CASE REPORT

Aneurysmal bone cyst of the ethmoid bone

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SUMMARY

A 90-year-old woman presented with headache and diplopia, and a mass in the nasal cavity and paranasal sinuses was found on CT scan. The patient underwent examination under anaesthesia showing a mass arising from the ethmoid, apparently from the nasal septum. Biopsies taken showed an aneurysmal bone cyst. The patient opted against surgery and has been managed conservatively. A second CT scan 4 years later showed significant increase in size.

BACKGROUND

Aneurysmal bone cysts (ABCs) are non-neoplastic tumours, first described by Jaffe and Lichtenstein in 1942.¹ The pathophysiology is so far poorly understood, but are known to affect mainly metaphysis and diaphysis of long bones as well as spinal vertebrae and pelvis. Skull ABCs are rare and represent a diagnostic and treatment challenge. In this case the lesion originated from the ethmoid bone, and intraoperatively appeared to originate from the nasal septum. The lesion caused a significant erosion of the surrounding structures and documented a relatively fast growth. To the best of our knowledge, a case of ABC arising specifically from the nasal septum has not been reported before, and those arising from the ethmoid bones more generally are also very rare.

CASE PRESENTATION

A 90-year-old woman presented to the acute medical unit with a 2-week history of right temporal headache and diplopia. Background medical conditions included ischaemic heart disease, stage 3 chronic kidney disease and a recently excised squamous cell carcinoma of the right lower leg. An ischaemic stroke was suspected and a CT of the head was performed, revealing a large mass seemingly centred on the ethmoid and sphenoid sinuses.

A full system-based examination of the cardiac, respiratory and nervous systems revealed no abnormalities. There were no signs of raised intracranial pressure or meningism. The patient remained non-feverish throughout the admission.

Cranial and peripheral nerve examination was performed and showed no abnormality other than diplopia with right ophthalmoplegia. However these seemed to have settled spontaneously within 2 days of admission. No proptosis or chemosis was seen. There was no nystagmus and visual field testing did not reveal any bitemporal hemianopia. No loss in visual acuity was noted.

Examination of the ear, nose and throat revealed a mass in both nostrils with no abnormality in the ears or throat.

INVESTIGATIONS

The initial CT of the brain and paranasal sinuses (figure 1) showed a 4.2 cm×3.6 cm heterogeneous mass centred on the ethmoid and sphenoid sinuses causing expansion and erosion of the lateral walls of the ethmoid air cells. High-density fluid level was noted within the mass suggestive of some proteinaceous material or blood products. The mass was abutting the sella turcica superiorly; however, the optic chiasm was not compressed.

The patient's full blood count and biochemistry were entirely normal. Erythrocyte sedimentation rate was not raised and there was no scalp tenderness suggestive of temporal arteritis.

DIFFERENTIAL DIAGNOSIS

On the basis of radiology, the mass was thought to be an inflammatory mucocoele or a slow growing sinonasal mass compressing and eroding surrounding structures. No intracranial abnormality or ischaemic stroke was noted.

Other clinical differential diagnoses would include inverting papilloma, low-grade sinonasal carcinoma, fibrodysplasia or cemento-ossifying/ossifying fibroma, giant cell lesion or fungal mycetoma.

Formal diagnosis would require histological confirmation.

TREATMENT

The patient was booked urgently for an examination of the nose and biopsies under general anaesthetic, with consent to proceed to a debulking/excision procedure if amenable. Propofol induction was used by the anaesthetist, with desflurane and nitrous oxide maintenance. Examination revealed a cystic mass appearing to arise from the bony (posterior) nasal septum portion of the ethmoid and expanding bilaterally from the nasal septum to block both nasal cavities by approximately 80%. Adhesions of the mass to both middle turbinates were noted. The nasal septal cartilage was partially eroded by the mass.

During the surgery multiple biopsies were taken; the absence of an amenable plane between the nasal cavity and the mass meant that full excision was not attempted and the mass was debulked to give the patient symptomatic relief of nasal obstruction.

The patient was discharged home the following day with a short course of nasal steroids to reduce postoperative inflammation.

OUTCOME AND FOLLOW-UP

Histopathology showed a composition of spindle cell and giant cell proliferation interspersed with haemosiderin and small trabeculae of bone.

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Figure 1 Axial section of the CT scan at initial presentation showing cystic, soft tissue mass causing expansion of the nasal cavity.

A provisional diagnosis of giant cell lesion was made and the sample was sent to the regional bone tumour service for confirmation. The histology was subsequently reported as an aneurysmal bone cyst with fairly typical features. There was no evidence of fibrous dysplasia, cemento-ossifying or ossifying fibroma, or other findings often associated with ABCs.

The histological diagnosis was consistent with previous imaging.

The patient was kept under regular ENT (ear, nose and throat) review following the debulking procedure. The lesion was noted to be slowly increasing in size. After appropriate consultation the patient rejected further surgical intervention,

relying on nasal steroids for symptomatic relief of nasal obstruction.

Four years after initial presentation, the patient was admitted to the hospital for management of a haemorrhagic stroke. During the workup for her stroke, a repeat CT of the brain and paranasal sinuses was organised (figures 2 and 3). This showed a significant increase in the size of the ABC with posterior extension to the pterygoid plates and significant thinning of the lamina papyracea. The haemorrhagic stroke has left her severely dysarthric and she has significant mobility problems.

After discussion with the patient and members of the multi-disciplinary team, the decision was to manage this conservatively with symptomatic relief only. She reports new symptoms of midfacial pain and bilateral ocular discomfort. These have been well controlled with regular analgesia. She remains under regular ENT follow-up.

DISCUSSION

Aneurysmal bone cysts were identified in 1942 and named descriptively for their characteristic gross physical features: a cystic, blood-filled cavity within an expanded or 'aneurysmal' bony wall.^{2 3} CT scan will show a thinned or interrupted cortex with a well-defined soft tissue mass (figure 2).^{2 3} A 'soap bubble' appearance and fluid–fluid levels are considered characteristic (figure 3).^{3 4} Imaging methods such as CT are useful in evaluating the extent of the lesion, especially in areas such as the head and neck, where the bony anatomy is complex.³ Definitive diagnosis is made histologically, and this typically shows irregular, blood-filled chambers with islands of bone and fibrous tissue.² These spaces are surrounded by rare spindle-shaped endothelial cells and multinucleated giant cells.⁴

There is limited knowledge of the pathophysiology and natural history of these tumours. They are more commonly encountered in women, and in the second decade of life. The rate of growth is usually relatively slow although, especially when arising from the skull, ABCs can be locally destructive.

Figure 2 Coronal sections of the CT scan at 4-year follow-up delineating the cyst and exemplifying the 'aneurysmal' bony wall with cortical thinning and interruption.

Figure 3 Axial sections of the CT scan at 4-year follow-up, showing increased size and the typical features of expanded bony wall and fluid–fluid levels within the cysts.

ABCs arising from the head and neck region are rare, and those arising from the nose rarer still. These have been reported to be possibly associated with 17p13 translocation⁵ and previous nasal fracture.⁶ Vascular, traumatic and genetic origins have all been suggested, and all remain controversial.⁷ One theory that the ABCs are a result of vascular degeneration within another benign lesion, that is, as secondary lesions, is no longer felt to be sufficient to explain most of these tumours.²

Treatment for ABCs in general is usually removal, often by curettage, although adjuvant radiotherapy is also sometimes used, and more rarely, cryotherapy, sclerotherapy and radio-nuclide ablation.⁷ Recurrence rates have been quoted at 10–30% after treatment.⁶

The case that we have presented adds to the literature by showing the natural history of an aneurysmal bone cyst if it is not completely excised or debulked regularly. Although these are felt to be benign neoplastic lesions, our case shows that they

tend to compress rather than invade neighbouring structures and our serial imaging can give an estimation of the rate of growth of the lesion. It is felt that patient's main symptoms of diplopia and headache resulted from the expansile nature of the tumour exerting pressure on the orbits and possibly sella turcica, although the optic chiasm was noted not to be compressed. The tumour did not invade any of these structures, and debulking was able to afford the patient some relief.

It is also one of very few reported cases to arise from the ethmoid bone. There are several case reports of aneurysmal bone cysts arising from the skull, including the various paranasal sinuses. There are a few reports of ABCs arising from within the nasal cavity such as the middle turbinate⁴ but as far as the authors are aware, not from the nasal septum, the involvement of which caused the patient's bilateral symptoms.

Contributors NJ and MC performed the literature search for the report and took main responsibility for writing the article. RS was involved in patient management, suggested the case report and contributed to critically reviewing and modifying the written article. AM was the consultant in-charge of patient's ENT management and also contributed to critically reviewing the written article.

Competing interests None.

Patient consent Obtained.

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Learning points

- ▶ Awareness of aneurysmal bone cysts as a differential diagnosis for space occupying lesions in the skull and skull base.
- ▶ Appreciation that although a benign tumour, aneurysmal bone cysts may cause symptoms due to its expanding, compressive and locally destructive nature, particularly within a contained area such as the skull.
- ▶ Understanding of the natural history of untreated aneurysmal bone cysts
- ▶ Particular physical, radiological and histological characteristics of aneurysmal bone cysts that can aid diagnosis. These include:
 - An expanded bony wall surrounding blood-filled cysts.
 - Fluid–fluid levels within the cysts on imaging.
 - Islands of bony and fibrous tissue in blood-filled chambers surrounded by spindle and giant cells.

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