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ONLINE ONLY ABSTRACT SELECTION

Proceedings of the 152nd Semon Club, 14 November 2016, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Chairpersons: Miss Elfy B Chevretton, and Miss Irumee 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 Catherine Chatfield-Ball for 'When the diagnosis of a tracheal tear has already been made'.

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 and anterior skull base section

Chairperson: Mr Shahzada Ahmed

Unilateral sudden-onset epistaxis

A O Alade, P Singh, A Levene

From the Luton and Dunstable University Hospital

Introduction

Primary sinonasal renal cell-like adenocarcinoma is a very rare tumour that bears histological resemblance to the clear cell type of renal cell carcinoma. We report a case presenting with unilateral epistaxis.

Case report

A 55-year-old male presented with brisk right-sided epistaxis, which failed to resolve with conservative management. He was subsequently scheduled for sphenopalatine artery ligation, which was unsuccessful because of excessive bleeding and deformed anatomy. A red polypoidal lesion was seen endoscopically emerging from the right ethmoid cavity.

Radiological findings

Computed tomography angiography identified a $5 \times 3 \times 3$ cm soft tissue mass centred on the ethmoidal air cells. There was significant localised bone destruction, which involved the nasal septum, ethmoid bone, both of the lamina papyracea, medial walls of the maxillary sinuses and anterior walls of the sphenoid sinuses. In addition, there was localised bone destruction of the cribriform plate.

Dr Connor demonstrated how cellular lesions such as sinonasal tumours exhibit a low T2-weighted signal on magnetic resonance imaging scans. He highlighted that the periosteum was intact, indicating no invasion into the brain (Supplementary Figure 1). He also demonstrated the blood vessels on magnetic resonance angiography, indicative of a vascular tumour.

Histological findings

The tumour was composed of sheets, islands and nests of epithelial cells with clear cytoplasm in the vascular stroma. There were focal duct formations. No necrosis was seen and mitoses were few. Periodic acid–Schiff and periodic acid–Schiff diastase stains confirmed abundant glycogen in the cytoplasm (Supplementary Figure 2).

Dr Sandison explained how sinonasal renal cell-like adenocarcinoma had only recently been included in the World Health Organization classification, and that it is rare to find clear cell tumours in the nose. She also said that the expression of cytokeratin 7 and S100 protein indicate that this tumour was of nasal origin as opposed to renal origin.

Management

The patient underwent successful right extended lateral rhinotomy and craniofacial resection.

Conclusion

Sinonasal renal cell-like adenocarcinoma and renal cell carcinoma can be distinguished successfully via immunohistochemistry. The tumour is characteristically highly vascular and therefore pre-operative embolisation is a viable option to reduce intra-operative bleeding.

A serious incidentaloma in a patient presenting with sudden hearing loss

A Kaura, C Rennie, V J Lund

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

Introduction

Olfactory neuroblastoma is a rare sinonasal tumour. It may give rise to paraneoplastic syndromes, of which the syndrome of inappropriate antidiuretic hormone secretion is the commonest.

Case report

A 38-year-old woman presented with left-sided sudden sensorineural hearing loss (SNHL). She had been diagnosed with syndrome of inappropriate antidiuretic hormone secretion 12 years earlier. The sudden SNHL was treated with oral and intratympanic steroids, but showed no improvement.

Radiological findings

Magnetic resonance imaging (MRI) demonstrated an irregular soft tissue mass in the left nasal cavity, expanding the left middle meatus (Supplementary Figure 3). The appearances were suggestive of an inverted papilloma, but other sinonasal tumours could not be excluded.

Histological findings

Biopsy demonstrated olfactory neuroblastoma (Hyams' grade 1) (Supplementary Figure 4). Surgical excision confirmed this, showing focal deposits in the anterior lamina papyracea, nasolacrimal bone and posterior ethmoid, with clear margins; the tumour was positive for synaptophysin and S100 protein.

Dr Sandison demonstrated the round cell tumour under high power magnification. She commented that the current grading system for olfactory neuroblastoma is based on morphology and is controversial. She also suggested that an ectopic pituitary tumour could not be excluded at this stage and advised that the patient have pituitary markers checked.

Management

The patient underwent endoscopic medial maxillectomy and resection of the tumour seven weeks postpartum, and completed a course of chemoradiotherapy. Her plasma sodium and osmolality levels normalised post-operatively.

Discussion and lessons learned

Mr Ahmed commented that it appears aggressive for the multidisciplinary team to recommend chemoradiotherapy in a low-grade lesion that was completely excised. Professor Lund would advocate adjuvant radiotherapy for all olfactory neuroblastomas in view of published evidence of improved survival and reduced recurrence rates. Dr Sandison explained that if vascular or perineural invasion were seen on histology, chemotherapy would be recommended.

Conclusion

In cases of unexplained syndrome of inappropriate antidiuretic hormone secretion, rare causes including sinonasal tumours need to be excluded. Patients should undergo computed tomography of the head, and/or MRI scanning of regions including the paranasal sinuses and nasal cavity.

Unilateral nasal discharge and mild nasal obstruction

T Williams, N Stobbs, M Nussbaumer

From the Barnsley Hospital NHS Foundation Trust

Introduction

Chondrosarcomas are rare malignancies, requiring imaging and biopsy for histology to confirm diagnosis and establish tumour extent. If detected early enough, wide excision of the tumour with possible adjuvant treatments can improve five-year survival rates.

Case report

A 78-year-old female presented with a 1-year history of unilateral nasal discharge and obstruction. Nasendoscopy revealed abnormal bony swellings in the floor of the nose. Examination under anaesthetic was performed and biopsies were conducted.

Radiological findings

In light of patient co-morbidities, computed tomography was undertaken (Supplementary Figure 5) instead of magnetic resonance imaging.

Dr Connor demonstrated a lesion arising from the inferior nasal septum, with invasion of the underlying maxilla and hard palate, and encroachment on the medial wall of the right maxillary sinus. The lesion was solid with patchy calcification. There was no evidence of lymphadenopathy or metastasis.

Histological findings

Dr Sandison demonstrated respiratory mucosa, with cartilage and atypical increased cellularity, with myxoid areas (Supplementary Figure 6). She explained that this indicated a low-grade lesion with cartilaginous elements, suggestive of low-grade chondrosarcoma. The specimen was sent for review at the specialist sarcoma multidisciplinary team (MDT) in Birmingham, who confirmed the diagnosis.

Management

The MDT outcome recommended extensive resection with curative intent. The patient has been referred to the maxillofacial team to discuss surgical resection options, and to the restorative dentistry team for pre-emptive moulds because of the likely collapse of the hard palate.

Discussion

Mr Ahmed noted that these tumours are very slow growing and so patients tend to have good five-year survival rates. He said that when operating around the clivus, there are limits to surgical resection. He enquired as to whether radiotherapy was a treatment option. Dr Sandison advised against it given the risk of sarcomatous transformation. She also explained that chemotherapy is also not advised because chondrosarcomas have poor vascularity.

Progressive loss of vision and eye movement in an 83-year-old woman

D V Sinclair, G Lloyd, C Hopkins

From the Guy's Hospital, London

Case report

An 83-year-old woman with a history of malignant melanoma of the right cheek presented with a 2-year history of gradual onset epiphora, loss of vision, and loss of movement and pain in the right eye. On examination, the globe was non-functional, necrotic and fixed. Autoimmune screening was negative.

Radiological findings

Dr Connor demonstrated a very destructive lesion on magnetic resonance imaging. The lesion extended into the sphenoid and ethmoid sinuses, and involved the extraocular muscles and medial orbital soft tissue, with dystrophic changes of the globe.

Pathology

Biopsies of the ethmoid bone and orbital apex showed scanty immunoglobulin G4 (IgG4)-positive plasma cells, with no evidence of neoplasia (Supplementary Figure 7). Serum IgG4 levels were subsequently measured, which were double the upper limit of normal. Oral prednisolone brought only modest radiological disease resolution. The patient elected to undergo orbital exenteration, with a free-flap reconstruction, because of the severe pain.

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Dr Sandison explained how a ratio of immunoglobulin G:IgG4 greater than 30–40 per cent was required to make a diagnosis of IgG4-related disease and advised a multidisciplinary team approach to diagnosing these lesions. Mr Ahmed commented that organ-specific criteria for the orbit require only 10 cells per high powered field, making diagnosis in this case one of the clearest he has seen. Dr Sandison was in agreement. She also warned that these lesions can present as pseudotumours and can easily be misdiagnosed as fibrous lesions.

Discussion

Mr Ahmed related that, in his experience, patients had usually received high-dose steroids prior to biopsies being taken, and this frequently resulted in difficulties reaching a diagnosis.

Conclusion

Immunoglobulin G4 related disease is a rare immune-mediated condition affecting multiple organ systems, including the orbit, pancreas, salivary glands and retroperitoneum. Diagnosis is based on histological and immunohistochemical findings. ENT surgeons should consider this diagnosis as a cause of orbital disease.

Chronic unilateral rhinorrhoea and otorrhoea

A O Alade, A Vijendren, N Clifton
From the Luton and Dunstable University Hospital

Introduction

In cases of a unilateral nasal mass, care must be taken to exclude neoplastic or intracranial pathology.

Case report

A 60-year-old female presented with a 1-year history of clear right-sided rhinorrhoea following functional endoscopic sinus surgery (FESS) the previous year. The FESS had been performed by another ENT team after the patient had presented with chronic right-sided facial pain, rhinorrhoea and a discharging right ear (which was her only hearing ear).

Radiological findings

Dr Connor demonstrated on the computed tomography (CT) scan a dehiscence in the right lateral wall of the sphenoid sinus, lucency of the sphenoid wing and arachnoid granulations throughout the skull base (Supplementary Figure 8).

A subsequent magnetic resonance imaging scan identified a soft tissue mass and fluid filling the sphenoid sinus, continuous with the extracerebral cerebrospinal fluid (CSF) in the right middle cranial fossa, consistent with a sphenoid meningoencephalocele (Supplementary Figure 9). A separate 5.5 × 5 5× 54.4 cm left parafalcine meningioma was also identified. A CT scan of the temporal bones identified a 2 mm defect in the right tegmen tympani, with opacification of the mastoid air cells.

Histological findings

The tissue removed from the right sphenoethmoidal recess during endoscopic biopsy was identified as an inflammatory nasal polyp.

Management

The sphenoid meningoencephalocele was resected endoscopically, with repair of the skull base defect jointly with a neurosurgical team. The meningioma was excised as a separate procedure. Given the high risk of meningitis, the patient also underwent repair of the tegmen defect and closure of the tympanic membrane perforation.

Discussion

The meningioma was an incidental finding, whereas the meningoencephalocele and CSF leak were suggestive of chronic idiopathic intracranial hypertension. Mr Ahmed questioned the necessity of removing the asymptomatic meningioma. He also observed that idiopathic intracranial hypertension is associated with the female gender and elevated body mass index (BMI). This patient did not have an elevated BMI. Mr Ahmed also reminded the audience that the skull base defects should only be closed when there are specific indications.

Otology and skull base section

Chairperson: Miss Irumee Pai

An unusual fibro-osseous temporal bone mass

J Faulkner, M Whittaker, R Obholzer
From the Guy's Hospital, London

Case report

A 65-year-old man was referred with an incidental temporal bone mass following a computed tomography (CT) scan of the head. The patient had been suffering with tinnitus and migraines, with some distortion of balance. Examination findings were unremarkable.

Radiological findings

The CT imaging showed a destructive mass in the left temporal bone that had features suggestive of malignancy, with erosion of the mastoid air cells and erosion into the posterior cranial fossa. Positron emission tomography showed increased activity limited to the left temporal bone, with cortical erosion most prominent in the left mastoid.

Histological findings

Dr Sandison demonstrated bone trabeculae within fibrous tissue, and irregular islands of immature bone. She felt this was most consistent with a diagnosis of fibrous dysplasia, but said that the differentials included a brown cell tumour, a reparative granuloma or a reactive condition. She advised that such specimens should ideally be sent fresh in formalin to the Royal National Orthopaedic Hospital in Stanmore for confirmation of the diagnosis.

Management

Following excision of the lesion, the patient's hearing has been unaffected and he has recovered well. He will undergo a post-operative CT, with annual review.

Discussion

Miss Pai enquired if a magnetic resonance imaging (MRI) scan may have been helpful in the management of this patient, with a view to avoiding surgery in a relatively asymptomatic patient. Dr Connor advised that fibrous dysplasia can look invasive and hence worrying on an MRI scan, and felt that the MRI scan would probably not have changed the management of this patient. In any event, the patient was unsuitable for MRI scanning given that he has a cardiac pacemaker.

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Time heals – balance disturbance and severe unilateral hearing loss progressing with acute symptom and radiological changes

E Crossley, D Argiris, C Hardwidge

From the Royal Sussex County Hospital, Brighton and Sussex University Hospitals NHS Trust

Introduction

Acoustic neuromas are the most common cerebellopontine angle tumours. A spontaneous macroscopic haemorrhage into this tumour is very rare, particularly in tumours smaller than 25 mm.

Case report

An otherwise healthy 40-year-old man was referred to the ENT clinic with a 6-month history of balance disturbance and distorted left-sided hearing. Symptoms including facial palsy, poor spatial awareness and pain had progressed over a nine-month period. As a result of the patient's commitments, the proposed surgery was delayed by a further five months, during which time the symptoms improved.

Radiological findings

The initial magnetic resonance imaging (MRI) revealed a lesion, 11 × 17 mm in size, consistent with an intracanalicular acoustic neuroma (Supplementary Figure 10). Three subsequent MRI scans (at 2, 8 and 11 months after presentation) demonstrated three separate episodes of haemorrhage, initially causing significant tumour enlargement, essentially liquefying its solid component. The MRI scan performed at 14 months demonstrated a reduction in size of the acoustic neuroma (Supplementary Figure 11).

Management

The patient was managed conservatively with high-dose oral prednisolone and hearing aids.

Discussion and lessons learned

Miss Pai noted that it is unusual for a relatively small lesion of this nature to cause such extensive symptoms. Dr Connor explained how slow-flowing venous blood can produce similar high T1-weighted signals; he suggested a venous angioma as a possible differential diagnosis. He also noted that microhaemorrhages are seen in 90 per cent of acoustic neuromas.

Conclusion

This case demonstrates the importance of pre-operative scans being undertaken close to the operation date, especially when there are changing symptoms or findings on imaging. In addition, even in the context of an intralesional haemorrhage causing enlargement, delaying surgery could be considered.

Sudden-onset hearing loss: lightning can strike twice

S Khosla, M Whittaker, C Skilbeck

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

Introduction

Sudden-onset hearing loss can be a life-changing condition, particularly if it occurs bilaterally. Here, we present an extremely unusual case of sequential sudden-onset hearing loss due to pneumolabyrinth secondary to minor pressure changes.

Case report

A 56-year-old male presented with left, sudden-onset hearing loss, unsteady gait and horizontal nystagmus following an international flight, with an upper respiratory tract infection on a background of severe chronic sinusitis. Two months previously, a similar event had caused severe sensorineural hearing loss (SNHL) in the contralateral ear.

Radiological findings

The magnetic resonance imaging scan showed no infarction or internal auditory meatus lesion. However, there was poor definition of the left lateral and posterior semicircular canals and cochlea, but sparing of the VIIth and XIIIth cranial nerves. Computed tomography of the mastoid showed gas replacing the cochlear and labyrinthine inner-ear fluid, suggestive of a round or oval window fistula (Supplementary Figure 12a).

Management

Pure tone audiometry showed profound SNHL (Supplementary Figure 13). The findings of haematological and biochemical investigations were normal. A high-dose steroid trial had no impact. Subsequently, the patient underwent left exploratory tympanotomy, fascial plugging of the round window niche and steroid instillation. After significant input from vestibular physiotherapy and speech therapy, psychological input, and counselling, he has compensated well for his vestibular dysfunction, with mild improvement in hearing on pure tone audiometry.

Discussion

Miss Pai noted the patient's marginal hearing improvement; however, she recommended consideration of early cochlear implantation given the risk of subsequent cochlear ossification.

Conclusion

Exploratory tympanotomy and round window niche plugging aided resolution of this unusual barotraumatic pneumolabyrinth, with slight improvement in hearing. However, the patient remains within the threshold for cochlear implantation.

Paediatric section

Chairperson: Mr Christopher Pepper

'My son has a tumour on the right side of his face'

G Oikonomou, C Tornari, H Daya

From the St George's Hospital, London

Introduction

Facial lumps in children are commonly treated as an infectious disease by general practitioners, but the differential diagnosis includes a broad spectrum of diseases.

Case report

We report the diagnosis and management of a four-year-old child who presented with a six-week history of a rapidly enlarging right parotid area mass. Symptoms included pain, dysarthria, dysphagia and weight loss that prompted referral.

Radiological findings

Dr Siddiqui demonstrated a large right infratemporal tumour, which was pushing the ramus of the mandible and parotid

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gland laterally. There was intracranial extension without brain parenchymal involvement. There was some mass effect on the upper airway (Supplementary Figure 14). Positron emission tomography suggested metastatic disease.

Histological findings

Pathology demonstrated stage 3 (clinically staged as a T_{2b} tumour, and pathologically staged as N_0 and M_0 in terms of nodal classification and metastasis), group III translocation-negative alveolar rhabdomyosarcoma of the right infratemporal fossa (Supplementary Figure 15).

Management

Following initial imaging and biopsy, the patient underwent chemotherapy and proton beam therapy.

Discussion

Mr Pepper enquired whether fine needle aspiration cytology would provide an adequate histological specimen. Dr Sandison advised that the loss of architecture would be unhelpful, particularly in low-grade tumours, and that the volume of tissue would be inadequate for immunochemistry. She recommended a core biopsy, but warned that whenever taking a biopsy of any sarcoma, there is a risk of metastatic seeding of the tumour along the path of the biopsy. She therefore advised carefully planning the surgical approach prior to taking the biopsy.

Conclusion

Any progressively enlarging, symptomatic facial mass should always raise suspicion for soft tissue malignancy.

Chronically discharging ear with a cochlear implant in situ

J Faulkner, H Powell, I Pai

From the Guy's Hospital, London

Introduction

Cochlear implantation may rarely be complicated by a devastating infection many years after surgery, and patients and/or parents should be counselled of the risk appropriately.

Case report

A 12-year-old girl presented with a 2-year history of chronic discharge from the right ear and cochlear implant non-use. She had received a cochlear implant in the right ear at the age of four years for profound deafness and had become a non-user aged eight years. She had presented to primary care several times and received multiple courses of antibiotics over the two years. Otoscopy revealed a right ear full of infected keratinous material and aural polyps. Integrity testing showed the implant to still be functioning.

Radiological findings

Computed tomography showed significant bony destruction and extension of the electrode beyond the cochlea, consistent with aggressive tympanogenic labyrinthitis.

Management

The patient underwent explantation of the cochlear implant. Intra-operatively, the middle ear was filled with granulation tissue, with destruction of the ossicles. The implant was removed and granulation tissue cleared. An attempt was made to insert a 'dummy' electrode to keep the cochlear lumen patent for potential re-implantation; this was not possible and the parents were counselled regarding other modes of communication.

Discussion

This destructive process had occurred over many years. Patients with a cochlear implant receiving multiple courses of antibiotics should be investigated for deeper-seated infections. Miss Pai suggested that all patients undergoing cochlear implantation should be counselled on chronic infections as a long-term complication.

When the diagnosis of a tracheal tear has already been made

C Chatfield-Ball, N Amin, V Possamai

From the Evelina London Children's Hospital

Introduction

Tracheal tears are a rare but life-threatening cause of respiratory distress, usually attributable to trauma. Patients may present with subcutaneous emphysema, haemoptysis or respiratory compromise.

Case report

We present the case of a 15-year-old girl who developed severe neck and thoracic subcutaneous emphysema following an asthma-like coughing fit. Flexible bronchoscopy revealed an extensive posterior tracheal tear, presumed to be secondary to the violent coughing. This was managed conservatively with prolonged intubation for 10 days, during which the tear healed. Chest X-rays demonstrated minor hyperinflation with a normal mediastinum, supporting the theory of undiagnosed asthma. She was successfully extubated and discharged with asthma medications.

At a two-month respiratory out-patient review, the patient reported experiencing cough and respiratory distress once more. This prompted further imaging.

Radiological findings

Dr Connor demonstrated a posterior mediastinal soft tissue mass on the computed tomography scan. Loss of fat planes is indicative of an aggressive invasive process. The magnetic resonance imaging scan showed involvement of the posterior tracheal and oesophageal walls. Further microlaryngoscopy and bronchoscopy demonstrated an obstructive distal trachea lesion, with significant posterior tracheal compression from the mass (Supplementary Figure 16).

Histological findings

Tracheal biopsy indicated an anaplastic large cell lymphoma.

Management

Chemoradiotherapy was commenced, with a good initial response.

Discussion

Although bronchoscopy remains the 'gold standard' for diagnosis of tracheal tears, we should remember that early cross-sectional imaging can also be of value diagnostically. In the past, early surgical intervention has been seen as the cornerstone of management in tracheal injury, but there is a growing body of evidence to suggest that conservative management has better outcomes.

Conclusion

The diagnosis of a tracheal tumour should be considered in any paediatric patient with refractory asthma-like symptoms.

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Sustaining a tracheal injury, without history of iatrogenic damage or external trauma, should make physicians wary of alternate pathologies given the rarity of a spontaneous tear.

Problematic recurrent neck abscess

E Molena, N Hughes, L Pitkin

From the Frimley Park NHS Foundation Trust

Introduction

We present the case of an eight-year-old girl with a recurring left anterior neck abscess.

Case report

An eight-year-old girl, previously fit and well, presented in 2015 with an anterior left neck abscess that was incised and drained. Microbiology grew *Streptococcus intermedius*. She re-presented in February 2016 with identical symptoms, which settled with intravenous antibiotics. A further episode occurred in August 2016 and was treated with a course of oral antibiotics.

Radiological findings

Initial ultrasound showed an ill-defined mass anteriorly in the left side of the neck. Magnetic resonance imaging did not show any evidence of a branchial cleft cyst. A further ultrasound scan showed an oval structure in the sternal notch, between the brachiocephalic veins, thought to be a cyst. Barium swallow study demonstrated a trickle of contrast from the left hypopharynx.

Histological findings

The cystic structure found at surgery was confirmed to be the thymus gland on histology.

Management

The patient underwent exploration of the neck and upper mediastinum. Panendoscopy findings were normal. A large cystic structure was found in the left root of the neck surrounded by scarring (Supplementary Figure 17). This extended down behind the brachiocephalic vein.

Lessons learned

Pre-operatively, this had been suspected to be a third branchial cleft cyst. Discussions from the Semon Club meeting suggested that it was likely this had been a recurrent thymus abscess. Dr Sandison advised that the specimen be cut in different planes, as the lack of a cavity may have been a sampling error.

Conclusion

Although rare thymus abscesses can occur, recurring neck abscesses should be investigated for an underlying cause.

Head and neck section

Chairperson: Miss Lisa Pitkin

New-onset, enlarging, left-sided neck swelling

K Vijayasurej, N Foden, R Terry

From the Princess Royal University Hospital, Orpington

Introduction

Cervical sympathetic chain schwannomas are exceptionally rare. Most extracranial schwannomas of the head and neck are found in the parapharyngeal space, with the vagus nerve accounting for 3 per cent of these. First described as 'neurinoma' by Verocay in 1908, this encapsulated nerve sheath tumour is considered benign and slow growing, with few cases of nerve invasion and pre-operative Horner's syndrome reported. Surgical excision frequently results in poor preservation of nerve function.

Case report

A 55-year-old male presented with a 4-week history of a painless, left-sided neck swelling increasing in size. Cranial nerve function remained intact.

Radiological findings

An ultrasound scan revealed a large left, level II/III lesion, measuring $6 \times 4 \times 3$ cm. Fine needle aspiration was performed. Magnetic resonance imaging (MRI) with gadolinium identified a well-defined, oval, soft tissue mass in the left side of the neck, displacing the carotid vessels anteriorly, and lying within the carotid space from the level of C3 to C6 (Supplementary Figure 18). No lymphadenopathy was noted.

Histological findings

Subsequent histology depicted a well-circumscribed spindle cell tumour composed of wavy spindle cells, with interlacing fascicles and classic Verocay body formations, with Antoni type A and B patterns.

Management

Video fluoroscopy revealed normal swallowing. Management involved excision of the lesion (Supplementary Figure 19). Post-operative partial Horner's syndrome (miosis only) was apparent.

Discussion

Interestingly, the MRI displayed separation of the common carotid artery and internal jugular vein, with displacement anterolaterally (normally consistent with a vagal schwannoma). Dr Sandison explained that the diagnosis was dependant on finding both Antoni A and B areas, and advised biopsy under radiological guidance to increase the chances of a representative sample.

Conclusion

Pre-operative imaging is crucial in distinguishing vagal from cervical sympathetic chain schwannoma. Careful attention of vascular anatomy and associated displacement can guide operative management.

A rare cause of progressive hoarseness

R Thomas, C Xie, D Kim

From the St George's Hospital, London

Case report

A 58-year-old woman presented with a 6-month history of dysphonia. Apart from age, there were no other risk factors. She was found to have right vocal fold paralysis. Computed tomography (CT) was undertaken. Fine needle aspiration cytology suggested parathyroid carcinoma (Thy4). She underwent right hemithyroidectomy with excision of the mass, and right lateral neck dissection of levels II–IV. Intra-operatively, there was tracheal invasion, which was repaired with a sternocleidomastoid flap. Three days post-operatively, she suffered sudden subcutaneous emphysema with bilateral pneumothoraces, requiring a tracheostomy and chest drains. Following recovery, the patient was decannulated.

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Radiological findings

The CT scan showed a 6 mm calcified lesion in the right thyroid lobe, and excess soft tissue mass of different enhancement lying in contact with the right side of the trachea (Supplementary Figure 20). There was no radiological lymphadenopathy.

Histological findings

Histology showed a 30 mm focus of classical papillary thyroid carcinoma, a diffuse sclerosing variant, and another area of transformation to squamous cell carcinoma (SCC) (Supplementary Figure 21), with extensive perineural and adjacent soft tissue (including parathyroid) invasion. This was pathologically staged $T_{4a}N_{1a}M_0$, with positive margins.

Management

As completion thyroidectomy would be delayed because of her tracheostome, the multidisciplinary team opted for radiotherapy, followed by completion thyroidectomy, plus or minus radioactive iodine.

Discussion

Primary SCC of the thyroid represents less than 1 per cent of thyroid carcinomas. Dr Sandison explained that primary SCC of the thyroid is most commonly described with diffuse sclerosing variant papillary carcinoma, which is biologically aggressive. It usually presents with rapid and diffuse thyroid enlargement, and is more common in women. Miss Pitkin mentioned it is generally radiotherapy-resistant. Dr Sandison explained that the radioactive iodine was suggested because of the focus of papillary carcinoma.

A rare cause of subcutaneous thickening in the head and neck

K Karamali, D Lother, N Choudhury

From the East Surrey and Sussex Hospital

Case report

A 40-year-old gentleman presented with a 6-month history of persistent swelling in the right parotid region, with associated discolouration of the overlying skin and regional discomfort. There was no history of fluctuations in size whilst eating, and no associated systemic illness or weight loss. However, he later reported night sweats but no fever. Clinical examination revealed thickening and induration of the skin in the right parotid region and level II lymphadenopathy.

Radiological findings

Ultrasound and computed tomography scans of the neck (Supplementary Figure 22) both confirmed diffuse infiltration and thickening of the subcutaneous tissue.

Histological findings

A histological analysis of the subcutaneous tissue in the right parotid area identified small round histocytes, mature plasma cells and atypical lymphoid cells. A number of lymphocytes were associated with rimming and atypia. They were positive for cluster of differentiations 3, 4, 8 and 43, and negative for cluster of differentiations 30 and 56, although many lymphocytes seemed not to express beta F1. The overall histological findings suggested a subcutaneous panniculitis T-cell lymphoma overlapping with lupus profundus, which is a well-recognised phenomenon.

Discussion

Dr Sandison recommended analysis of cell markers to confirm a monoclonal origin.

Management

The patient has been referred to haematology for further treatment.

Conclusion

Meticulous examination is essential in the case of panniculitis as the diagnosis of subcutaneous panniculitis T-cell lymphoma can be easily missed.

A posterior oropharyngeal mass: a difficult diagnosis

G Thompson, B Yu, O Mirza, V Pothula

From the Salford Royal NHS Foundation Trust

Background

Myoepitheliomas make up 1–1.5 per cent of all salivary gland tumours. We present a case that illustrates the ongoing difficulty in diagnosing such tumours.

Case report

A 56-year-old man presented to the ENT out-patient department with a swelling of the posterior oropharyngeal wall causing snoring. Examination revealed a 2 cm non-ulcerated, pedunculated mass.

Radiological findings

A computed tomography scan showed a 17 mm, well-defined polypoidal mass arising from the posterior oropharynx (Supplementary Figure 23). There were no signs of invasion.

Histological findings

Dr Sandison demonstrated a spindle and epithelioid cell lesion that was not obviously malignant. There was no necrosis, and the lesion was monomorphic, with no mitosis.

The initial histology report suggested an aggressive paraganglioma in light of a Zellballen pattern of cells. Immunohistochemistry demonstrated patchy staining for vimentin, suggesting a diagnosis of myoepithelioma.

Management

The lesion was excised, and initially histology reported clear resection margins. With an initial diagnosis of aggressive paraganglioma and evidence of tumour recurrence at four weeks' follow up, the patient was referred to the regional head and neck multidisciplinary team. In light of the second immunohistochemical opinion favouring myoepithelioma, it was felt that the tumour had recurred but did not have malignant potential. The patient underwent a repeat excision with radiological follow up.

Discussion

Miss Pitkin advised that lesions of the posterior pharynx can usually be resected with a wide margin, as this area usually re-epithelialises and heals well. Dr Sandison wondered if the reporting pathologist had inked the resection margin to prevent autolysis, leading to a misleading impression that the resection margins were clear. Heat artefacts during excision can also give misleading margins. Miss Chevretton advised nasendoscopy in all patients who present with snoring.

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Learning outcomes

To the best of our knowledge, this is the first reported case of myoepithelioma arising from the posterior oropharynx.

An unusual cause of progressive dyspnoea and stridor

D W Scholfield, J S Virk, P Stimpson

From the Whipps Cross University Hospital, London

Introduction

Neuroendocrine tumours account for approximately 1 per cent of laryngeal neoplasms. Our case demonstrates the challenges in achieving an early diagnosis and subsequent resection.

Case report

A 43-year-old gentleman was referred to the respiratory clinic with a 15-month history of cough and dyspnoea on exertion. He had been treated for asthma by his general practitioner. On examination, he was found to have soft expiratory stridor. Flow volume curves showed narrowed expiratory and inspiratory loops. Subsequent computed tomography (CT) of the neck and chest led to otolaryngology referral. A large pedunculated subglottic mass was visible on endoscopy.

Radiological findings

The CT scans demonstrated an exophytic polypoidal subglottic lesion with a maximum diameter of 1.8 cm (Supplementary Figure 24).

Histological findings

Histology revealed a grade III, poorly differentiated, large cell neuroendocrine carcinoma, with focal glandular differentiation, of likely local origin in view of thyroid transcription factor-1 negativity.

Management

The patient underwent microlaryngoscopy and transoral laser debulking of the subglottic mass, with ablation of the base (Supplementary Figure 25). No further lesions were identified in the trachea or at the carina. The airway appeared normal at subsequent follow up. Multidisciplinary team discussion planned for positron emission tomography scanning for formal staging, followed by chemoradiotherapy.

Discussion

The importance of maintaining effective cross-specialty communication was emphasised. Dr Sandison highlighted the forthcoming change in World Health Organization classification, with laryngeal large cell neuroendocrine carcinomas being classified as their own entity.

Conclusion

Specialist referral is appropriate if a patient's symptoms do not resolve with initial management.

A rare cause of dysphonia

M Ally, A Kinshuck, G Sandhu

From the Charing Cross Hospital, London

Introduction

Lipoid proteinosis also known as Urbach–Wiethe disease, is a rare autosomal recessive disorder caused by mutations in the extracellular matrix protein 1 gene. It is characterised by deposition of hyaline in the skin and mucous membranes.

Case report

A 51-year-old ex-smoker, who was a former intravenous drug user, with a background of asthma, presented with dysphonia. Examination of the oral cavity revealed submucosal lesions, of less than 1 mm in size, on the hard palate, soft palate, tonsils and pharyngeal wall. Flexible nasoendoscopy revealed multiple exophytic lesions affecting the supraglottis.

Radiological findings

Computed tomography revealed a thickening and irregularity of the laryngeal mucosa.

Histological findings

Histopathological analysis with periodic acid–Schiff stain (Supplementary Figure 26) demonstrated mucosa with stromal hyalinisation, acutely inflamed hyperplastic squamous epithelium and candida-type organisms, consistent with the diagnosis of acute inflammation associated with candida on a background of lipoid proteinosis. There was no evidence of dysplasia or malignancy.

Management

The patient underwent suspension microlaryngoscopy with laser debulking of the supraglottic lesions using a 'pepper pot' technique. This has been performed on two separate occasions, with the patient reporting an improvement in their voice and breathing.

Discussion

Dr Sandison highlighted the characteristic deposition of perivascular eosinophilic material on biopsy (Supplementary Figure 26). She emphasised that a cutaneous or mucosal biopsy is required to confirm this rare diagnosis, and that this systemic disease can affect other organs.

Conclusion

Although there is currently no known cure for lipoid proteinosis, laryngeal symptoms can be controlled with the judicious use of laser debulking. The 'pepper pot' technique spares mucosa and prevents stenosis, whilst controlling the disease.

Supplementary Material

The supplementary material for this article can be found at https://doi.org/10.1017/S0022215117002559.