programmes, improved techniques in imaging, and advances in cochlear implantation. Congenital syphilis was proposed as a solution to the mystery of Lac Thien, but an autosomal recessive disorder or ototoxicity are possible alternatives. In view of the financial implications of thorough investigation, the low yield and an inability to treat most congenital causes it was felt that extensive searches for the underlying aetiology is impractical outside of specialist centres. However, the early diagnosis and aiding of children with congenital sensorineural hearing loss should remain a priority.

Case 8: An unusual cause of recurrent unilateral cervical lymphadenopathy

C. Ryan, C. Dutta, R. Simo, M. Khan (University Hospital Lewisham, London)

A 63-year-old lady presented in 1998 with right cervical lymphadenopathy. She had been previously managed for the same problem in 1984 by another hospital. The neck mass had been present for three months and was mildly uncomfortable but was not associated with any other symptoms. Examination revealed isolated firm lymphadenopathy in the upper right posterior and anterior triangles only. All blood screens and tests for infectious agents were negative. Fine needle aspiration cytology (FNAC), ultrasound (US) and CT were consistent with benign reactive hyperplasia. Following a period of observation without resolution a panendoscopy and open neck biopsy were performed. Histological examination required exclusion of lymphoma and a final diagnosis of reactive hyperplasia with progressive transformation of germinal centres (PTGC) was made. The patient was kept under close observation and again relapsed in 2001. Excision biopsy was repeated. Lymphoma could not initially be excluded, however, further histological staining confirmed recurrence of her previous diagnosis. She remains under close review.

Discussion: Progressively transformed germinal centres are thought to be morphological variants of follicular hyper-

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plasia and are of unknown cause. They are not felt to be pre-neoplastic lesions, however there is an ill-defined association with Hodgkin's lymphoma, in particular nodular lymphocyte predominance Hodgkin's disease (NLPHD). Clinically and histologically the diagnosis can be very difficult to differentiate from lymphoma, especially follicular lymphoma and NLPHD, requiring a high level of clinical suspicion and rigorous histological examination. Current management consists of observation with repeat excision biopsy in case of recurrent or progressive lymphadenopathy. The differences between PTGC and HIV-related lymphadenopathy were discussed.

Case 9: Post-traumatic laryngeal incompetence

A.S. Banerjee, A.M. Shaaban, D. Gibson, M.G. Dilkes (St Bartholomew's Hospital, London)

A 53-year-old man presented to the A & E department with self-inflicted knife wounds to the anterior and lateral neck and initial attempts at conservation of the laryngo-skeletal structures were performed. Later, it became evident that the patient had an insensate, hypofunctioning larynx. There was a fracture of the right-sided thyroid lamina, and the left vocal cord had been transected anteriorly. The damaged pharynx and trachea were repaired and a size 10 endotracheal tube was shortened and secured within the laryngeal lumen as a makeshift stent. Following recovery, video-fluoroscopic imaging suggested up to 95 per cent of food and saliva was being aspirated. Vocal quality was poor. Bilateral superior laryngeal palsies were also present contributing to the aspiration. Despite intensive speech therapy, the problem persisted. Cricopharyngeal myotomy, along with anterior hyoid suspension and augmentation of the vocal folds with Bioplastic® was performed. The rationale for this was that anterior hyoid suspensions would pitch the laryngeal introitus forward, helping the epiglottis to close over the larynx, and deflect food into the hypopharynx. The cricopharyngeal myotomy was performed to allow ingested food to drop freely through into the oeosphagus. The vocal folds were augmented to improve glottic closure thus strengthening the voice and further reducing aspiration. After surgery, significant improvement in aspiration occurred, such that one week later, the tracheostomy was removed. There was a good cough reflex present. A further video-swallow test showed a significant improvement in aspiration. After further speech and physiotherapy, the patient was fit enough to go home.

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Case 10: An aggressive thyroid lump

R.A.L. Di Cuffa, A. Singh, M.G. Dilkes (Whipps Cross Hospital, London)

A 62-year-old lady presented to A & E with a two-day history of hoarseness associated with a six-month history of a left thyroid lump, weight loss, lethargy and odynophagia. A CT scan found an invasive lesion of the left lobe of the thyroid with lymph node involvement and metastatic deposits in the liver. A bone scan was found to be normal. Excision of a local lymph node was found initially to show a pink cell variant of papillary carcinoma. However, on further evaluation, histology showed a Hurthle cell carcinoma. A total thyroidectomy with radioiodine ablation was subsequently performed. She was progressing well until a recurrence of a lump in the region of the thyroidectomy scar was found and she is now booked for external beam radiotherapy.

Discussion: Histology was reviewed by Professor Michaels who confirmed that it was a Hurthle cell carcinoma. The treatment of Hurthle cell neoplasia is controversial because of its rarity and the inconsistent criteria for diagnosis. Tumours which are greater than 2 cm should be treated with total thyroidectomy. Hurthle cell tumours do not respond very well to radioiodine ablation or external beam irradiation. Professor Cheeseman felt that despite the presence of liver metastases, a total thyroidectomy should be performed as this would reduce subsequent morbidity and provide an opportunity to determine whether the liver deposits would respond to radioactive iodine ablation therapy.

Case 11: Multisystem masses presenting a management challenge – NK cell lymphoma

R Singh Deol, A. Singh, M.G. Dilkes (Whipps Cross University Hospital, London)

A 39-year-old Afro-Caribbean gentleman presented to A & E with a three-week history of hoarseness, odynaphagia, general malaise, weight loss, subcutaneous lumps over his limbs and scrotal enlargement. Upon presentation he looked generally unwell, had stertor, a grossly enlarged painless scrotum and numerous lumps on his body, in particular a right forearm lesion causing ulnar nerve compression. He had no palpable lymphadenopathy. Nasendoscopy showed further lesions in the left nasal cavity, tonsil and hypopharynx. A biopsy was taken from one of the peripheral lesions and a NK cell lymphoma-nasal type was diagnosed. The patient was transferred to a specialist oncology centre. After a good initial response to an anthracycline-based chemotherapeutic regimen, progressive disease developed. Three further regimens were tried, each with short-lived benefit. The patient died seven months after presentation from overwhelming disease.

Discussion: This case illustrated non-typical presentation of nasal type NK cell lymphoma and the initial diagnostic difficulty encountered, particularly in a young man with gross testicular swelling. Some discussion centred on the possible use of localized radiotherapy in combination with the chemotherapy. It was felt that this might be of benefit in such cases, although the rapidly progressive and aggressive nature of this disease was appreciated by the group. This case clearly highlighted the need for multi-disciplinary team involvement in the management of such cases.

Case 12: To stent or not to stent? Endoluminal stent for bronchial malacia

L. Liew, S.P.A. Blaney (Guy's Hospital, London)

A male, with a cardiac anomaly diagnosed antenatally, was born by induction at 39 weeks. Soon after birth there were intermittent respiratory difficulties with recession and desaturations. Magnetic resonance angiogram and flexible bronchoscopy showed antero-posterior vascular compression of the distal trachea and proximal left main bronchus by the left pulmonary artery arising from the right pulmonary artery in the right lung forming a sling passing between the trachea and oesophagus. The sling was surgically corrected, but the respiratory difficulties continued. Further investigations, showed the malacic segment compressed between the aorta and left ventricle. A left

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main bronchopexy was performed, but the airway remained precipitous and therefore an endoluminal Palmaz stent was placed under direct visualization and X-ray guidance. This was successful, with an immediate airway improvement. Later the patient developed respiratory failure secondary to pneumonia and recovered, but unfortunately then suffered a cardiac arrest and died.

Discussion: A difficult airway compounded by complex cardiovascular anomalies provides a multidisciplinary team challenge. Endobronchial stents are very useful in maintaining airway patency secondary to segmental malacia, but they are not without its complications. Early complications include dislodgement and infection. Longer term complications include granulations that can cause bleeding and obstruction and erosion of the tracheo-bronchial tree and stenosis. Surgical correction of the primary cause should be the first option. The discussion group felt that even if the prognosis was poor, endoluminal stenting should be considered, as this allows the patient to be extubated.

Case 13: A challenging neck mass

R. Dasgupta, N. Patel, M. Papesch (Whipps Cross University Hospital, London)

A 65-year-old male smoker presented with a right-sided neck lump. He underwent excision biopsy and the histology was reported as 'ruptured epidermoid cyst'. He subsequently developed a discharging lump over the same site and was referred to us. Examination showed a 6 cm, firm, non-tender, fixed discharging right neck mass. FNA was non-diagnostic and CT scan showed a large multilobulated mass with a 'predominantly cystic component' and extension into the parapharyngeal space. Endoscopy revealed a lesion in the right nasopharynx. Biopsies from the nasopharynx and neck mass were reported as benign papilloma and inflammatory neck mass respectively. Due to clinical suspicion, he underwent further biopsies under GA, which were reported as 'sino-nasal carcinoma with neck metastases'. He has been offered radical radiotherapy.

Discussion: Professor Michaels commented that the histology showed a predominantly undifferentiated carcinoma with areas of basaloid differentiation consistent with primary nasopharyngeal carcinoma with neck metastases and added that 'sinonasal carcinoma' is not a histological entity. Professor Cheeseman advised that the best technique for biopsing the nasopharynx is to make a crescentic incision from left to right side and remove a whole strip of tissue to increase chances of a representative sample of tissue, as the nasopharynx is notorious for inadequate biopsies.

Case 14: Tullio's phenomenon due to superior semicircular canal dehiscence

D. Gibson, M. Wareing, A. Banerjee (St. Bartholomew's Hospital, London)

A 34-year-old man presented, initially with a three-year history of episodic balance loss and short-lived visual disturbances on hearing loud noises. Exercise was also noted to provoke symptoms. Of note, two years before the onset of these symptoms he had sustained a head injury requiring admission. He also gave a long history of episodic migraines. The patient felt that the hearing in his right ear was 'muffled', with the sensation of aural fullness. Neuro-otological examination revealed only that he was unsteady whilst performing Unterburger's test. An audiogram showed a mild, flat, right-sided sensori-neural hearing loss. Abnormalities from the vestibular function tests included borderline amplitude ratios for the electrocochleography (SP:AP ratio 29 per cent). Glycerol dehydration testing did not result in any change in hearing thresholds. The fistula test was negative. Calorics were unremarkable. In view of these inconclusive results a CT scan of the petrous temporal bones was requested. This suggested the presence of a dehiscent right semicircular canal. After discussing the possibility of a middle cranial fossa exploration with the patient a decision was made to manage the problem conservatively.

Discussion: The superior canal dehiscence syndrome has recently been described by Minor *et al.*[1] It has been demonstrated, with video-oculography and magnetic field coil recordings, that torsional eyes movements are precipitated in the plane parallel to the superior semicircular canal. Vestibular evoked myogenic potentials (VEMP's) are used to confirm the diagnosis in tertiary centres only. High-resolution computerized tomography is the investigation of choice in demonstrating superior semicircular canal dehiscence. Severe, disruptive symptoms, with this aetiology, will probably be best served with a middle cranial fossa exploration and plugging of the dehiscence. Over a short-term period the results of surgery are very encouraging.[2] Long-term outcomes are eagerly awaited. Professor Cheeseman was of the opinion that the symptoms would have to be fairly disruptive to warrant surgery with all its attributable risks.

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