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Proceedings of the 136th Semon Club, 14 November 2008, Otolaryngology Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

Chairman: Miss Elfy B Chevretton, Consultant ENT Surgeon, Guy's and St Thomas' NHS Foundation Trust.
Pathologists: Professor Leslie Michaels, and Dr Ann Sandison, University College London.
Radiologists: Dr Steve Connor, Consultant Radiologist, and Dr Daniel Bell, STR in Radiology, Guy's and St Thomas' NHS Foundation Trust.
Secretary: Mr Sherif Haikel, STR in ENT, Guy's and St Thomas' NHS Foundation Trust.
A prize was awarded for the best presentation of the meeting, to Mr Colin Butler.

Otology and skull base session

Chairman: Professor Michael Gleeson

Longstanding otorrhoea with facial nerve palsy

K Patel, J R Tysome, D Jiang
From Guy's and St Thomas' NHS Foundation Trust, London.

Introduction
In patients presenting with otorrhoea and facial nerve palsy, diagnoses to consider include malignant otitis externa, cholesteatoma and neoplasms. We present a case in which the histological diagnosis was challenging, and had not been established at the time of writing.

Case report
A 65-year-old man presented to the otology unit with a three-month history of right-sided hearing loss, malodorous otorrhoea, intermittent vertigo and weight loss. On examination, his external ear canal was inflamed and occluded; he had a mixed severe hearing loss and a House–Brackmann grade five right facial nerve palsy.

Radiology
Computed tomography and magnetic resonance imaging scans revealed a mass filling the right external ear canal and extending into the middle ear, with minimal bony erosion. Extra-axial extension was seen into the middle cranial fossa. A bone scan suggested osteomyelitis of the right petrous temporal bone.

Histology
A 1×1 cm biopsy taken from the right ear canal mass revealed numerous non-caseating granulomas with a predominance of mature lymphocytes.

Management
Oral ciprofloxacin and gentamicin and steroid ear drops were commenced to treat possible malignant otitis externa. Clinical improvement of the facial nerve palsy to grade two was observed.

Conclusion
Although the initial impression was of malignant otitis externa, Professor Michaels agreed that the histopathology was not conclusive, and a specialist review was recommended to exclude a rare form of lymphoma.

An interesting presentation of multiple cranial nerve palsies in an elderly diabetic

S Patil, I Rana, N Patel
From Whipps Cross University Hospital, London.

Introduction
Skull base osteomyelitis has high morbidity and mortality rates and is often difficult to treat.

Case report
An 80-year-old diabetic man presented with left ear pain, discharge and signs consistent with left necrotising otitis externa associated with IXth and Xth cranial nerve palsies. Microbiology confirmed pseudomonas. The patient was treated with 30 sessions of hyperbaric oxygen and adjuvant intravenous antibiotics, and was subsequently discharged home on oral ciprofloxacin for eight months. Six months later, he re-presented with increasing pain, ear fullness and deafness in the opposite ear. He was found to have a right middle-ear effusion, an oedematous right nasopharynx, and right IXth, Xth and XIIth cranial nerve palsies.

Radiology
Computed tomography (CT) imaging suggested a soft tissue swelling at the right skull base with bony erosion around the foramen magnum, clivus and jugular foramen.

Histology
A nasopharyngeal biopsy showed normal lymphoid tissue.

Management
At a multidisciplinary team meeting, it was decided to treat the relapse of skull base osteomyelitis with hyperbaric oxygen therapy and adjuvant intravenous antibiotics.

Conclusion and lessons learned
Professor Gleeson commented that the treatment of this condition is primarily medical, with antibiotics for at least one year. Regarding follow up, magnetic resonance imaging would help confirm soft tissue resolution, the CT being of use to detect early stage bony erosion. Professor Gleeson advised serial indium-labelled white cell scans to confirm disease control. The best indicator of resolution was felt to be reducing pain.

Parapharyngeal tumour presenting as stroke and diagnosed by ultrasound-guided, trans-oral biopsy

A Riskalla, S Khemani, P O'Flynn

From University College Hospital, London.

Case report

A 42-year-old woman was initially admitted with a right-sided stroke.

A magnetic resonance imaging scan revealed a mass most probably arising from the deep lobe of the parotid and extending to the parapharyngeal space.

Clinically, there was no evidence of any mass on palpation of the neck or intra-orally.

An angiogram confirmed complete occlusion of the right internal carotid artery. An ultrasound-guided fine needle aspiration was unsuccessful due to the deep nature of the tumour.

Under general anaesthesia, a prostatic ultrasound attachment was used trans-orally to obtain a tissue diagnosis, which suggested pleomorphic adenoma.

The mass was successfully excised via a trans-cervical approach. Histological analysis confirmed it to be a pleomorphic adenoma.

Discussion

Professor Gleeson thought it highly unlikely that this mass had caused a dissection which had progressed to stroke, particularly because of the mass's moderate size and low position within the neck. He also advised excision of the mass without biopsy, due to the high risk of seeding.

Cochlear implantation in a patient with otosclerosis

A Lee, D Jiang, A Fitzgerald O'Connor

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

Introduction

In otosclerosis cases, hearing loss can be secondary to stapes ankylosis and progressive sensorineural hearing loss. High resolution computed tomography (CT) has been shown to have a high sensitivity and specificity for the diagnosis of otospongiosis (see below). Cochlear implantation (CI) is a well established and cost-effective means of rehabilitating patients with deafness. Here, we present a case of CI in a patient with advanced otosclerosis.

Case report

Our patient was a 58-year-old man with bilateral progressive hearing loss due to otosclerosis. He had received a stapedectomy in 1982 yet his hearing continued to deteriorate. Audiological testing showed him to be a candidate for CI.

Radiology

This patient's CT scan showed bilateral fenestral otospongiosis with marked encroachment on the round window niches. There was no intrinsic cochlear calcification.

Management

A left-sided CI procedure was performed on 9 September 2008 using a straight electrode (Nucleus Freedom implant). The patient's round window was obliterated, yet we were able to create a cochleostomy and achieve full electrode insertion.

Conclusion

With careful pre-operative planning and good knowledge of the round window anatomy, it is possible to implant patients with advanced otosclerosis and to achieve satisfactory results.

Unilateral hearing loss and facial spasm

D Whitehead, Y Samam, M Gleeson

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

Introduction

A 15-year-old girl presented with progressive sensorineural hearing loss following a viral upper respiratory tract infection in 2002. Her hearing continued to deteriorate, and she noticed occasional tinnitus and vertigo that settled.

A magnetic resonance imaging (MRI) scan performed in South Africa at the time was reported as normal. Six years later, the patient presented to a UK ENT clinic with an intermittent, left-sided facial twitch and bilateral otalgia. A further MRI scan was arranged, which showed what appeared to be a vestibular schwannoma.

Management

A diagnosis of vestibular schwannoma was made, and the patient underwent a translabyrinthine resection in October 2008. However, at surgery an arteriovenous malformation was found at the internal acoustic meatus. The procedure was abandoned, with Surgicel being placed over the arteriovenous malformation. The dura was closed with temporalis fascia and the surgical defect obliterated with a free muscle and fat graft. The patient developed partial, left-sided hemiplegia five days post-operatively. An MRI scan showed no intracranial bleeding, but suggested that the arteriovenous malformation had thrombosed. This was confirmed by angiography. The patient's left-sided weakness resolved several weeks later.

Conclusion

Patients presenting with the classical symptoms and signs of a vestibular schwannoma but with the addition of facial twitching or pain should prompt suspicion that the lesion may be an arteriovenous malformation. Where there is suspicion of a vascular element, angiography should be considered pre-operatively.

Paediatric session

Chairman: Mr Ian Hore

A rapidly enlarging neck mass in an infant

S J Broomfield, I A Bruce, M P Rothera

From the Royal Manchester Children's Hospital.

Background

Although most neck lumps represent common and benign disease, it is important for otolaryngologists to consider malignancies and rare conditions.

Case report

A five-month-old male infant presented with a posterior triangle neck lump. Initial ultrasound scan and biopsy suggested a benign, self-limiting condition. Rapid enlargement of the lump prompted referral to a tertiary centre for definitive surgical excision.

Radiology

Initial ultrasonography showed a 3.5 cm, solid, vascular lymph node suggestive of tuberculosis or lymphoma. Four months later, a computed tomography scan showed an 8 cm lesion with features suggesting a soft tissue tumour.

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Histology

The initial biopsy was reported as a lymph node with reactive sinus histiocytosis. Subsequent, definitive histological analysis showed a tumour with morphological and immunohistochemical features of extra-abdominal fibromatosis.

Management

The lesion was completely excised. Tumour adherence to surrounding structures made facial nerve identification difficult, although the initial post-operative facial weakness subsequently resolved.

Conclusion and lessons learned

The treatment of fibromatosis was discussed, including the role of radiotherapy and chemotherapy for benign disease in children. Dr Sandison highlighted the importance of the proliferative index in distinguishing benign from malignant tumours in fibromatosis, where a spectrum of disease exists. Optimal post-treatment follow up was felt to comprise regular clinical examination for at least five years, with no role for routine imaging. This case demonstrates the importance of timely referral to a tertiary centre in cases of rapidly enlarging or atypical neck lumps.

Nasal septal pleomorphic adenoma in an adolescent, excised via a midfacial degloving approach combined with endoscopic assistance

I McKay-Davis, L Pitkin, A Hosni

From Frimley Park Hospital, Frimley.

Introduction

Pleomorphic adenomas originate from the minor salivary glands, and are rare in the nasal cavity. Surgical excision is the preferred treatment, and approaches include lateral rhinotomy, transpalatal, midfacial degloving and, more recently, endoscopic surgery.

Case report

A 16-year-old female presented with progressive, left-sided epistaxis over several months, associated with ipsilateral reduced nasal patency and cachosmia.

Clinical examination revealed a 2×3 cm, well circumscribed mass arising from the left septum areas two to five.

Radiology

Radiological examination revealed a soft tissue mass in the anterior aspect of the left nasal cavity in continuity with the nasal septum, extending laterally to the bony nasal wall, which showed some early demineralisation.

Histology

Histological analysis showed a mixed salivary adenoma with a predominant myoepithelial component. Tumour cells were positive for epithelial cell markers.

Management

The patient underwent excision via a combined endoscopic and midfacial degloving approach. The contralateral mucoperichondrium was left intact and supported with temporary, bilateral, Silastic® splints. The patient made an uneventful recovery.

Conclusion and lessons learned

The combined use of both an endoscopic and an open approach gave good access to the tumour, whilst reducing unnecessary resection and preserving cosmesis. Prof. Howard advised 25 years of endoscopic surveillance for such patients, as the prognosis regarding recurrent disease (which is known to present many years later) is poor regardless of treatment modality; the recurrence rate is in the order of 5–10 per cent.

A neutropenic child: a rare sequela of right-sided otalgia and squint

C R Butler, M S Ferguson, E Benjamin

From Charing Cross Hospital, London.

Introduction

We present the case of a child who developed rare complications of suppurative otitis media.

Case report

A seven-year-old boy presented with a two-week history of right-sided otalgia and frontal headaches, initially treated conservatively. Subsequently, he developed a rapid onset, right-sided squint. Clinical examination demonstrated a right abducens nerve palsy with ipsilateral otitis media.

Radiology

Computed tomography and magnetic resonance imaging revealed right-sided apical petrositis, mastoiditis, and extradural and infratemporal fossa abscesses with cavernous sinus thrombosis.

Management

Emergency cortical mastoidectomy was undertaken to allow surgical toilet. Post-operatively, the child developed a delayed, severe neutropenia and thrombocytopenia. A bone marrow biopsy confirmed a diagnosis of haemophagocytic lymphohistiocytosis secondary to the apical petrositis. The child was managed with conservative, supportive treatment. At four months follow up, he had completely recovered.

Histology

The bone marrow trephine was hypocellular for age, but all three haemopoietic cell lines were represented. Reduced numbers of granulocytes and red cell precursors were observed. Megakaryocytes were normal in number. Immunostaining for cluster of differentiation 68 glycoprotein showed increased numbers of macrophages demonstrating haemophagocytosis.

Conclusion

To our knowledge, there are no previously published case reports of Gradenigos syndrome complicated by haemophagocytic lymphohistiocytosis. The consensus was that this condition was probably precipitated by viral infection. The Semon Club panel agreed that this case presented a uniquely challenging phenomenon which had been optimally managed by a multidisciplinary team.

Recurrent neck mass following complete excision of a benign lesion

J C Fleming, R Moorthy, J Weighill

From the Royal Sussex County Hospital, Brighton.

Background

Castleman's disease is a rare, benign, lymphoproliferative condition of unknown aetiology characterised by lymph node hyperplasia. The neck is the second most common site of presentation. Histological classification includes hyaline-vascular, plasma cell or mixed types, whereas clinical presentation divides the disease into localised or multicentric types. Fine needle aspiration cytology results are unreliable, and diagnosis requires a tissue biopsy. Surgery

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is the mainstay of treatment for localised disease, and complete excision was previously thought to be curative.

Case report

A 16-year-old female presented with an enlarging right neck swelling. Magnetic resonance imaging (MRI) revealed bulky, right-sided cervical lymphadenopathy. Surgical excision was performed. Histological analysis demonstrated a completely excised, encapsulated mass consistent with hyaline-vascular Castleman's disease. Six years later, the patient re-presented with a one-year history of a similar enlarging right neck mass.

Radiology

Repeated MRI scans showed several soft tissue masses consistent with pathological lymph nodes at levels II and III. Dr Connor noted a characteristic, low signal, stellate pattern on the T2-weighted scan.

Management

A repeat excision of the lesion was performed in September 2007. The patient had an uncomplicated post-operative course, with no clinical evidence of recurrence.

Histology

Histological analysis revealed the typical depleted follicles with surrounding 'onion skin' pattern confirming Castleman's disease of the hyaline-vascular type.

Conclusion

This case represents the first published report of recurrence of localised Castleman's disease (hyaline-vascular type) following complete excision. The importance of follow up to detect recurrence was emphasised, and interval ultrasound monitoring was recommended.

Exit procedure for intra-oral teratoma

V Teoh, A Donne, I Hore

From Evelina Children's Hospital, Guys' and St Thomas's NHS Foundation Trust, London.

Introduction

Teratomas are the commonest congenital neoplasms, and the head and neck are frequently involved. These midline lesions can occur within the mouth, resulting in extra-partum airway compromise.

Case report

A 27-year-old, primigravida woman was found to have a fetus with a mass arising from its mouth at her 13-week antenatal scan. Computed tomography and magnetic resonance imaging scans at 34 weeks' gestation demonstrated a large cleft palate and an obstructing lesion arising from the left alveolar margin. An elective caesarean section was undertaken at 37 weeks' gestation, during which the fetus was partially delivered and an endotracheal tube sheathed, rigid endoscope intubation was performed before the delivery was completed. The airway was then secured with a tracheostomy.

Result

Histological analysis confirmed the typical findings of a mature teratoma.

The baby survived, and was decannulated some weeks after complete excision of the teratoma.

Conclusion

Intubation with a sheathed endoscope is very safe and allows direct visualisation of the vocal folds from an angle not possible using standard laryngoscopic intubation techniques.

Rhinology session

Chairman: Mr David Howard

Unusual nasal swelling after trauma

S Gupta, R Talwar, S Paun

From St Bartholomew's and the Royal London Hospitals.

Giant cell tumour of soft tissue is a rare, low grade sarcoma with clinicopathological features similar to those of giant cell tumour of bone. No aetiological factors have been confirmed. There are rare reports of giant cell tumour of soft tissue involving various head and neck sites, including the nasal cavity and sinuses. We report the first case of post-traumatic giant cell tumour of soft tissue in this region.

A 42-year-old male trader presented with a painless nasal tip swelling, having sustained a boxing injury three months previously. Examination revealed a 1.5-cm, subdermal, cystic lesion in the nasal tip region, ballooning into the nasal vestibule. Needle aspiration was unsuccessful. Magnetic resonance imaging of the lesion demonstrated solid gadolinium enhancement extending across the dorsal cartilaginous septum. Histological correlation was advised.

An external septorhinoplasty approach enabled complete excision of a multilobulated, fibrotic mass lying between the medial crurae of the lower lateral cartilages. Nasal tip reconstruction was performed with dome sutures and cephalic trim.

Histological analysis confirmed a benign giant cell tumour of soft tissue, composed of polyhedral, spindle and giant cells with small amounts of intervening collagen.

Twelve-month follow up demonstrated excellent wound healing and no recurrence.

In conclusion, soft tissue tumours must be considered in the differential diagnosis of nasal swelling. Prof. Howard suggested that the lesion may represent a giant cell reparative granuloma, as it has a similar histopathological appearance; this was verified by Dr Sandison. This case provides evidence that trauma may be an aetiological factor in the development of giant cell tumour of soft tissue. With meticulous surgical excision, a benign clinical course can be expected.

Three-week epistaxis following chronic nasal obstruction in a 14-year-old boy

M J D Dunstan, A P Patel, A A Adeniran

From King's College London School of Medicine.

Case report

A 14-year-old boy presented with a three-week history of frequent, left-sided epistaxis, plus complete right nasal obstruction for 18 months. Examination under anaesthesia revealed a posterior right nasal mass extending through the posterior septum into the left nasal cavity. Computed tomography and magnetic resonance imaging (MRI) demonstrated a $39 \times 20 \times 22$ mm mass centred in the posterior right nasal cavity, with no apparent intracranial extension or invasion of the skull base. A diagnosis of stage II juvenile nasopharyngeal angiofibroma was made, although the typical finding of extension through the sphenopalatine foramen was not observed radiologically. Radiological embolisation and endoscopic resection of the mass were conducted. Histological analysis revealed a mixed tubular–cribriform adenoid cystic carcinoma.

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Discussion

A further examination under anaesthesia and debridement demonstrated no residual macroscopic disease, and external beam radiotherapy (60 Gy in 30 fractions) was administered. Follow-up MRI scans demonstrated no disease recurrence. Adenoid cystic carcinoma is extremely rare in children, and there is thus little literature regarding its treatment. Surgery and radiotherapy are the accepted treatment of adenoid cystic carcinoma in adults. Prof. Howard commented that several typical radiological features of juvenile nasopharyngeal angiofibroma were missing in this case. These included extension into the pterygopalatine fossa, bowing of the maxillary antrum and erosion of the basisphenoid. Rare malignancies of the nasal cavity should be considered in adolescents, particularly when typical radiological features are absent.

Expectoration of a soft tissue mass in an otherwise asymptomatic woman

A A Adeniran, A P Patel, M J D Dunstan

From King's College London School of Medicine.

Case report

A 40-year-old woman presented after expectorating a bloody tissue mass, which she took to her general practitioner. Histological examination showed submucosal aggregates of plasma cells. Immunohistochemical analysis revealed polyclonal immunoglobulins. A reactive cause was suspected. Computed tomography (CT) revealed a 1.4-cm, enhancing mass related to the lateral wall of the left nasopharynx. The patient was diagnosed with extramedullary plasmacytoma, following excision biopsy and histological analysis. Bone marrow aspirate and trephine were normal. Post-operative positron emission tomography showed no local uptake or bone lesions. Radiotherapy was declined by the patient and a 'wait and see' approach was adopted, with the option of radical radiotherapy on local recurrence.

Discussion

Nine months post-operatively, the patient was asymptomatic with no clinical or radiological evidence of recurrence. A CT scan carried out nine months prior to admission to investigate a conductive hearing loss was available. On retrospective review of this scan, Dr Connor commented that the neoplasm was macroscopically similar in size, providing an insight into the natural history of extramedullary plasmacytomas. Professor Michaels confirmed the finding of plasma cells under stratified squamous epithelium, and the likelihood of extramedullary plasmacytoma, but emphasised the need for confirmatory immunohistochemistry. Prof. Howard and Professor Michaels discussed rates of progression to multiple myeloma; previous retrospective literature reviews have found conversion rates of 40 per cent (Wiltshaw, 1976) and 16 per cent (Alexiou *et al.*, 1999).

An unexpected diagnosis in a patient with bilateral nasal polyps

R Harris, D B Mitchell, M Black

From the William Harvey Hospital, Ashford.

Case report

A 27-year-old woman presented with recurrent, spontaneous, right-sided epistaxis. Symptoms of bilateral nasal congestion, anosmia and frontal headache persisted for three months, despite oral antibiotics and topical steroids in primary care. Bilateral nasal polyposis and left septal deviation were identified on examination. Septoplasty and bilateral functional endoscopic sinus surgery were arranged; however, these were abandoned upon finding a friable, polypoidal mass in both nostrils, which bled extensively on touch intra-operatively.

Radiology

Computed tomography (CT) scanning of the paranasal sinuses demonstrated a large, polypoidal mass completely opacifying the left maxillary, ethmoid and sphenoid sinuses. The mass traversed the nasal septum to partly opacify the right maxillary antrum, with evidence of bony remodelling. No metastatic lesions were identified.

Histology

Histological analysis revealed scanty, nasal respiratory type mucosa and multinodular masses of neoplastic cartilage with focal calcification. Grade two chondrosarcoma was diagnosed.

Management

The patient underwent craniofacial resection and enblock left orbital exenteration with subsequent orbital reconstruction.

Discussion and lessons learnt

This case emphasises the importance of reviewing CT images personally and of maintaining a high index of clinical suspicion when managing recurrent epistaxis with nasal polyposis, irrespective of age. Prof. Howard suggested that the differential diagnosis of tissue calcification might include inverted papilloma or fungal infection. He felt that enucleation of the orbit would not prolong the patient's life, and proposed that future treatment options include proton therapy.

A lesion on the right nasal ala: part of a rare syndrome

J Watson, M Karela, M Terry

From the Princess Royal University Hospital, Orpington.

Background

Ferguson–Smith syndrome, or multiple self-healing squamous epitheliomata, is a rare cancer-associated genodermatosis transmitted via autosomal dominant inheritance. Most cases are recorded in patients originating from West Scotland, and the gene is located on chromosome 9q22. The disease occurs on exposed areas of skin, causing lesions that spontaneously regress leaving deep, pitting scars.

Case report

A 79-year-old woman presented with a 3 mm lesion on the right nasal ala extending into the nasal mucosa. Biopsies were taken.

The patient had been diagnosed with multiple self-healing squamous epitheliomata aged 16 years, and had had multiple lesions excised over the years, the majority from her face. Her mother and two of her five siblings had the same condition.

Histology

Professor Michaels felt that the lesion showed downgrowth of well differentiated stratified squamous epithelium, which could represent a keratocanthoma or a well differentiated, very low grade squamous cell carcinoma (SCC).

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Management

The right nasal ala lesion was excised, and a partial thickness skin graft from the right post-auricular region was applied. Mr Paun noted that a partial thickness graft in this area often leaves a notch around the alar rim as it heals, and recommended that a composite with cartilage would have been more appropriate.

Conclusion

Multiple self-healing squamous epitheliomata is a rare syndrome causing lesions over the head and neck which closely resemble SCCs, and which cause severe scarring if left untreated. Therefore, the appropriate treatment is local excision.

Head and neck session

Chairman: Mr Ricard Simo

Neck mass as initial presentation of testicular seminoma

A Jacob, A Riskalla

From University Hospital Lewisham, London.

Case report

A 36-year-old man presented with a three-month history of general malaise, poor appetite, fever, rigor and night sweats. He also reported a one-month history of a left-sided neck mass and left abdominal pain. Examination revealed a large, firm, supraclavicular mass. Computed tomography scanning revealed a 5×5 cm mass in the left side of the neck, extending into the superior mediastinum and displacing the airway.

A full blood count analysis revealed neutrophilia. There were also abnormal liver function tests and a markedly raised c-reactive protein level. An incisional biopsy of the mass confirmed metastasis of a testicular seminoma. The patient was referred to the urology department for further treatment of the testicular seminoma.

Discussion

The early diagnosis of a neck metastasis from a testicular seminoma will depend on a high index of clinical suspicion. Prof. Howard and Mr Simo held the opinion that a thorough history, examination, and possibly further imaging of the abdomen, pelvis and groin, should be routinely performed in order to detect such a rare cause.

Rare neoplasm of the tonsil in pregnancy

J Magill, M Ferguson, W Grant

From the Charing Cross Hospital, London.

Introduction

We present the case of a 29-year-old, non-smoking, pregnant woman who had a persistent right tonsillar lesion which was excised. Histological examination demonstrated an inflammatory myofibroblastic tumour.

Case report

The patient presented with a 10-day history of odynophagia despite a course of antibiotics. Intensification of antimicrobial therapy had no effect on her symptoms. Examination revealed a right, grade II, erythematous tonsil with ulceration on the upper pole. A biopsy was arranged, and initial evaluation was suggestive of spindle cell carcinoma. However, this diagnosis was reviewed after a panel of immunohistochemical stains confirmed inflammatory myofibroblastic tumour (a benign pseudosarcoma).

Histology

Macroscopically, an ulcerated, polypoid tumour was present. On histological analysis, the tumour consisted of atypical, large spindle cells mixed with inflammatory cells. The tumour expressed smooth muscle actin, vimentin and cyclin D1. There was no expression of cytokeratin.

Management

Carbon dioxide laser excision of the right tonsil was performed with clear margins. Considering the risk of local recurrence, careful follow up was arranged.

Conclusion

Pseudosarcoma of the inflammatory myofibroblastic type rarely occurs in the tonsil. We present the first reported case appearing during pregnancy. This entity is known to be locally aggressive, both clinically and pathologically, and can present with similar features to high grade carcinoma of the tonsil. We recommend that first line treatment of this condition consist of complete local excision. This case highlights the need for expert review of these unusual cases within a specialist, multidisciplinary setting.

Involuting pleomorphic adenoma of the submandibular salivary gland

R Rolph, B Conn, J-P Jeannon

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

Case report

A 41-year-old, Caucasian British man presented with a right submandibular swelling. The patient's history was suggestive of calculus disease. Clinical examination revealed a firm, right-sided, 2-cm, submandibular gland swelling. The rest of the ENT examination was unremarkable.

Ultrasonic imaging revealed a 10-mm, hypo-echoic region within the right submandibular gland. Fine needle aspiration cytology was non-diagnostic, and an excision biopsy was performed for diagnostic and therapeutic reasons.

Histological analysis revealed a well circumscribed tumour within the gland, in which the epithelial elements were obscured by a heavy lymphocytic infiltrate. The presence of small, cystic spaces imparted a microcystic appearance, and there were focal areas of stromal scarring among myxochondroid matrix. In places, there were groups of large cells with pleomorphic, hyperchromatic nuclei. Immunohistochemical analysis revealed epithelial ductal structures and myoepithelial cells with strong positivity for calponin. The tumour was subsequently diagnosed as an involuting pleomorphic adenoma, on the basis that there appeared to be active destruction of the neoplastic elements by the lymphoid infiltrate.

Discussion

Interpretation of the unusual histological features proved difficult, and the initial report proffered a differential diagnosis including benign and malignant entities: acinic cell carcinoma, carcinoma ex-pleomorphic adenoma or a variant of pleomorphic adenoma.

To our knowledge, there have been no previous reports of pleomorphic adenoma with these distinctive features. Involuting pleomorphic adenoma should be considered in the differential diagnosis of epithelial salivary gland tumours with an associated heavy lymphoid infiltrate.

It is of paramount importance to differentiate this condition from a malignant pathology, in order to initiate appropriate management and follow up.

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A rare cause of retrosternal goitre

H Wong, J R Tysome, R Simo

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

Introduction

Metastases of solid tumours to the thyroid gland are uncommon, with renal cell carcinoma being the most frequently reported.

Case report

An 82-year-old woman presented with a 12-month history of dysphonia, midline neck swelling and progressive stridor. She had undergone a radical right nephrectomy for renal cell carcinoma 20 years previously. On examination, she had a large goitre and was clinically euthyroid. Flexible nasendoscopy revealed compression of the posterior tracheal wall.

Radiology

Computed tomography revealed a multinodular goitre with retrosternal extension and tracheal compression.

Histology

The results of ultrasound-guided fine needle aspiration cytology were suspicious of metastatic renal clear cell carcinoma.

Management

The patient underwent incisional biopsy, which confirmed the diagnosis. Thyroidectomy was not advised in view of the diagnosis, due to medical comorbidity. Following diagnosis, the patient was referred to the renal oncology team, which advised palliative management.

Conclusion and lessons learned

Renal cell carcinoma metastases to the thyroid gland may often have a delayed presentation. This diagnosis should be considered in any patient with a history of renal cell carcinoma who presents with a thyroid mass. Prof. Howard commented that important differential diagnoses included anaplastic thyroid cancer and other sources of metastasis. Professor Michaels confirmed the histopathological diagnosis. Thyroidectomy can be performed for local control in selected patients, although the survival of these patients depends more on their comorbidity than on tumour-related factors.

Advanced, metastatic thyroid cancer presenting as hyperthyroidism, goitre and spinal cord compression

W Y Lee, J-P Jeannon, M O'Connel

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

Introduction

Hyperthyroidism due to poorly differentiated cancer of the thyroid is rare. The complications from distant metastatic disease and the management of tri-iodothyronine thyrotoxic storm are discussed.

Case report

A 66-year-old Nigerian woman presented with a large goitre, lower back pain and weakness of the left leg. On examination, she had a 10-cm thyroid mass and a 5-cm sternal mass. Biopsy and histological analysis revealed follicular thyroid carcinoma. Magnetic resonance imaging of the neck showed masses in the head of the clavicle, sternum, mandible and multiple level cervical lymph nodes. Positron emission tomography showed bony metastases in the left femoral head, lungs and pelvis. A sacral mass was causing cauda equina compression.

Urgent radiotherapy and steroids were given to reduce the cauda equina compression, and medical therapy for thyrotoxicosis was commenced. Thyroidectomy with bilateral modified radical neck dissection was performed to alleviate tracheal compression. Radical dose external beam radiotherapy was given to the neck and the bony deposits.

Conclusion

We could find only one previous report of poorly differentiated cancer of the thyroid presenting with thyrotoxic storm. Treatment of such extensive metastatic disease can be difficult, and a multidisciplinary approach is needed to accurately stage and treat such advanced cases.

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