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Proceedings of the 144th Semon Club, 1 November 2012, ENT Department, Guy's and St Thomas' NHS Foundation Trust, London, UK

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

Secretaries: Mr Sherif Haikel, and Miss Simone Hadjisymeou, Guy's and St Thomas' NHS Foundation Trust

Invited panel for pathology: Professor Leslie Michaels, and Dr Ann Sandison, University College London

Invited panel for radiology: Dr Steve Connor, and Dr Ata Siddiqui, Guy's and St Thomas' NHS Foundation Trust

Professor Leslie Michaels awarded a prize for the best presentation of the meeting to Mr Adebayo Alli for 'Recurrent tracheoesophageal fistula following laryngeal cleft repair in a female infant'.

Otology section

Chairman: Mr Alec Fitzgerald O'Connor

An unusual cause of parotid pain and swelling

E Head, L McClymont

From Raigmore Hospital, Inverness

A 75-year-old man presented with a left external jugular vein thrombosis secondary to a left parapharyngeal mass. Unfortunately, two fine needle aspirates were inconclusive (both producing a blood-filled syringe). Magnetic resonance imaging showed an unusual lesion in the left parapharyngeal space with irregular peripheral enhancement and signs suggestive of haemorrhage. The appearances were not diagnostic, and a vascular, inflammatory or neoplastic lesion was considered possible. Open biopsy was felt to be potentially hazardous (due to the risk of bleeding); we therefore decided to closely monitor the lesion.

In the meantime, the patient was diagnosed with a lung mass, suspicious of malignancy. Open biopsy of the parapharyngeal lesion, now felt to be necessary, showed a high-grade spindle cell sarcoma. Positron emission tomography (PET) scanning showed a positive lesion in the pharynx and two lung lesions consistent with metastases. The tumour was widely excised but unfortunately several margins were involved with tumour. After deliberation, the patient elected for palliative treatment.

At the 144th Semon Club meeting, Dr Steve Connor felt that the tumour was more likely to be a metastasis, as primary tumours of the parapharyngeal space are very rare. He also suggested that a computed tomography guided biopsy could have been useful to obtain an earlier histological diagnosis. Mr Grant Radcliffe (from the Royal Free Hospital) suggested that an earlier PET scan may have also assisted with management.

External jugular vein thrombosis is an unusual presentation of a parapharyngeal neoplasm. Additionally, spindle cell carcinomas of unknown origin are particularly rare in the head and neck. Early histological diagnosis (if possible) and appropriate imaging are essential aids in the management of these difficult cases.

A conchal bowl lesion in a 17-year-old male, with unusual histology

F Ryba, T Odutoye, J Williams

From St George's Hospital, London

Introduction

Seborrhoeic warts are a common non-malignant lesion of the ear in the older population, but are rare in young adults. This case highlights the importance of sufficient biopsy size, and of considering all diagnoses irrespective of the patient's age.

Case report

A 17-year-old male student with no significant family history presented with a warty lesion in his right conchal bowl, on return from a trip to India. The lesion had been present and increasing in size for six months.

Histopathology

A 2 mm, incisional biopsy was reported as a benign, mildly inflamed seborrhoeic wart.

A 15×6 mm, excisional biopsy demonstrated a pT_1, moderately differentiated squamous cell carcinoma without perineural, lymphovascular or cartilage invasion. The specimen excision margins were positive.

A subsequent resection, performed with intra-operative frozen section analysis, produced tumour-free margins.

Radiology

Magnetic resonance imaging of the head and neck showed no cervical lymphadenopathy.

Management

Following the recommendation of the multidisciplinary team, the conchal bowl of the right ear was fully excised and the defect reconstructed with a Wolfe graft. Mr Grant Radcliffe commented that he would have also excised cartilage.

Conclusion

Seborrhoeic keratoses are rare in teenagers. This case highlights the fact that, when faced with an incongruous diagnosis, an alternative diagnosis should always be considered, and that a representative biopsy is essential in the diagnosis of any pathology, especially unusual lesions. Professor Leslie Michaels reviewed the initial histology, and felt that

spindle cell hyperplasia could be seen. He pointed out that although this was not diagnostic of malignancy, any ear lesion that shows atypia should be treated with a high degree of suspicion. He suggested a basal cell papilloma as an alternative diagnostic possibility for the first biopsy. Mr Alec Fitzgerald O'Connor asked if sunlight may have played a part in the aetiology, and enquired about the risk of metastasis or recurrence. Professor Leslie Michaels responded that exposure to sunlight may have played a part, but that metastasis and recurrence were unlikely.

An atypical lesion of the outer ear canal

D Subramaniam, S Baer

From East Sussex Healthcare NHS Trust

Introduction

A variety of cutaneous lesions and dermatological conditions can affect the outer ear. We present the case of an unusual lesion affecting a patient's ear canal.

Case report

A 57-year-old man was referred with watery discharge and bleeding from his left ear. He had been hit by a tennis ball two months previously. He denied any otalgia or hearing loss.

His medical history was significant for a tympanic membrane perforation that had closed spontaneously, as well as a bilateral pinnaplasty as a child.

On examination, a granulomatous lesion was noted on the posterior aspect of the left outer ear canal. This was removed under local anaesthetic.

Three years later, significant regrowth of the lesion occurred, and the patient now reported reduced hearing in his left ear.

Histological findings

The lesion was described as polypoid tissue comprising ulcerated, keratinising stratified squamous epithelium. Occasional foamy macrophages and giant cells were present. Staining for cluster of differentiation 68 protein was strongly positive. A diagnosis of non-X histiocytosis was made at St John's Institute of Dermatology in London.

Management

The lesion was removed under general anaesthetic via an endaural incision.

Learning points

Professor Leslie Michaels emphasised that, although there were histological features consistent with the diagnosis, the cluster of differentiation 68 protein stain was not specific for non-X histiocytosis and could also be consistent with a granuloma. Dr Ann Sandison pointed out that cutaneous histiocytosis may herald systemic histiocytosis, and that genetic counselling was required.

To scan or not to scan: an unusual case of tinnitus

P Touska, C J Skilbeck, P M Clarke

From the Charing Cross Hospital, London

Introduction

Tinnitus represents a common otological complaint with a wide differential diagnosis. It is often associated with sensorineural hearing loss, and infrequently with life-threatening or malignant processes. However, on occasion, decisions surrounding imaging may be difficult.

Case report

A middle-aged man was referred with persistent, bilateral tinnitus, slightly worse on the left side. Otological and cranial nerve examinations were unremarkable. However, pure tone audiometry revealed a mild hearing loss of 25–30 dB at 6 and 8 kHz. Due to the laterality of the audiological findings, a magnetic resonance imaging (MRI) scan was arranged.

Radiological findings

Magnetic resonance imaging of the cerebellopontine angles revealed a small, lobulated, cystic lesion in the pre-pontine region abutting and slightly eroding the clivus and displacing the basilar artery. An atypical cystic chordoma was the suggested diagnosis. However, subsequent expert review confirmed the diagnosis of a retroclival ecchordosis physaliphora.

Management

Following discussion of the patient's case in a multidisciplinary meeting, a conservative approach was taken, with annual MRI scanning.

Discussion

Dr Steve Connor highlighted the relative rarity of the finding, and noted that it is primarily a radiological diagnosis. Mr Alec Fitzgerald O'Connor led the discussion, which raised the point that some of the consultants at the meeting would not have requested imaging for this patient, given his age, the mild degree of audiometric asymmetry and the lack of any other symptoms.

Conclusion

Retroclival ecchordosis physaliphora represents a remnant of the fetal notochord and is predominantly asymptomatic and slow-growing. It should be considered if radiological reports suggest the presence of a small clivus chordoma.

A unique neoplastic lesion of the external ear canal

R Wong, B Fu, S Lew-Gor

From Brighton and Sussex University Hospitals NHS Trust

Introduction

Ceruminous gland tumours are rare neoplasms of the external ear canal which usually present in a benign manner. We present a classic case of this unique neoplastic entity.

Case report

A 49-year-old woman presented to our clinic with a soft tissue mass obstructing over 90 per cent of her left ear canal and causing a conductive hearing loss. She had no other otological symptoms.

Radiology

Computed tomography showed non-specific opacification of the left external auditory meatus without any bony destruction or aggressive features. There were retained secretions in the left petromastoid air cells.

Histology

The polypoid tumour was confined to the dermis and displayed lobar architecture. It had glandular structures lined with apocrine glands with small amounts of secretions.

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Despite the very small number of myoepithelial cells seen, histology was consistent with an adenoma of a ceruminous gland.

Management

Complete excision of the lesion was performed. The patient recovered fully, and at the time of writing was under outpatient monitoring.

Discussion

Professor Leslie Michaels and Dr Ann Sandison agreed that the histological appearances were unusual for ceruminous adenoma, and felt that the diagnosis of neuroendocrine adenoma should be excluded as this tumour can also arise from the middle ear or mastoids. Dr Steve Connor stated that radiological appearances favoured a lesion originating from the external auditory canal.

Conclusion

Neoplasms of the ceruminous glands are rare, and their benign behaviour allows many to go undetected. However, large intra- and extra-cranial lesions have been described which posed a surgical challenge. Early clinical suspicion, adherence to investigation guidelines and resection can prevent growth to such a size, and allow the patient to achieve the excellent prognosis this type of neoplasm typically confers.

Paediatric section

Chairman: Mr Nico Jonas

Recurrent tracheoesophageal fistula following laryngeal cleft repair in a female infant

A B Alli, M D Elloy, B E J Hartley

From Great Ormond Street Hospital for Children, London

Introduction

Laryngeal cleft accounts for approximately 1 per cent of congenital laryngeal malformations and arises as a failure of rostral extension of the tracheoesophageal septum in week five of embryonic development. It may be associated with other congenital anomalies, including those of the gastrointestinal tract.

Case report

We present the unusual case of an infant born with a congenital left-sided diaphragmatic hernia, hiatus hernia and a type three laryngeal cleft, which recurred despite repeated surgical intervention because of an undiagnosed upper oesophageal hamartoma that disrupted the cleft repair suture line.

Imaging

Video documentation, shown at the meeting, demonstrated the recurrent tracheoesophageal fistula, recurrent laryngeal cleft and the oesophageal hamartoma seen at microlaryngo-bronchoscopy.

Histopathology

Histological inspection revealed a benign, pedunculated, hamartomatous polyp originating at the level of the cricoid cartilage, with a central elastic cartilaginous core surrounded by adipose tissue, mucinous glands and lymphoid aggregates, and lined with non-keratinising squamous and respiratory epithelium. Professor Leslie Michaels commented that its appearance represented a replica of a developing trachea.

Management

The cleft and fistula were repaired via laryngofissure on three occasions. The hamartoma was eventually identified on oesophagoscopy and resected. The subsequent cleft and fistula repair was successful.

Discussion

The consensus of the meeting was that, given its histological composition, the polyp probably arose as part of the same process that gave rise to the laryngeal cleft and, as such, was congenital in origin. Formal oesophagoscopy is recommended prior to the repair of large laryngeal clefts to exclude the presence of an oesophageal lesion that may compromise the surgical intervention. Mr Nico Jonas asked if there was a risk of recurrence following incomplete excision, and Professor Leslie Michaels confirmed that this was possible.

A rare case of paediatric epistaxis

S Shah, R Nash, G Mochloulis

From the Lister Hospital, Stevenage

Case report

We report a rare presentation of an epithelioid haemangioendothelioma found in the nasal cavity.

A 15-year-old girl who had presented intermittently to her local hospital with repeated episodes of epistaxis was admitted with a haemoglobin concentration of 5.8 g/dl. She underwent examination under anaesthesia and cautery, at which time a polyp was found on the right side, which was excised. The sample was sent to Addenbrooke's Hospital for histological analysis, and was reported as an epithelioid haemangioendothelioma. The patient underwent subsequent magnetic resonance imaging (MRI), and at the time of writing was being followed up in the clinic.

Radiology

The MRI showed a normal appearance of the nasal cavity on the right side, with apparently normal turbinates, apart from a 2-mm artefact on the floor of the cavity, likely to be related to the resection.

Histology

The lesion comprised plump, eosinophilic, epithelioid cells with large, vesicular nuclei and small nucleoli. There was no significant cytological atypia. There were abundant red blood cells, but well-formed vascular channels were inconspicuous. The lesional cells stained strongly positive for cluster of differentiation 31 protein and vimentin. Overall appearances were most consistent with an angiomatous endothelial nodule.

The slides were reviewed by Professor Leslie Michaels, who agreed with the diagnosis of epithelioid haemangioendothelioma.

Conclusion

There are only two previously reported cases of epithelioid haemangioendothelioma occurring in the nose.

Ganglioneuroma presenting as a large parapharyngeal mass

A Vijendren, I Eladawy, P Kothari

From Luton and Dunstable Hospital

Background

Tumours of the parapharyngeal space are uncommon, comprising less than 1 per cent of all head and neck neoplasms.

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Seventy per cent of them are benign, and classically salivary or neurogenic in origin. Their incidence is further reduced in the paediatric population.

Case report

A 15-year-old girl was referred with an incidental finding of a bulge in the left lateral oropharyngeal wall. On examination, a smooth mass was seen on the left side of the pharynx extending from the skull base to the epiglottis, causing a shift of the oropharyngeal structures.

Radiological findings

A magnetic resonance imaging (MRI) scan of the neck showed a large mass in the left retropharyngeal and parapharyngeal space. The lesion had medialised the lateral and posterior pharyngeal wall while lateralising the internal carotid artery and jugular vein.

Histological findings

Histological inspection showed a spindle cell matrix with interspersed mature ganglionic cells, consistent with a ganglioneuroma.

Management

The patient underwent a transoral excision during which 80 per cent of the mass was removed. Post-operatively, the patient developed a left-sided Horner's syndrome and mild vocal fold palsy, which were managed conservatively. Repeat MRI scanning three months later showed a much smaller mass in the left carotid space, partially encasing the left internal carotid and displacing the left internal jugular vein laterally.

Discussion

In this patient, the entire mass could not be removed due to its close proximity to the skull base and major vessels in the neck. Close monitoring of this patient will be required as ganglioneuromas can recur or, rarely, become malignant. Professor Leslie Michaels recalled he had diagnosed one case of transformation from an aggressive to a more benign lesion. Mr Grant Radcliffe suggested that an external approach would have allowed complete tumour excision. The initial operation was performed with the intention of obtaining tissue for histology. Subsequently, a joint decision was made with the patient to monitor the situation. Mr Nico Jonas enquired if this lesion could have been diagnosed via fine needle aspiration cytology, and Professor Leslie Michaels confirmed that this may have been possible.

Conclusion

These benign masses are usually asymptomatic; thus, management options should be thoroughly discussed with the patient as surgery can cause significant morbidity.

Profound hearing loss after blunt trauma to the head in an 11-year-old

T Jacques, R Nash, S J Quinn

From the Lister Hospital, Stevenage

Introduction

Enlarged vestibular aqueduct syndrome is a rare congenital disorder causing audiovestibular symptoms in response to minor head trauma. Its management is challenging due to a lack of effective surgical treatment and an unpredictable clinical course. Its presence is associated with mutations in the SLC26A4 (pendrin) gene, both as an isolated disorder and as part of a syndrome leading to congenital deafness (e.g. Pendred's syndrome).

Case report

An 11-year-old girl presented with a long-standing, profound, left-sided hearing loss. Enlarged vestibular aqueducts were identified on magnetic resonance imaging (MRI). She had no other syndromic features or abnormal findings. The hearing loss was managed conservatively with advice regarding head injury avoidance. However, whilst trampolining she sustained a trivial blow to the right side of the face. This caused a sudden, profound hearing loss on the right side, leaving her with no useful hearing. She had no significant vertigo.

Radiology

At the time of diagnosis, MRI revealed bilateral enlarged vestibular aqueducts and endolymphatic sacs (the right was more severely enlarged than the left). There were no cochlear or neural abnormalities.

Management

The patient's sudden hearing loss was unsuccessfully treated with systemic steroids and intratympanic dexamethasone. The patient's profound hearing loss was persistent, and she was therefore referred for cochlear implantation.

Conclusion

Enlarged vestibular aqueducts should be excluded in cases of sensorineural deafness in young patients, particularly in cases of stepwise deterioration. If detected, patients should be counselled effectively and monitored. In the event of profound deafness, these patients are felt to be good candidates for cochlear implantation.

A 14-year-old, hypothyroid patient with enlarged right thyroid lobe

F Ryba, T Odutoye, J Williams

From St George's Hospital, London

Introduction

Paediatric thyroid disease is known to be more aggressive in nature. We present a rare variant of papillary thyroid cancer, diffuse sclerosing papillary carcinoma, which demonstrated typical features of this unusual form first described in 1985.

Case report

A 14-year-old girl presented with a right-sided thyroid swelling. She had a strong family history of thyroid disease and had been treated with thyroxine for many years for idiopathic hypothyroidism. She also had a known protein C deficiency.

Histopathology

Professor Leslie Michaels pointed out the classical histological features found in the thyroidectomy specimen: papillary epithelium, clear cells with a clear nucleus (so-called 'Orphan Annie' cells), psammoma bodies (concentric calcified bodies), fibrosis and lymphocytic infiltration.

A selective neck dissection removed 25 nodes, 13 of which had metastases.

Radiology

An initial uptake scan showed multinodular change on the right, with a normal left lobe. Magnetic resonance imaging showed suspicious lymphadenopathy on the right side of the neck.

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Management

Initially, a right hemithyroidectomy was performed. Following multidisciplinary team discussion and further imaging, the patient underwent a completion thyroidectomy and right selective level III–IV and bilateral level VI neck dissection, with post-operative radioactive iodine therapy. The patient was discharged on thyroxine, calcium and alphacalcidol.

Conclusion

Diffuse sclerosing papillary carcinoma is a rare but aggressive variant form of papillary thyroid cancer more commonly found in younger females. It metastasises early to regional lymph nodes. An aggressive therapeutic approach can result in a similar prognosis to that of the classical type.

Rhinology section

Chairman: Mr Ravinder Natt

A rare, benign growth causing progressive and complete destruction of the nose and nasal cavity

N Foden, S Vasani, C A East

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

Introduction

Eosinophilic angiocentric fibrosis is an extremely rare fibrosing vasculitis of unknown aetiology. It is most commonly seen in the upper airways.

Case report

A 66-year-old woman presented with a history of a swelling on the lateral aspect of her nose. A computed tomography scan was performed followed by an incisional biopsy, establishing a diagnosis of eosinophilic angiocentric fibrosis.

Radiological findings

Computed tomography of the sinuses showed non-specific changes. There was marked, heterogeneous thickening of the anterior cartilaginous septum with extension to the right side of the nose including the alar region. The overlying soft tissues were also involved.

Histological findings

Histological analysis revealed concentric whorls of perivascular fibrosis ('onion skin') and a mixed inflammatory cell infiltrate with frequent eosinophils. Both inflammation and fibrosis could be seen in the same slides, indicating an evolving disease process.

Management

The patient opted for a rhinectomy followed by an autologous repair. Reconstruction was made using a rib graft, radial forearm flap and a forehead flap for cover.

Discussion

Eosinophilic angiocentric fibrosis is extremely rare, and the diagnosis will most likely be made by excluding other causes, such as Wegener's granulomatosis, Churg–Strauss syndrome, infection, Kimura disease and granuloma faciale. Professor Leslie Michaels stated that, although rare, there is probably some overlap with the aforementioned differential diagnoses.

Management of these patients is not easy. No evidence of pharmacological benefit has been found. Surgical resection appears to offer the best outcomes but recurrence rates are high. Professor Michaels commented that historically these lesions may have been termed midline granulomas, and that steroid treatment will often cause exacerbation, with radiotherapy being used to induce regression.

Unilateral nasal mass in a 53-year-old: a 'red flag' sign?

N Amin, R Roplekar, S Lew-Gor

From Worthing Hospital

Introduction

Unilateral sinonasal masses are suspicious for malignancy. However, some benign pathologies present similarly.

Case report

We report a 53-year-old man who presented with progressive unilateral nasal congestion, clear rhinorrhoea, bilateral facial pain and symptoms of eustachian tube dysfunction. He had a history of septoplasty and endoscopic sinus surgery eight years previously, but no other past medical history. Anterior rhinoscopy showed right-sided mucopus and an obstructing lesion. Flexible nasendoscopy identified a polypoidal mass originating from the right choana.

Radiological finding

A computed tomography scan demonstrated a large, lobulated, polypoidal mass filling the nasopharynx and extending into the right nasal passage. There were some areas of calcification but no invasive features. No radiological diagnosis was offered.

Management

The patient underwent urgent endoscopic resection without complication. The mass was fully resected and sent for histopathological analysis. At follow-up appointments, the patient reported symptom resolution with no evidence of recurrence.

Histological findings

Histological analysis demonstrated hypertrophic mucosal glands lined with multi-layered, ciliated respiratory epithelium. Deep ducts connected to the surface, with periglandular hyalinisation of the basement membrane. The diagnosis was respiratory epithelial adenomatoid hamartoma.

Conclusion

Although often a sign of malignant disease, a unilateral nasal mass may be benign, and careful clinical, radiological and histopathological studies are required to avoid unnecessary intervention.

A rare, life-threatening aetiology of a common presentation, epistaxis

R Wong, S P Patel, S Watts

From the Royal Sussex County Hospital, Brighton

Introduction

We present a rare case of clival chordoma, the surgery of which was complicated by torrential bleeding. Clival chordomas are slow-growing tumours of the notochord remnants within the axial skeleton, and are often managed by debulking and radiotherapy.

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Case report

A 32-year-old man underwent endoscopic debulking of a clival chordoma; due to the extent of the disease, it was estimated that a 20 per cent resection had been achieved. The patient presented two weeks later with sudden-onset vertigo, hearing loss and severe epistaxis, and progressed to unconsciousness. Following resuscitation, examination revealed a right eye divergent squint and left temporal nerve paralysis.

Radiological findings

Magnetic resonance imaging showed a residual, large ($29 \times 21 \times 31$ mm), lobulated, enhancing mass arising from the clivus, with compression and displacement of the brainstem and pons.

Management

Initial management included haemorrhage control with local measures and fluid resuscitation. The underlying dilemma was whether to continue conservative management, with the risk of further haemorrhage from the sphenoidal sinus, or to take a more aggressive approach including surgical clipping or angio-embolisation. We adopted a conservative approach and closely monitored the patient. No further haemorrhage occurred.

Discussion

Professor Leslie Michaels confirmed the diagnosis of a clival chordoma.

Conclusion

Currently, treatment strategies are not well defined in the acute setting. This case demonstrates the importance of considering a conservative approach in patients who are haemodynamically stable.

Chronic nasal obstruction in a patient with rheumatoid arthritis

E Warner, S Ahluwalia, C A East

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

Introduction

We report a case of chronic nasal obstruction secondary to mucocutaneous leishmaniasis. A 53-year-old Caucasian man presented with a 4-year history of worsening nasal obstruction, rhinorrhoea, anosmia and vestibulitis. His only co-morbidity was rheumatoid arthritis, well controlled with methotrexate and adalimumab.

Case report

The patient's nasal symptoms progressed despite treatment with combinations of douches, steroid creams and antibiotics. Skin prick testing was negative and bloods tests (including anti-neutrophil cytoplasmic antibodies and angiotensin-converting enzyme concentrations) were normal. Initial biopsies showed 'mild nuclear atypia'. A magnetic resonance imaging scan was performed and showed a 'non-specific appearance, soft tissue thickening … no obvious bone destruction'. Subsequent nasal biopsies showed no neoplasia and special stains detected no micro-organisms. The patient was treated for probable anti-neutrophil cytoplasmic antibody negative, atypical Wegener's disease. Biopsies showed pathology was 'not pathognomonic, but in keeping with Wegener's disease'. The swelling reduced on treatment.

Six months later, the symptoms recurred. Nasal biopsies were taken and sent to the Hospital for Tropical Diseases for analysis. Leishmaniasis organisms were seen on microscopy and *Leishmania donovani* was diagnosed on polymerase chain reaction. This disease had probably been acquired on annual trips to Greece, rather than a more exotic location. Twenty-eight days of pentavalent antimonial treatment lead to complete resolution of the lesion.

This case highlights the difficulty of diagnosing this treatable disease: the presentation varies depending on the infecting species, and the clinical, radiological and histopathological features are non-specific. Inexpensive airfares have increased the number of UK inhabitants exposed to leishmaniasis, and it should be considered in the differential diagnosis of non-resolving mucosal lesions, particularly in patients taking immunomodulatory therapies.

Professor Leslie Michaels commented that Wegener's granulomatosis is often over-diagnosed and labelled as 'atypical'. He reminded the meeting that Wegener originally described lesions that affected the nose, lungs and kidneys.

A unilateral, enlarging, hard swelling over the frontal sinus

E H C Wong, W Y Yang, D Lowe

From James Cook University Hospital, Middlesbrough

Introduction

Sinonasal lymphoma is rare and can mimic infectious, non-lymphomatous neoplastic and granulomatous processes in its initial presentation, thus delaying diagnosis.

Case report

A 61-year-old woman presented with a 4-week history of a gradually enlarging, right-sided forehead swelling, which was smooth and firm with no tenderness, fluctuation or overlying skin changes.

Radiological findings

Initial computed tomography (CT) of the paranasal sinuses revealed a mildly opacified right frontal sinus with an overlying soft tissue swelling. A repeated CT scan one week later showed evidence of osteomyelitis of the medial wall of the right frontal bone and orbit, suggesting a diagnosis of Pott's puffy tumour. No pus drained from a frontal sinus trephine, and biopsies were taken. Following a period of antibiotic treatment, the lesion continued to enlarge. A third CT scan demonstrated a collection over the frontal sinus.

Histological findings

Histopathological analysis of the frontal sinus biopsy showed diffuse large B cell lymphoma.

Management

The patient was treated successfully with six cycles of chemotherapy.

Discussion

Pott's puffy tumour is primarily a clinical diagnosis, and care should be taken not to assume a benign diagnosis based on radiology alone. Dr Ata Siddiqui noted that in the first CT scan there was no expansion of the frontal sinus and most

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of the soft tissue was outside the sinus, which would point to a diagnosis other than Pott's puffy tumour.

Head and neck section

Mr Jean-Pierre Jeannon

A progressively enlarging goitre

H Dixon, S Maskell, M Black
From William Harvey Hospital, Ashford

Introduction

Thyroid masses can present in a variety of manners, with patients often noting a neck lump or a change in the size of the gland. This report presents an unusual thyroid mass.

Case report

A 59-year-old woman was referred with a progressively enlarging goitre. It had been present for 10 years and had increased in size over the last 4 years. Initial examination and investigation, including a fine needle biopsy, suggested benign disease. However, continued expansion of a dominant nodule prompted a diagnostic right hemithyroidectomy.

Radiological findings

The initial computed tomography (CT) scan showed a large, heterogeneous, vascular mass in the right lobe of the thyroid, measuring 70 mm. Post-operative CT scans of the neck and chest showed no lung lesions, and highlighted a small right axillary sebaceous cyst.

Histopathological findings

Histopathological analysis with immunohistochemical staining suggested a biphasic malignant tumour with a large sarcomatoid and smaller epithelioid component. An opinion from the Royal Marsden Hospital agreed with a histological diagnosis of poorly differentiated carcinoma with extensive sarcomatoid change (carcinosarcoma).

Management

A right hemithyroidectomy with clear margins was deemed to be therapeutic, and the head and neck multidisciplinary meeting recommended completion radiotherapy.

Discussion

Discussion at the Semon club meeting agreed with the management plan instigated. Professor Leslie Michaels commented on the unusualness of a diagnosis of carcinosarcoma within the thyroid, and noted that this had contributed to the complexity of the patient's management.

A history of preceding goitre does not exclude new thyroid disease, and unusual diagnoses should always be discussed in an appropriate multidisciplinary forum to ensure optimal patient management.

A neck lump presenting with acute dysphagia and dyspnoea in a young man

R M Naik, A A Qureshi, A Hannan
From the Royal Free Hampstead NHS Trust, London

Introduction

Synovial sarcoma is a rare soft tissue tumour most frequently found in the limbs. We present an occult case of this uncommon entity presenting acutely within the thyroid and surrounding structures.

Case report

A 20-year-old, Australian man presented with acute symptoms of sore throat, dysphagia and dyspnoea. A 3×3 cm, soft, left-sided neck mass was palpated. Nasolaryngoscopy revealed an erythematous mass arising from the left pyriform fossa.

The differential diagnosis included a parapharyngeal mass and thyroglossal or branchial cyst.

Sudden airway deterioration resulted in emergency neck exploration, revealing a mass arising from the left thyroid lobe and extending into the neck. An incision biopsy was taken and a tracheostomy tube inserted.

Radiology

Prior to surgery, ultrasonography of the neck demonstrated a 3.2×2.9 cm, lobulated lesion contiguous with the left upper pole of the thyroid, featuring intermediate echogenicity, which yielded acellular, blood-stained fluid on aspiration. Computed tomography revealed a 7.5×3.6 cm mass extending from the left pyriform fossa up to the thyroid.

Histology

Biopsy revealed islands of epithelial proliferation, composed of plump spindle cells and focal glandular differentiation. Immunohistochemistry pointed towards a diagnosis of biphasic synovial sarcoma. Professor Leslie Michaels agreed with this diagnosis.

Management

Definitive management included total laryngectomy and radiotherapy.

Discussion

Masses within the neck in young adults are commonly attributed to congenital causes. However, it is paramount to maintain a high index of suspicion for malignancy, and to be cautious of sudden airway deterioration.

Dysphagia, weight loss and haematemesis

J K C Chan, S Kadirkamanathan, A Pace-Balzan
From Broomfield Hospital, Chelmsford

Introduction

We report an unusual case of a hypopharyngeal lipoma extending per oesophagus into the stomach.

Case report

A 71-year-old woman presented with a long history of progressive dysphagia, weight loss and recent haematemesis. She had previously been diagnosed with achalasia, for which she had been treated with botulinum toxin injection. Symptoms persisted, and oesophagoduodenoscopy revealed pseudo-achalasia secondary to a pedunculated, post-cricoid lesion that extended the full length of the oesophagus into the stomach, with bleeding distally.

Radiology

Computed tomography (CT) confirmed the presence of a large, fatty, $165 \times 40 \times 45$ mm mass extending from the post-cricoid area, through the full length of the oesophagus, to the greater curve of the stomach.

Histology

Histological analysis revealed a large, pedunculated mass composed of normal oesophageal-type squamous epithelium stretched over an unencapsulated but well circumscribed

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mass of mature adipose tissue. No malignancy was evident. A diagnosis of benign lipoma was made.

Management

The tumour was excised. The hypopharyngeal pedicle was exposed using a Weerda bi-valve diverticuloscope, and was stapled and divided endoscopically. The tumour was delivered through a gastrotomy by an upper gastroenterological surgeon. Recovery was uneventful, and no recurrence was evident on repeated oesophagoduodenoscopy at six months.

Conclusion

We present a case of pseudo-achalasia secondary to a hypopharyngeal-oesophageal-gastric lipoma. It is necessary to differentiate between pseudo-achalasia and idiopathic achalasia as the two conditions are managed differently.

Discussion

The value of CT in identifying the fatty nature of this unusual tumour was highlighted.

An unusual case of rapidly progressive neurological deterioration

L Prichard, M Rollin, F Vaz

From University College Hospital, London

Introduction

Paraneoplastic cerebellar degeneration is a rare syndrome known to be associated with small cell lung cancer, Hodgkin's lymphoma, thymoma, and testicular and gynaecological cancers. It presents with progressive truncal ataxia, limb ataxia and cerebellar dysarthria occurring within 12 weeks. Symptoms often predate the discovery of the tumour and response to treatment is rare. We present a case of paraneoplastic cerebellar degeneration associated with papillary thyroid carcinoma, with rapid neurological improvement in four weeks following tumour resection.

Case report

A 71-year-old man was referred for a neurological opinion following a 4-month history of pain across his shoulders, bilateral paraesthesia of his fingers and increasing unsteadiness of gait. His sensory and motor system rapidly deteriorated, forcing him to use a wheelchair. Cerebrospinal fluid sampling disclosed a weakly positive Collapsin response mediator protein 5 ('CRMP5') titre. This onconeuronal antibody result, in combination with neurological symptoms and thyroid malignancy, helped to confirm the diagnosis of paraneoplastic cerebellar degeneration syndrome.

Radiological findings

Magnetic resonance imaging of the brain and spinal cord was normal. However, a whole-body positron emission tomography computed tomography scan revealed a 1.1×0.9 cm, right-sided, fluorodeoxyglucose-avid thyroid lobe nodule.

Histological findings

Formal histology confirmed the diagnosis of papillary thyroid carcinoma, with a 15 mm, macroscopic lesion in the right lobe graded as T_{1b} N_0 M_0.

Management

The patient underwent total thyroidectomy and radioiodine ablation, followed by lifelong levothyroxine treatment.

Discussion

Colleagues at the meeting reported that they had not encountered such an unusual case, and expressed pleasant surprise at this patient's dramatic recovery four weeks after tumour resection.

An unusual subglottic swelling presenting with dysphonia

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From Stepping Hill Hospital, Stockport

Introduction

We present the case of a 61-year-old man with a 6-month history of dysphonia, who was found to have a left subglottic swelling on endoscopic examination.

Case report

Flexible nasendoscopy revealed a smooth, submucosal, subglottic swelling contiguous with the left vocal fold. Direct examination was performed and the lesion was biopsied.

Radiological findings

A computed tomography scan of the neck identified a swelling in the left subglottic region, arising from the inferior aspect of the left vocal fold, with no associated lymphadenopathy. Incidentally, a large left thyroid nodule was noted.

Histological findings

The histological appearance was in keeping with a benign tumour of neural origin, likely to be a schwannoma due to positive S100 staining. A tertiary centre review of the histology confirmed the lesion to be a benign peripheral nerve sheath tumour with appearances consistent with an extracranial schwannoma.

Management

At the time of writing, the patient was awaiting definitive subglottic excision, postponed due to radioiodine ablation therapy for thyroid cancer. The thyroid fine needle aspiration result was Thy3f, and he underwent a diagnostic left hemithyroidectomy. Histology revealed a Hurthle cell carcinoma, and completion thyroidectomy was performed. Subsequent histology revealed an additional area of follicular variant papillary micro-carcinoma.

Discussion and lessons learned from the meeting

The surgical options for benign tumours in this location are endoscopic or external 'cold steel' excision, or endoscopic laser excision.

'Take-home' message

This case illustrates the importance of fibre-optic examination of all patients with persistent dysphonia, together with histological assessment of all subglottic masses. Additionally, all incidental thyroid masses should be investigated.

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