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Proceedings of the 127th Semon Club, Otolaryngology Department, Guy's and St Thomas' NHS Foundation Trust, London, UK, 28 May 2004

Chairman: Ms E B Chevretton, Consultant Otolaryngologist, Guy's and St Thomas' Hospital, London
Pathologists: Professor L Michaels, University College London, and Dr A Sandison, Charing Cross Hospital, London
Radiologist: Dr G Rottenburg, Guy's and St Thomas' Hospital, London
Minutes: Mr P S Phillips, Specialist Registrar in Otolaryngology, Guy's and St Thomas' Hospital, London

Unilateral sphenoiditis causing bilateral papilloedema

F C Van Wyk, J A McGilligan
From the Royal Sussex County Hospital, Brighton, UK

Case report

A 70-year-old woman presented with a two-week history of diplopia, frontal headache and blurred vision. Bilateral papilloedema was noted. Computed tomography revealed right-sided opacification of the sphenoid and posterior ethmoid sinuses. Decompression of a right-sided sphenoid pyocoele was followed by complete resolution of symptoms. No mucosal abnormality was evident at surgery. Unexpectedly, the histology revealed adenocarcinoma. Immunohistochemical analysis suggested an unusual primary or a secondary metastasis from breast or the gastrointestinal tract. The patient denied exposure to wood dust or leather. Further investigation revealed lower abdominal pain, a colonic polyp, and a family history of colonic cancer. Raised carcino-embryonic antigen levels inferred visceral malignancy. The patient developed a right lateral sinus thrombosis and, despite intensive medical treatment, died eight weeks later. A post mortem autopsy demonstrated a posterior fossa metastasis but no other primary.

Discussion

Dr Sandison commented that this was a very unusual tumour, although a lower gastrointestinal tract primary could be excluded on the basis of the stains obtained. Ms Chevretton commented that once adenocarcinomas metastasise, patients do deteriorate over weeks rather than months. The lateral sinus thrombosis was thought to be due to the prothrombotic effect of cancer.

A rare cause of lump in the tongue

M Craig, M J Gleeson, R Simo
From Guy's and St Thomas' Hospital and Lewisham Hospital, London, UK

Case report

A 16-year-old boy of Pakistani origin presented with a three-year history of a firm, painless mass in the anterior tongue in the midline. The mass was within the tongue muscle, extending to the floor of mouth, with no epithelial changes and no lymphadenopathy. Contrast magnetic resonance imaging showed a highly vascular mass, and incisional biopsy indicated a glomus tumour. Transmandibular excision of the encapsulated mass was performed; the histopathology, confirmed by Professor Michaels, showed an immature glomus tumour, approaching the excision margins, with potential for recurrence and maturation.

Discussion

Glomus tumours are rare lesions of the head and neck. They are benign hamartomas, originating from the pericytic cells of the neuromyoarterial bodies (which are involved in temperature regulation in the skin by the shunting of blood). They should not be confused with paragangliomas, which are informally called glomus tumours and are more commonly seen in otolaryngology. Glomus tumours usually present to hand or orthopaedic surgeons as small and extremely painful lumps in the digits or limbs, occasionally being multiple or familial.

Presumed angiofibroma of the inferior turbinate

R Kumar, S Lo, J Blanshard
From the North Hampshire Hospital, Basingstoke, and St George's Hospital, London, UK

Case report

A 29-year-old man presented with a six-week history of worsening, left-sided nasal obstruction, with blood-stained nasal discharge. Anterior rhinoscopy revealed a soft, fleshy mass filling the left nostril, with crusting over its anterior surface. Rigid nasal endoscopy indicated that the soft tissue swelling was arising from the inferior turbinate. The remainder of the ENT examination was normal. A provisional diagnosis of pyogenic granuloma was made. The patient underwent excision of the mass. Intra-operatively, a lobulated mass was found arising from the anterior end of the inferior turbinate. The mass was removed using monopolar diathermy. Recovery was uncomplicated. Histological analysis revealed a proliferation of variable-sized vascular spaces surrounded by a stroma of varying cellularity, lacking a lobular pattern. The histological features were consistent with an angiofibroma.

Discussion

The inferior turbinate is a very unusual site of origin for an angiofibroma, with only two previously reported cases in the medical literature. Professor Michaels reviewed the histology slides and, considering the clinical features, held the opinion that the histological appearance favoured a diagnosis of pyogenic granuloma. He emphasised that an unusual histology report that does not match the clinical features should be carefully reviewed. A second opinion may be appropriate in order to gain an accurate diagnosis.

Angiosarcoma of maxillary sinus

A K Golagani, M H Wickham, R E Quincy
From the Barnsley District General Hospital, Barnsley, UK

Case report

A 90-year-old man presented with epistaxis from his left nasal cavity and numbness over the left side of his face of four weeks' duration. Clinical examination of the left nasal cavity was unremarkable. A computed tomography (CT) scan revealed a large, homogenous opacity filling the left maxillary antrum, and eroding its medial and lateral walls and the floor of the left orbit. A biopsy was reported as low-grade angiosarcoma, further confirmed by immunohistochemistry (being positive for CD31, CD34 and factor eight antigen). As the patient was not fit for surgery, radiotherapy was advised. However, unfortunately, three months after the diagnosis, the patient died of an unrelated cause.

Discussion

Only 19 cases of angiosarcoma of the maxilla have been reported in the literature. Dr Sandison discussed the possibility of an organised haematoma eroding the bony walls of the maxilla, but confirmed the diagnosis based on histopathology and immunohistochemistry as a low-grade maxillary angiosarcoma. Dr Rottenberg further supported the diagnosis based on the CT findings. Correlating clinical, radiological, histopathological and immunohistochemistry findings, the diagnosis was confirmed as maxillary angiosarcoma.

Parotid and submandibular masses associated with cervical lymphadenopathy: malignant or benign?

V Prasad, R Persaud, D Howard
From the Royal National Throat Nose & Ear Hospital, London, UK

Case report

A 58-year-old Chinese man presented with a seven-year history of prominent soft tissue swellings in the head and neck region. On examination, a soft, 3×2.5 cm mass was palpable in the tail of the right parotid gland, causing displacement of the ear. There was also a 2×1.5 cm lymph node in the right jugulo-digastric area and another soft mass (4×2 cm) superficial to the left masseter muscle. The chest X-ray appeared normal. Previous blood results revealed eosinophilia and elevated immunoglobulin E levels. The patient was diagnosed with Kimura's disease.

Discussion

Professor Michaels confirmed the diagnosis, due to the presence of inflammatory granulomas with a large amount of lymphocytes and eosinophilic infiltration. Kimura's disease is a rare, idiopathic condition that usually affects people of Oriental origin, especially young men. The condition is characterised by painless, solitary or multiple soft tissue swellings in the major salivary glands, subcutaneous tissue and lymph nodes in the head and neck region. Kimura's disease, which usually presents bilaterally, is often misdiagnosed as a salivary gland tumour with associated regional lymphadenopathy. Histological confirmation is needed from an incisional or excisional biopsy.

Excised nasopharyngeal tumour recurring as an infra-temporal metastasis

L Peiris, D Jiang, D Mitchell
From the William Harvey Hospital, Ashford, UK

Case report

We report the case of a 61-year-old man who presented with a one-year history of nasal congestion and discharge. Examination of the postnasal space revealed a large, fibrous lesion occupying the superior nasopharynx. Biopsy of this tumour showed characteristic physalipherous cells, compatible with a diagnosis of chordoma. Access was gained to the nasopharynx via a left maxillary swing approach. The tumour was based on the left basisphenoid, extending inferiorly and prolapsing through the posterior choanae into the nasal cavity. The patient received post-operative radiotherapy. Eighteen months after resection, the patient re-presented with a mass in the left temporal region. A magnetic resonance imaging scan revealed a large mass in the left infratemporal fossa extending into, and destroying, the lateral wall of the orbit. There was no evidence of recurrence in the nasopharynx. Biopsy of this mass was histologically similar to the initial biopsy and excision specimens. At the time of writing, the patient was awaiting ophthalmic assessment with a view to excision of this recurrence.

Discussion

The point was made that the recurrence may have arisen due to peri-operative seeding – a known complication of chordoma excision. The benefit of wide access enabled by a maxillary swing approach should be weighed against the consequences of seeding to adjacent structures.

Amyloidosis of the larynx

H Narayanareddy, S T Baer
From the Conquest Hospital, St Leonard's on Sea, UK

Case report

A 58-year-old woman presented with an 18-month history of voice change. Her voice was worse on using the telephone and returned to normal occasionally. She was a non-smoker. Examination, including nasendoscopy, showed her left vocal fold mobility to be slightly impaired. Investigations, including a full blood count, electrolytes, thyroid function tests and computed tomography scan of the chest, were normal. Initially, the patient was given speech therapy, with no improvement in her symptoms. She was examined again, and a swelling was noted below the free edge of both vocal folds; the mobility of the folds was found to be normal. She underwent microlaryngoscopy, biopsy and laser ablation of the swelling. Histopathology revealed amyloidosis.

Discussion

Amyloidosis of the larynx is rare; however, the larynx is the commonest site of amyloidosis in the head and neck region. Laryngeal amyloidosis can be local or, rarely, part of systemic amyloidosis. The commonest presentation is change of voice and the commonest site is the vocal folds. Diagnosis is clinched by histopathology. Treatment options are surgery and laser. Recurrence is common. In patients with voice change and vocal fold swelling, it is important to keep in mind that amyloidosis of the larynx can present with vague symptoms; it can be ruled out by biopsy and histopathology.

A rare case of bilateral nasal blockage

F I Abdulkader, A Alshammari, H S Kaddour
From the Harold Wood Hospital, London, UK

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Case report

A 63-year-old woman presented with a three-month history of bilateral nasal blockage, right eye proptosis and diminished right eye vision. Examination of the nose revealed a large mass occupying the right nasal cavity and extending into the postnasal space, causing obstruction to the left side. Computed tomography and magnetic resonance imaging scans confirmed a large mass sitting on the ethmoid air cells, extending to the nasopharynx and oropharynx, and compressing the right optic nerve. The histological appearance was described as a malignant myxoid tumour; a 'second opinion' described it as a spindle cell sarcoma. The management was discussed in a multidisciplinary team meeting. The patient declined the surgical option and was treated with chemotherapy and radiotherapy.

Discussion

The challenges of this case were discussed: late presentation, extensive disease and the role of a multidisciplinary approach to treatment. The histological appearance of the tumour was also considered, and Professor Michaels agreed that the findings were difficult to interpret and that spindle cell sarcoma would be the best description. To the authors' knowledge, this is the first reported case of such pathology being found in the paranasal sinuses.

An unusual pharyngeal mass extending to the middle ear

R J D Hewitt, S A Hannan, G Alusi
From St Bartholomew's & The Royal London Hospitals, London, UK

Case report

An 18-year-old female student presented with a two-week history of right otorrhoea, fever and sore throat. Ear examination revealed a dull, inflamed right tympanic membrane with a 20 dB conductive loss on audiogram and a type B tympanogram. Examination of the oropharynx revealed a bilobar mass emerging from the nasopharynx and descending into the laryngopharynx. The patient reported a long-standing nasal voice but had never noticed the swelling. Computed tomography (CT) scanning showed a posterior pharyngeal mass extending from the nasopharynx to the hypopharynx. Surgical excision revealed a pedunculated globular/cystic mass, which was removed at its stalk, medial to the right eustachian tube. Aspirated clear fluid grew no organisms. A right tympanotomy revealed 'glue ear', and a grommet was inserted. Post-operative magnetic resonance (MR) imaging and temporal bone CT imaging were sought.

Discussion

Professor Michaels remarked on the respiratory epithelial lined structure containing lymphoid stroma, and proposed that it was most likely to be a first branchial cleft cyst, but could also be a Torwaldt's cyst. Dr Rottenberg noted soft tissue extension through the inferior margins of the temporal bone, contiguous with soft tissue opacification in the right aditus antrum, mastoid air cells, and hypo-, meso- and epitympanum. Discussion with the forum concluded that this represented a branchial cleft anomaly compressing the eustachian tube from the medial aspect and causing its dysfunction. It was felt that the eustachian tube dysfunction would only be assessable when the current grommet became displaced. A plan to monitor this benign condition with clinical examination and repeated CT and MR imaging was recommended, due to the difficulty of surgical excision of the residual cyst.

An extensive parapharyngeal granular cell tumour: a unique surgical challenge

K Amonoo-Kuofi, R Persaud, L Michaels
From the Royal National Throat, Nose and Ear Hospital, London, UK

Case report

A 49-year-old woman presented with a 12-month history of mild dysphagia. There were no other aero-digestive symptoms of note. Oral examination revealed a swelling in the lateral pharyngeal wall, displacing the tonsil medially. Cranial examination was unremarkable. Palpation revealed a firm, non-tender parapharyngeal mass. There was no cervical lymphadenopathy. The rest of the ENT examination, including flexible endoscopy, was normal. A computed tomography scan (neck and chest) showed a 3.75×2 cm, non-enhancing, homogenous mass arising in the parapharyngeal space, extending from the nasopharynx to the oropharynx. No clear plane distinguished the tumour from the prevertebral musculature, and there was also close adherence to the right carotid sheath and contents. An intra-oral biopsy confirmed a diagnosis of significant oro-naso-parapharyngeal granular cell tumour. Excision of the tumour by conventional 'cold steel' technique proved impossible, due to the gristle-like consistency of the tumour. The tumour was subsequently resected via a trans-oral route using a CO_2 laser.

Discussion

Granular cell tumours are rare, benign tumours of uncertain origin and have previously been reported in the head and neck region (the tongue is the most commonly affected site). There is a sexual preponderance in females, with a higher incidence in people of African origin. Granular cell tumours presenting as a parapharyngeal mass have not been previously reported. Although it has been reported that seven per cent of these tumours may be malignant, Professor Michaels held the view that this was due to misinterpretation of pseudoepitheliomatous hyperplasia (i.e. granular cell tumours in submucosal locations with hyperplastic epithelia) as well differentiated squamous cell carcinoma.

Accidental contamination of traumatic tracheostomy by extraneous neoplastic tissue

A A Khattab, S K Duvvi, A Ali
From the Southmead General Hospital, Bristol, UK

Case report

Contamination of extraneous neoplastic tissue in the neck is unusual, but always misleading and potentially detrimental to the patient's health. We present a case of accidental contamination with squamous cell carcinoma in a patient with a tracheostomy, following road traffic accident neck trauma. The objective is to highlight the confusion and potential surgical risks of tissue contamination. Histology showed a fragment of markedly atypical cells, suggestive of an epithelial neoplasm. However, the fragment was very poorly preserved and showed air-drying artefact. Clinical, endoscopic and deoxyribonucleic acid testing proved that the tissue was unrelated to the patient.

Discussion

This is the first reported case of tracheostomy contamination with neoplastic tissue. Contamination of pathology samples must be investigated thoroughly, and policies need to be reviewed in order to prevent misdiagnosis as a result.

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Lemierre's syndrome secondary to paranasal sinus pathology: a real pain in the neck!

T Maani, I Friedrichs, A Toma
From St George's Hospital, London, UK.

Case report

A 14-year-old girl with a known history of chronic blockage of the nose was admitted to the paediatric department with nausea, vomiting and neck pain. Blood cultures grew *Fusobacterium necrophorum*, and computed tomography (CT) scanning showed right sphenoid sinusitis and cavernous sinus thrombosis. Broad-spectrum antibiotics were initiated and the patient was transferred to the ENT services with a diagnosis of Lemierre's syndrome, in order to treat the underlying sinus pathology. Lemierre's syndrome is known to be a complicated parapharyngeal and retropharyngeal infection, with internal jugular vein and venous sinus thrombophlebitis. Repeated CT and magnetic resonance imaging scanning confirmed the underlying pathology. Endoscopic sphenoid sinus exploration revealed only oedematous sphenoid sinus mucosa. The patient improved post-operatively and was discharged on high-dose antibiotics and warfarin. Lemierre's syndrome can be an extreme complication of a neglected sore throat. It starts with a pain in the neck and if left without prompt treatment, can become a real pain in the clinicians' neck!

Discussion

Dr Rottenberg suggested that the status of the internal jugular vein should be checked with a neck ultrasound. Ms Chevretton suggested that the duration of anticoagulation and antibiotic therapy should be discussed in a multidisciplinary team setting, with paediatric and microbiology input.

A case of a right external nasal mass

V Singh, M Farag
From the Barnet and Chase Farm Hospital, London, UK

Case report

A three-year-old girl presented with a 2×2.5 cm, soft, non-tender, non-pulsatile, cystic mass on the right lateral aspect of the nose, beneath the medial canthus. Anterior rhinoscopy ruled out intranasal extension of the mass. Radiological examination showed no bony erosion. The lesion was excised via an intranasal approach in order to avoid an external scar. Histopathological examination revealed a spindle cell lesion, possibly a low-grade leiomyosarcoma. The patient developed a smaller, local recurrence five months post-operatively. At the time of writing, the child was under the care of the paediatric oncologists.

Discussion

Dr Sandison emphasised that this is indeed a very worrying spindle cell tumour, characterised by its aggressive invasion of surrounding structures and tendency to recur. Professor Gleeson added that equally aggressive surgical management should be adopted at an early stage. Radiotherapy has no place in the management of these soft tissue tumours.

Neurofibromatosis presenting as post-traumatic deformity of the pinna

C Hopkins, S Khemani, K Uplands, D Golding-Wood
From the Princess Royal University Hospital, Farnborough, UK

Case report

We report the case of a plexiform neurofibroma of the pinna in a 15-year-old boy. The presenting history was of trauma sustained when the patient was only 18 months old, with the resultant abnormality of the left pinna and external auditory canal leading to recurrent otitis externa and a conductive hearing loss. Examination findings were thought to be consistent with a post-traumatic, organised haematoma. The pinna had continued to develop abnormally, with rotation and gross secondary lymphoedema. A histological sample demonstrated the lesion to be a plexiform neurofibroma. This lesion is pathognomic for neurofibromatosis type one, although there were no associated features in this case. Three months post-surgery, the cosmetic appearance of the pinna had improved, but there was evidence of recurrent disease at the resection margins.

Discussion

Histological analysis showed myxoid-like tissue with spindle cells in an anastomosing mass of tissue, sometimes arising from nerve bundles. The forum discussed the best setting for management of these patients, and emphasised the need to examine the eyes on a yearly basis, as well as the need for further scanning to check for other lesions. (Following discussion of this case at the Semon Club, the patient was referred to an ophthalmologist, and also to a specialist neurofibromatosis clinic for follow up.)

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