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Proceedings of the 147th Semon Club, 12 May 2014, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Chairperson: Miss Elfy B Chevretton, 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, Guy's and St Thomas' NHS Foundation Trust

Dr Ata Siddiqui, Guy's and St Thomas' NHS Foundation Trust

Professor Leslie Michaels prize for the best presentation of the meeting was awarded to Maha Khan for 'Truly malignant otitis externa'.

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.

Otology section

Chairperson: Mr Rupert Obholzer

Chronic sinus discharge secondary to a parotid 'rock'

J Long, R Oakley

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

Case report

A 67-year old lady presented to the clinic with a 55-year history of a left parotid lump. This had become increasingly troublesome over the last year and was discharging onto the skin. There was weakness of the left marginal mandibular nerve and a 1 cm lesion in the parotid tail that was ulcerating onto the skin surface. Oral examination findings were unremarkable. There was no lymphadenopathy. The patient underwent imaging and an excision biopsy two months later.

Radiological findings

Ultrasound showed a large irregular calcified mass within the parotid measuring 3×3 cm. It was not amenable to fine needle aspiration. Computed tomography and magnetic resonance imaging showed a destructive lesion within the parotid, with loss of cortical bone from the mandible.

Histological findings

Left total parotidectomy and parapharyngeal space dissection were performed. A 6 mm Warthin's tumour was discovered incidentally within the gland. Other sections showed patchy, chronic, non-specific inflammation and multiple foci of mineralisation, of unclear origin, extending onto the ulcerated skin surface. There was no evidence of malignancy. Six reactive lymph nodes were identified.

Management

Despite no origin or cause of the significant calcification being identified, and no definitive diagnosis being made, the patient continues to make a good recovery.

Discussion

Professor Howard commented that the long history is in keeping with a congenital aetiology. If the patient had a skin dimple near the lobule of the ear, or a pitted ear canal, that would point towards a first branchial arch anomaly. Dr Sandison commented that dermatomyositis could have caused the calcification.

Conclusion

The surgical sieve remains an essential diagnostic aid. In addition, one must not forget congenital causes.

Management of a parotid mass of uncertain diagnosis

F Holt, P Tsirevelou, N Gibbins

From the University Hospital Lewisham

Introduction

A case of a rare parotid mass that defied pre-operative investigations was presented.

Case report

A 60-year-old ex-smoker presented with a 3×3 cm left parotid mass that had developed over the preceding month. There was no facial nerve involvement. Co-morbidities included type II diabetes mellitus and hypertension.

Radiological findings

Left parotid ultrasound revealed a 2.8 cm cystic mass. Necrotic material was aspirated from this mass. Neck computed tomography (CT) confirmed this finding, but no primary source of the suspected squamous cell carcinoma (SCC) was identified.

Histological findings

Initial aspiration cytology showed atypical squamous cells suggestive of SCC. However, failure to find a primary tumour on CT prompted repeat aspiration, which yielded neither atypical nor malignant cells.

Following multidisciplinary team discussion, superficial parotidectomy was performed. Histopathology demonstrated Warthin's tumour with squamous metaplasia, with surrounding fibroblastic proliferation and chronic inflammation. Dr Sandison commented that fine needle aspiration cytology (FNAC) can cause a tract of granulation tissue that is easy

to misdiagnose histologically as SCC. When asked, she also commented that there was no evidence that FNAC can cause infarction. Mr Obholzer noted that in this case the swelling and cystic expansion started five weeks prior to the FNAC, so this was a case of spontaneous infarction.

Management

After discussing the benignity of the diagnosis with the patient, no further management was required.

Discussion

Infarcted metaplastic Warthin's tumours are clinically important and diagnostically difficult: appearances can masquerade as malignant tumours such as mucoepidermoid and SCC (primary and secondary). Furthermore, malignancy and Warthin's tumour can co-exist.

Professor Howard noted that although SCC of the parotid is very rare, primary SCC of the scalp and external auditory canal, with metastasis to the parotid area, are increasing in incidence.

This case challenged suggestions that aspirations cause infarction of Warthin's tumours. It is possible that there were relevant patient risk factors for microvascular disease.

Conclusion

Disentangling infarcted metaplastic Warthin's tumours from malignant differentials is challenging: a multidisciplinary approach is key.

Truly malignant otitis externa

M M Khan, R Hall, S Agrawal

From the Pennine Acute Hospitals NHS Trust, Manchester

Introduction

We present a case of malignant otitis externa, which, in retrospect, had atypical clinical and radiological features. This led to an alternative diagnosis.

Case report

An 82-year-old, non-diabetic lady presented with a 3-month history of right-sided otorrhoea and painful external ear canal granulations. Symptoms were refractory to microsuction and topical treatment. Whilst awaiting examination under anaesthesia, she developed complete facial nerve palsy with cessation of otalgia. Intravenous antibiotic therapy for malignant otitis externa was commenced.

Radiological findings

The findings of computed tomography and magnetic resonance imaging (MRI) of the temporal bones were considered consistent with malignant otitis externa. High signal intensity in the mastoid antrum on MRI was thought to represent a collection of symptoms. Drainage was advised. Careful pre-operative re-evaluation suggested spherical bone destruction consistent with tumour, rather than the linear canal erosion more commonly associated with malignant otitis externa.

Histological findings

Out-patient biopsies demonstrated inflammatory tissue with scant atypical cells, consistent with granulation tissue. Subsequent intra-operative debridement revealed necrotic rather than granulation tissue. Histology showed moderately differentiated squamous cell carcinoma (SCC) (tumour–node–metastasis (TNM) staging was $T_4N_0M_0$).

Management

Initial biopsy results were misleading. The short history of complaints in a previously normal ear culminating in facial nerve palsy was atypical of malignant otitis externa. Re-evaluation of imaging scans revealed findings consistent with malignancy.

Discussion and lessons learned

Dr Connor commented that malignant otitis externa often spreads anteriorly to affect the retrocondylar fat early on, whereas SCC is more likely to spread posteriorly and cause lytic lesions of the temporal bone. Dr Sandison commented that in view of the original clinical information, cellular dysplasia in early tissue samples could be interpreted as inflammatory, but in retrospect this represented malignant change. Mr Obholzer said it was not unusual for SCC of the external auditory canal to be initially misdiagnosed, and recommended not including any suspected diagnosis on investigative request forms, only the clinical details. Professor Howard felt that the clinical history, in terms of the rapidity of the facial nerve palsy and failure to respond to therapy, were not in keeping with malignant otitis externa.

Conclusion

Obtaining quality biopsies from multiple sites under general anaesthesia is essential.

Extra-axial chordoma of the paratracheal and paravertebral space

Z Nemeth, J Hughes, P M Clarke

From the Imperial College Healthcare NHS Trust, London

Case report

A 67-year-old gentleman presented with a large neck mass. Investigations revealed a malignant tumour involving the brachial plexus and vertebral artery.

Imaging findings

Magnetic resonance imaging showed a lesion within the left paratracheal soft tissues, extending from the larynx down to the thoracic inlet, with displacement of the trachea, carotid and jugular vessels. There was extension of the lesion into the C5–C6 epidural space and the C6–C7 vertebral bodies. The left C6 nerve root was splayed by the lesion.

Pathology findings

The tumour showed notochordal differentiation, with expression of cytokeratin, epithelial membrane antigen, S100 protein and brachyury, the latter of which is a transcription factor essential for the formation of posterior mesodermal tissue. Dr Sandison said the likely diagnosis was a chordoma, which had been confirmed by Professor Cyril Fisher from the Royal Marsden Hospital.

Management

The tumour was completely excised, in conjunction with the neurosurgical team, through a lateral neck approach, with stabilisation using a C5–T1 corpectomy cage and a plate.

Discussion

Mr Obholzer commented that perhaps the tumour was arising from the vertebral bodies. Dr Connor felt that the radiological findings were consistent with a vertebral origin. Dr Sandison commented that although chordomas which do not arise from the top or bottom of the axial

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skeleton are generally extra-axial, in this case it was an axial tumour, and it was unusual in that it presented as a neck mass. Professor Howard said that one-third of chordomas occur around the sphenoid bone, and the UK is in need of more clinical proton beam radiotherapy units to treat such patients.

Conclusion

Chordoma is a very rare, low-grade malignant bone tumour, arising from the remnants of the embryonic notochord. Small numbers of chordoma-like tumours develop in the extra-axial skeleton, called extra-axial chordomas.

Unilateral hearing loss and mandibular numbness

S Timms, R Obholzer

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

Introduction

Paraganglioma of the middle ear typically spreads either towards the mastoid or the jugular bulb. We present a paraganglioma extending anteriorly, through the eustachian tube into the post-nasal space.

Case report

A 45-year-old woman presented with a 4-month history of left-sided otalgia and hearing loss. On examination, an external ear canal mass was seen, along with a polypoid mass in the post-nasal space. Ipsilateral mandibular numbness was noted.

Radiological findings

Magnetic resonance imaging showed the ear and nasal mass to be continuous via the eustachian tube, with possible parotid involvement. Positron emission tomography computed tomography showed low activity, excluding malignancy.

Histological findings

A biopsy taken in clinic suggested a fibroepithelial polyp. However, histology of a specimen taken under general anaesthesia indicated paraganglioma with atypical features. The surgical specimen confirmed the diagnosis. Dr Ann Sandison pointed out the features of paraganglioma visible on S100 stain: cell clusters called zellballen, surrounded by sustentacular cells.

Management

The tumour was resected. During recovery on the ward, an axillary mass was noted and found to be part of a diffuse B-cell lymphoma, which has since been treated.

Discussion and conclusions

The atypical anatomical, radiological and histological features of the tumour, and the co-existence of lymphoma, created diagnostic difficulties in this case. Professor David Howard commented on the enormous range of pathology that can arise within ENT, and stressed the importance of obtaining quality biopsy material.

Rhinology section

Chairperson: Miss Jo Rimmer

A bleeding unilateral nasal polyp: cancer surely!

N Haloob, A Vijendren, M Salam

From the Ipswich Hospital

Introduction

Angiomyolipomas are hamartomas composed of smooth muscle, with vascular and fatty elements. They are commonly found in the kidney and liver, and rarely elsewhere. There is an association with tuberous sclerosis. To date, only eight cases in the nasal cavity have been reported.

Case report

A 49-year-old gentleman presented to the ENT out-patients department with a 2-year history of worsening unilateral nasal congestion, which was refractory to medical therapy. Clinical examination revealed a polypoidal mass within the right nasal cavity that bled upon contact. An urgent computed tomography scan of the sinuses and an excision biopsy were arranged.

Radiological findings

A large obstructing polypoidal mass was shown contained within the right nasal cavity, surrounding the inferior turbinate, which deviated the septum to the left. Bilateral sinonasal disease was also observed.

Histological findings

The tissue fragments consisted of a complex mix of adipose and spindle cells, and thick-walled abnormal blood vessels, in keeping with an angiomyolipoma.

Management

Intra-operatively, a smooth and firm mass was found arising from the lateral wall of the inferior meatus. It was excised in totem and bilateral functional endoscopic sinus surgery (FESS) was performed. Post-operative epistaxis was managed conservatively.

Discussion

Professor Howard and Miss Rimmer agreed that a diagnostic examination under anaesthesia and biopsy of the mass would have been adequate; contralateral FESS was unnecessary, as malignant pathology could not be excluded.

Conclusion and take-home message

Patients with a unilateral bleeding nasal polyp should be investigated on an urgent basis with appropriate imaging and high quality biopsies performed in the operating theatre, as both benign and malignant pathologies can have similar presentations.

Beware of adhesions – always consider a biopsy

M Lechner, N Choudhury, P Andrews

From the Royal National Throat, Nose and Ear Hospital, University College London Hospitals NHS Trust

Background

Nasal adhesions are a troublesome complication of nasal surgery. The post-operative use of Silastic splints is regarded as effective in the prevention of new nasal adhesions and to some extent when used for established lesions. If intranasal

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adhesions occur post-operatively, these are commonly divided under general anaesthesia (GA). However, biopsies are not performed routinely by the operating surgeon in most cases.

Case presentation

Here, we present two cases of patients who presented with recurrent adhesions, but who had rare underlying pathology.

The first case is that of a 33-year-old lady who underwent external septorhinoplasty and subsequently presented with nasal adhesions. These were divided under GA. Because they recurred, a biopsy was taken. The findings confirmed the diagnosis of eosinophilic angiocentric fibrosis.

The second case is that of a 51-year-old gentleman who presented with adhesions of the nose after septoplasty. These recurred over years. The patient was then referred to our centre, where biopsies revealed the diagnosis of angioleiomyoma.

Discussion

Here, we present two patients who suffered from recurrent intranasal adhesions despite the use of splints. These patients were found to have rare underlying conditions: eosinophilic angiocentric fibrosis and angioleiomyoma. The patients would have been diagnosed at an earlier stage if examination of underlying pathology and a biopsy had been considered. In the course of the discussion, Miss Jo Rimmer, who has recently published on eosinophilic angiocentric fibrosis, mentioned that the latest research shows eosinophilic angiocentric fibrosis is part of the spectrum of immunoglobulin G4 related systemic diseases.

Conclusion

These two exemplar cases clearly underscore the importance of considering biopsies in patients presenting with recurrent intranasal adhesions.

Progressively enlarging neck lump: when an abscess is not an abscess. A case report

M Ellabib, N Foden, T Jacob

From the University Hospital Lewisham

Introduction

Rhabdomyosarcoma is an aggressive tumour of skeletal muscle origin. It accounts for half of soft tissue malignancies in children, but is rare in adults.

Case report

We report the case of a 44-year-old lady, who presented with a 3-week history of a left-sided neck lump. The patient was systemically well. She was initially suspected to have a neck abscess and was treated with intravenous antibiotics. Flexible nasendoscopy findings were normal. The ultrasound suggested an abscess. Aspiration was performed, which revealed no growth on microbiology and inconclusive results on cytology. The patient also began to complain of epiphora.

Radiological findings

After an inconclusive initial computed tomography (CT) scan, a further head and neck CT scan showed a large, aggressive left nasal tumour extending to the left orbit, with multiple enlarged lymph nodes in the left side of the neck. Comparisons to the previous CT scan showed marked progression of the disease.

Histological findings

Incisional biopsy of a left cervical lymph node demonstrated a non-haemopoietic tumour, with small round cells, showing rhabdomyoblastic differentiation. Cluster of differentiation 56 positivity, a strongly positive staining for desmin and myogenin, confirmed skeletal muscle differentiation. These findings were in keeping with alveolar rhabdomyosarcoma.

Management

The patient was referred to a maxillofacial surgeon, who in turn referred her to a sarcoma specialist. The patient is currently undergoing chemotherapy.

Discussion

As discussed by Professor D Howard, understanding head and neck anatomy is key to elucidating clinical manifestations of disease. In our case, epiphora and node stage N_3 neck lymphadenopathy would suggest nasopharyngeal involvement. This was not realised until the second CT scan. Dr Sandison reminded the audience that sarcomas tend not to metastasise to lymph nodes, but rhabdomyosarcoma is an exception to the rule.

Conclusion

Because of its late, non-specific clinical manifestations, rhabdomyosarcoma is often difficult to diagnose and treat.

Maxillary mass associated with diplopia and proptosis following extraction of teeth

S Al-Hashemi, R Nash, G Watters

From the Southend University Hospital NHS Foundation Trust

Introduction

Maxillary masses may be caused by odontogenic tumours or carcinomas. Inflammatory maxillary pseudotumour is a very rare tumour-like lesion, which is easily mistaken for maxillary carcinoma. It is histologically characterised by variable proportions of spindle cells, foam cells, lymphocytes and plasma cells. Treatment involves surgical debridement and glucocorticoid therapy.

Case report

We present a case of a 59-year-old female with proptosis diplopia and left maxillary bone expansion. She initially presented with an oral antral fistula and non-healing sockets following the extraction of the upper left posterior teeth. The clinical findings were initially misinterpreted as being due to a malignant neoplasm.

Radiological findings

Computed tomography and magnetic resonance imaging confirmed a diffuse osteolytic lesion affecting the left maxillary antrum, destroying the left side of the hard palate and anterior part of the left zygoma. Soft tissue expansion had destroyed the orbital floor and infiltrated the inferior rectus muscle.

Histological findings

Initial histopathology was reported as showing a chronic inflammatory lesion with fungal infection. Subsequently, this was described as a hypercellular, spindle cell lesion associated with an inflammatory component. Dr Sandison demonstrated actinomyces and candida species on the surface of the lesion that had not penetrated too deeply. She agreed that this constituted an inflammatory pseudotumour.

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Management

A diagnosis of maxillary pseudotumour was made following discussion at the multidisciplinary meeting. There was a debate as to whether this condition constituted part of the spectrum of immunoglobulin G4 related diseases. The management for both, however, involves glucocorticoid therapy.

Surgical debridement with partial maxillectomy, followed by a course of prednisolone, improved the patient's symptoms. A prosthetic obturator was made to restore the alveolar defect. Disease activity was monitored clinically with imaging and serum inflammatory markers.

An unusual lateral nasal wall mass

J Long, J Rimmer, D Roberts

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

Introduction

This case highlights the importance of a wide differential diagnosis in the assessment of head and neck masses.

Case report

A 56-year-old male was referred with a 2-year history of a left lateral nasal wall swelling. There was no history of trauma and no nasal symptoms. He was otherwise fit and well, with no allergies. Flexible nasendoscopy and neck examination findings were normal.

Radiological findings

An ultrasound scan of the swelling showed a subcutaneous cystic lesion measuring 16×7 mm, with blood vessels crossing the lesion. Magnetic resonance imaging showed a non-specific lesion within the superficial soft tissues, with no evidence of deep extension.

Histological findings

The patient underwent excision of the lesion. Histological findings showed multiple fragments of skeletal muscle and adipose tissue, with large lymphoid follicles arranged around a central large artery.

Management

An elevated eosinophil count found on histology raised the suspicion of Kimura's disease and prompted measurement of serum immunoglobulin E (IgE). The diagnosis was subsequently confirmed as Kimura's disease. The patient is making a good recovery.

Discussion

Kimura's disease is a benign, rare, chronic inflammatory disorder that typically affects Asian males. It is characterised by: painless subcutaneous masses in the head and neck region, blood and tissue eosinophilia, and a raised serum IgE level. Patients are often extensively evaluated for neoplasia prior to this diagnosis.

Conclusion

Although Kimura's disease is rare in the UK, it is relatively common in Asia and should be considered in this patient population.

Head and neck section

Chairperson: Professor David Howard

An unusual cause of obstructive sleep apnoea like symptoms requires novel management

R J Glore, S Mortimore

From the Derby Teaching Hospitals NHS Foundation Trust

Introduction

We report the first case in the English literature of epiglottic polymorphous, low-grade adenocarcinoma treated using transoral laser excision.

Case report

A 38-year-old lady underwent tonsillectomy for obstructive sleep apnoea. An epiglottic mass was identified during the operation and was biopsied.

Radiological findings

A computed tomography scan confirmed a mass or tumour confined to the epiglottis, with no evidence of regional or distal metastasis.

Histological findings

First described in 1984, polymorphous, low-grade adenocarcinoma is a slow-growing, malignant, minor salivary tumour that is locally invasive. Despite being malignant, this tumour rarely metastasises. Polymorphous, low-grade adenocarcinoma is characterised by infiltrative growth, morphological diversity and cytological uniformity. The commonest sites affected are the palate and upper lip. The differential diagnoses are pleomorphic salivary adenoma and adenoid cystic carcinoma.

Management

The published evidence suggests that surgical excision offers the best chance of cure. There is no indication for neck dissection in a clinically node negative (N_0) neck. The patient underwent successful transoral laser resection of the epiglottic tumour. Histology results confirmed complete excision with good margins.

Discussion and lessons learned

Professor Howard agreed that this type of tumour should be surgically excised and that transoral laser resection is a reliable approach, with limited risks and morbidity. He also commented that surgical excision without adjuvant chemotherapy and/or radiotherapy is suitable for most low-grade adenocarcinomas.

Conclusion

The evidence suggests that the treatment of choice for polymorphous, low-grade adenocarcinoma is surgical excision. We have demonstrated that transoral laser resection allows full excision of the tumour with laryngeal preservation.

Acute airway obstruction in a young patient with uveitis and seronegative arthritis

J G Barr, K Kapoor, R Simo

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

Introduction

Relapsing polychondritis is a rare autoimmune condition that affects the airway in 20 per cent of cases.

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Case report

A 19-year-old man, with seronegative arthritis and uveitis, presented with stridor. He had a one-month history of worsening hoarseness, shortness of breath and night sweats. Inflammatory markers were raised. In light of imminent airway compromise, an emergency tracheostomy was performed. Panendoscopy showed a supraglottic mass, which was biopsied. The tracheal window and supraglottic biopsy samples were sent for histological analysis.

Radiological findings

Dr Siddiqui highlighted endolaryngeal circumferential thickening with a normal trachea on the computed tomography (CT) scan. Chest CT revealed minor inflammatory changes at the left lung base. A positron emission tomography scan did not show significantly increased uptake within the laryngeal mass.

Histological findings

There was extensive destruction of cartilage by a mixed inflammatory cell infiltrate in which macrophages and neutrophils were predominant. There was no evidence of vasculitis, granulomas, amyloid deposits or necrosis. The appearances were those of an inflammatory chondritis, likely to represent relapsing polychondritis.

Management

Following multidisciplinary input with the connective disorders team, treatment with oral corticosteroids was commenced. There has since been some resolution in the patient's symptoms. However, he remains tracheostomy-dependant.

Discussion

Immunosuppressive agents are key to managing relapsing polychondritis. Professor Howard reminded the audience that steroid-sparing agents such as azathioprine should be considered. He noted that polychondritis can cause both heart and tracheal disease, which can be life threatening. As the patient remains dependent on a tracheostomy, Professor Howard suggested referral to a specialist airway surgeon for consideration of laser treatment to the supraglottis or an airway stent. Overall, such patients have a poor prognosis.

Conclusion

Relapsing polychondritis is a rare condition. However, it is an important condition for the otolaryngologist to be aware of as it predominantly affects the cartilages of the ear, nose and laryngotracheobronchial tree. Multidisciplinary input is important for optimal management.

Exploding tyre blast injury causing persistent odynophagia

S M Field, J G Manjalay, T S Tatla

From the Northwick Park Hospital, Middlesex

Introduction

Pharyngoesophageal perforation secondary to barotrauma is a rare phenomenon that has serious complications if diagnosis is delayed. It is challenging to detect in the acute setting because of the non-specific symptoms. We present a case in which its detection proved difficult and led to a delay in diagnosis.

Case report

A 27-year-old male mechanic presented to the accident and emergency department with minor respiratory difficulty and haemoptysis after a car tyre exploded in his face. On examination, there was no other evidence of injury and he was systemically well. Flexible nasoendoscopy (FNE) revealed blood in the posterior pharynx, thought to be secondary to mucosal haemorrhage. He was started on intravenous antibiotics. In light of persistent odynophagia, with normal repeat FNE findings, a soft diet was commenced. However, after 3 days, complete dysphagia and odynophagia persisted, prompting computed tomography (CT) of the neck.

Radiological findings

The CT scan revealed a large defect in the posterior hypopharynx. There was extensive surgical emphysema in the deep neck tissues, down to the level of the mediastinum.

A contrast swallow test confirmed a posterior hypopharyngeal leak, showing liquid tracking down the prevertebral space.

Management

The patient was commenced on nasogastric feeding for one week. A repeat contrast swallow test confirmed resolution of the defect and he was discharged.

Discussion and conclusion

Pharyngeal perforation is not always detectable on FNE. Clinicians should have a low threshold for further investigation after barotrauma and consider keeping patients nil-by-mouth until a perforation is excluded. Professor Howard reminded us that it takes significant pressure for a tyre to explode – patients with this mechanism of injury should be observed closely, even if asymptomatic. He also noted that a lateral soft tissue neck X-ray may have aided an earlier diagnosis.

Progressive dysphagia to solids over a number of years

C McIntyre, J Hughes, G Sandhu

From the Charing Cross Hospital, Imperial College Healthcare NHS Trust, London

Introduction

Granular cell tumour, first described by Abrikossoff in 1926, is a relatively uncommon neoplasm. It is predominantly found in the head and neck region. Pharyngeal granular cell tumour is extremely rare, with only 3 cases reported in the literature in the last 50 years. Granular cell tumours are slow-growing, benign, subcutaneous tumours that can become symptomatic with increasing size. Surgical excision is the treatment of choice for these radio-resistant tumours.

Case report

A 50-year-old female was referred with a pharyngeal mass found on oesophago-gastro-duodenoscopy whilst investigating her symptom of progressive dysphagia to solids over a number of years. She reported having to tilt her head backwards whilst eating to manoeuvre food through her pharynx. Flexible nasopharyngoscopy revealed a large submucosal pharyngeal mass.

Radiological findings

A computed tomography scan of the patient's neck demonstrated a bulky mass centred on the posterior wall of the hypopharynx, measuring 2.5 cm in maximum craniocaudal extension. The mass severely narrowed the supraglottic airway and displaced the aryepiglottic folds anteriorly.

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Histological findings

Microscopic sections showed squamous epithelium overlying fibrous stroma, which contained a tumour. The tumour was composed of a proliferation of large cells, with abundant granular eosinophilic cytoplasm. The appearances were consistent with a granular cell tumour.

Management

Micro-pharyngoscopy and carbon dioxide laser excision were performed to completely excise the tumour. The patient made a good post-operative recovery and will continue to be followed up because of the risk of recurrence of these tumours.

Conclusion

Although rare, these tumours should be considered when faced with a pharyngeal mass.

Tongue lesion in a patient with metastatic adenocarcinoma of the bladder undergoing chemotherapy

A I Kaleva, R W A Hone, I J Nixon

From the William Harvey Hospital, East Kent Hospitals University NHS Foundation Trust, Ashford

Introduction

Chemotherapy may cause intra-oral ulceration, but a thorough investigation of symptoms is important to determine the underlying cause.

Case report

A 61-year-old gentleman presented with a painful tongue, causing dysphagia. He was receiving chemotherapy for a bladder adenocarcinoma with chest wall metastasis. His general practitioner had treated him with mouthwash and steroids. On examination, the tongue surface appeared normal, but there was a palpable mass on the left dorsum. The mass was biopsied under local anaesthetic.

Radiology

Magnetic resonance imaging of the head and neck revealed an isolated mass, $37 \times 31 \times 30$ mm in size, within the left tongue base, with no regional lymphadenopathy.

Histology

The biopsy showed an infiltrating, poorly differentiated adenocarcinoma. Dr Sandison noted that the lesion undermined the epithelium, but the basal layer was preserved. The findings were in keeping with a high-grade minor salivary gland adenocarcinoma or a metastatic deposit.

Discussion

After initial discussion at the head and neck multidisciplinary team meeting, the tongue adenocarcinoma was considered a second primary. Review of previous histology confirmed it as a metastasis from the bladder primary. The patient was referred back to the urology team, who continued his palliative chemotherapy.

There is only one other documented case report of a bladder primary metastasising to the tongue.

Conclusion

This case highlights the importance of comparing histological specimens to check metastatic spread (however unlikely) to allow appropriate treatment planning. Semon Club discussion emphasised the importance of quality histological samples. In this case, an adequate biopsy was obtained under local anaesthetic, but this is not always possible.

This case teaches us to assess each new symptom carefully and in the context of a patient's medical history.

Paediatric section

Chairperson: Mr Ian Hore

A rare cause of paediatric conductive hearing loss

J G Barr, P Singh, A Fitzgerald O'Connor

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

Introduction

We present a rare case of paediatric conductive hearing loss due to a high, lateralised jugular bulb.

Case report

A five-year-old boy presented with right-sided hearing loss. He had normal speech development, no tinnitus and no other medical history. On examination, his right tympanic membrane was retracted, and there was evidence of middle-ear effusion. His left tympanic membrane was normal. He had a conductive hearing loss down to 40 dB on his right side, with normal hearing in the left ear.

The patient continued to be reviewed and was consistently found to have right-sided conductive hearing loss. On subsequent examination, a pink bulge was seen towards the inferior part of the tympanic membrane.

Radiological findings

A computed tomography scan showed a high and lateralised right jugular bulb, which had a superolaterally pointing diverticulum that bulged into the lower mesotympanum and posterior external auditory meatus. Additionally, there was some subtle bony dehiscence overlying the diverticulum. The ossicular chains were intact bilaterally and the middle-ear clefts were otherwise normal.

Management

It was decided that no active management was needed. The child's parents were informed of the importance of not putting objects into the ear.

Discussion

If myringotomy was performed in error then Professor Howard recommended that packing the ear would be the best initial management for the bleeding.

Conclusion

Conductive hearing loss associated with a high, lateralised jugular bulb with bony dehiscence is rare. Correct diagnosis ensured that inappropriate intervention did not occur.

Unexpected findings in sudden-onset stridor with respiratory distress

E Schechter, I Bruce, M Rothera

From the Royal Manchester Children's Hospital

Introduction

Common causes of childhood stridor include laryngotracheomalacia, subglottic cysts, laryngotracheobronchitis and vocal fold palsy. This report highlights an unusual cause of airway obstruction in early childhood.

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Case report

A two-year-old girl was transferred to our tertiary referral centre with a one-month history of stridor, respiratory distress and blood-stained nasal discharge not responding to antibiotics. She had been developing normally, with no previous medical history or respiratory problems.

Rigid airway endoscopy demonstrated severe tracheitis, with a raised lesion extending from the distal trachea and occluding the left main bronchus. A representative biopsy was taken. The patient remained ventilated on intravenous antibiotics and steroids. Differential diagnoses included congenital narrowing, infection, a reactive lesion and tuberculosis. Bronchoalveolar lavage was sterile, and immunodeficiency and autoimmune screens were within normal limits. Gamma interferon tests were negative, but rhinovirus was detected on polymerase chain reaction analysis. Following extubation, a further endoscopy, balloon dilatation and repeat biopsy showed improvement in mucosal inflammation, with increased patency of the bronchus.

Radiological findings

A thoracic computed tomography scan (whilst intubated) showed: a small left-sided pneumomediastinum, areas of atelectasis and consolidation of both lungs.

Histological findings

Both biopsies showed respiratory and squamous epithelium, with underlying signs of inflammation.

Management

The patient is undergoing repeat endoscopy with high-dose methylprednisolone. Mucosal and tracheobronchial swelling is resolving.

Discussion and lessons learned

Mr Hore held the opinion that the diagnosis of idiopathic tracheitis and the ongoing management are correct.

A rare cause of cervical lymphadenopathy in adolescence

A Simmons, A Takhar, N Hyde

From the St George's Hospital, London

Introduction

This is the first reported case of an adolescent female with cribriform adenocarcinoma of the tongue and minor salivary glands affecting the hard and soft palate.

Case report

An 11-year-old girl presented to her general practitioner with a left, level II neck lump and symptoms of intermittent nausea. Following excision, histology indicated metastatic, polymorphous, low-grade adenocarcinoma within a lymph node, from an unidentified primary. The patient was subsequently referred to a tertiary head and neck cancer centre.

Examination at the tertiary centre demonstrated a firm, irregular mass in the left palate.

Radiology

Computed tomography and magnetic resonance imaging of the head and neck demonstrated a large lesion in the soft and hard palate, causing local bony destruction.

Histology

Histopathological analysis demonstrated an unencapsulated, malignant tumour. The predominantly solid and cribriform pattern was more accurately described by the term cribriform adenocarcinoma of tongue and minor salivary glands, thus the initial diagnosis was revised.

Management

The patient underwent a left Weber-Ferguson maxillectomy. She made a good recovery, with no immediate or early complications.

Discussion

Dr Sandison pointed out the histological features of cribriform adenocarcinoma of the tongue and minor salivary glands. Professor Howard commented that use of a mid-facial degloving technique may have avoided a facial scar. He also noted that the patient's face would continue to grow until the age of 16 years, but the fibula graft would not grow with the face. Professor Howard felt that the use of a prosthesis instead of the fibula graft may have allowed a more thorough examination of the patient for recurrent disease at follow up, and facilitated changes in the size of the prosthesis to accommodate facial growth.

Conclusion

This is the youngest reported case of cribriform adenocarcinoma of the tongue and minor salivary glands. However, a helpful lesson was learned from the need to revise the polymorphous, low-grade adenocarcinoma diagnosis.

A 12-year-old with nasal obstruction and rhinorrhoea

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From the Colchester Hospital

Introduction

The diagnosis of polypoid nasal lesions can be very challenging, especially in unusual or rare cases in which benign lesions mimic malignancy. A multidisciplinary approach is essential for those lesions presenting with unusual clinical or histological appearances.

Case report

A 12-year-old boy presented with a 2-year history of nasal obstruction and rhinorrhoea that was refractory to medical treatment with topical nasal steroids. Examination revealed a polypoid lesion, with atypical anatomy in the right nasal cavity.

Imaging findings

Computed tomography and magnetic resonance imaging demonstrated an expansile lesion, extending up to the skull base but not penetrating the cribriform plate. The differential diagnoses included fibrous dysplasia and rhabdomyosarcoma.

Pathology findings

The sections showed inflamed respiratory mucosa containing a variably cellular tumour. The tumour was composed of fibrous and myxoid stroma, with areas of nodular hyaline and chondroid tissue. Trabeculae of bone were present throughout and focally formed a rim at the periphery of the tumour. No necrosis was seen and mitoses were not a feature. Where it could be assessed, the tumour was well circumscribed in stroma, and no permeation of pre-existing bone was seen.

Immunostaining showed diffuse expression of S100, with focal expression of beta-catenin and clusters of differentiation 10 and 99. The proliferation rate, assessed with Ki-67, was less than 5 per cent. A wide panel of other immunostains were negative.

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The preferred diagnosis was benign chondromesenchymal hamartoma.

Management

The lesion was completely removed endoscopically. No further adjuvant treatment was recommended and the patient was followed up.

Discussion

The clinical setting here is unusual; the lesion usually occurs in young infants, but has been described in older children and adults. There is a predilection for males. The most common presentation is a mass lesion arising from the nasal septum or vestibule. In a small series of four cases occurring in older children and young adults, reported in the University of Pittsburgh (USA) literature, there was no recurrence following excision.

Conclusion

Chondromesenchymal hamartoma is an exceptionally rare tumour, most commonly reported in infants under the age of one year. It is benign, but may be mistaken clinically and histologically for malignancy. Awareness of the lesion is important in order to avoid unnecessary and potentially harmful therapies.

Rapid expansion of a previously benign neck mass in a child with neurofibromatosis type 1

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Introduction

A 13-year-old girl, known to suffer from generalised neurofibromatosis type 1 (NF1) since the age of 9 months, was referred to the Evelina Children's Hospital for an urgent assessment of a rapidly enlarging neck mass. This was causing severe compromise of the airway along with severe dysphagia. The swelling was initially slow-growing, and was first noticed when she was 16 weeks old. There was no family history of NF1.

Imaging

An 18 F-fluorodeoxyglucose positron emission tomography scan showed increased uptake in the region of the mass. On magnetic resonance imaging scans, Dr Siddiqui highlighted an initially lobulated tumour not respecting tissue planes, with later scans showing rapid growth and cystic changes, indicative of an aggressive process. Bronchoscopy revealed a fast, severe anterior displacement and clockwise rotation of the epiglottis. The patient underwent an urgent tracheostomy to secure her airway. Ultrasound-guided core biopsy of the mass was performed for tissue diagnosis.

Histology

Histology revealed a very large, benign plexiform neurofibroma, with areas of malignant peripheral nerve sheath tumour. The child was subsequently referred to a specialised paediatric oncology unit, where she is currently undergoing palliative chemotherapy treatment.

Discussion

Benign plexiform neurofibromas are often very large and can grow from nerves either in the skin or internal organs. The risk of change into malignant peripheral nerve sheath tumours is documented in the scientific literature. Patients with NF1 have a decreased overall survival rate because of this malignant potential. Between 8 and 13 per cent of patients with NF1 will develop malignant peripheral nerve sheath tumour over their lifetime. Some studies suggest that the mean age (in years) of onset of malignant peripheral nerve sheath tumour is around mid-30s.

Conclusion

Benign plexiform neurofibromas that change into malignant peripheral nerve sheath tumours in young children are challenging tumours to treat. This is because of the anatomical vicinity either to the airway or important vascular bundles and nerves. A definitive histological diagnosis and multidisciplinary team discussion are therefore of paramount importance for defining the treatment pathway. Furthermore, when the tumour mass is large, palliative chemotherapy and radiotherapy are of little value in the treatment of this malignant tumour.

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