CASE REPORT

Aneurysmal bone cyst of temporal bone

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SUMMARY

Aneurysmal bone cysts (ABC) are benign neoplasms frequently occurring in the long tubular bones. It is very rare in temporal bone. We report a case of ABC of the left temporal bone in an 8-year-old Asian boy who presented clinically with swelling over the left temporal region for 5 months. CT and MRI features were suggestive of ABC. Surgical resection was performed and on follow-up the patient was doing well. CT and MRI are the imaging modalities for proper evaluation of ABC, aiding to diagnosis and helpful in treatment planning.

BACKGROUND

Aneurysmal bone cysts (ABC) are rapidly growing, benign, non-neoplastic osteolytic lesions that consist of an intraosseous destructive tumour and an extraosseous aneurysm-like cyst. This entity was first described by Jaffe and Lichtenstein in 1942.¹ ABCs arise predominantly from the long tubular bones of the extremities, vertebrae or flat bones. ABC of the skull is very rare and it accounts for 3–6% of all ABCs.^{2–3} Only few cases of ABC of the temporal bone have been reported in the literature.^{2–5} These tumours are usually seen in young patients during adolescence presenting as a tender, externally visible lump at the site of the underlying tumour.² We report a case of ABC of the left temporal bone in an 8-year-old boy diagnosed on CT and MRI followed by surgical excision and histopathological examination.

CASE PRESENTATION

An 8-year-old Asian boy presented with gradually increasing painless swelling in the left temporal region (figure 1) for 5 months with intermittent pain in the swelling for the last 20 days. There was no history of any trauma, ear discharge, vertigo, nausea, vomiting or seizures. Neurological and ophthalmological examination results were normal. Local examination revealed a non-tender, firm and hard mass, measuring approximately 6×5×4 cm in the left temporal region which was fixed to the underlying bone. Skin over the swelling was normal.

INVESTIGATIONS

Routine laboratory investigations were within normal limits. Plain radiography of skull (antero-posterior and lateral views) revealed a well-defined, round, radiolucent lesion with thick sclerotic rim in the left temporal bone (figure 2). Non-contrast and contrast-enhanced volumetric CT of the head and paranasal sinuses were performed in axial sections followed by coronal and sagittal reconstructions. CT scan revealed a large, extra-axial, expansile,

Figure 1 The patient showing swelling in the left temporal region.

multiloculated, osteolytic mass of mixed density involving the squamous and petrous part of left temporal bone showing fluid–fluid level within and inhomogeneous enhancement on contrast administration (figure 3A–C). The underlined left temporal bone was thinned out with erosion of petrous bone and some part of mastoid air cells (figure 3D). There was erosion of the left tegmen tympani and mandibular fossa; however, articular surface of temporomandibular joint was normal. The mass was protruding intracranially causing compression of the left temporal lobe and ipsilateral lateral ventricle, left tentorium and brainstem. Mild

Figure 2 Plain radiography of skull (lateral view) revealed a well defined, round, radiolucent lesion with thick sclerotic rim in the left temporal bone (black arrowheads).

To cite: Ansari S, Ahmad K, Gupta MK, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009434

Figure 3 (A) Axial section CT image showing a large, expansile, multiloculated, osteolytic mass in the squamous and petrous part of the left temporal bone with fluid–fluid level within, protruding in intracranial direction and causing compression of the brain parenchyma. (B) Coronal reconstructed CT image showing mixed density mass in the squamous and petrous part of the left temporal bone protruding in intracranial direction, causing compression of the brain parenchyma and ipsilateral lateral ventricle with mild midline shift towards right side. (C) Sagittal reconstructed CT image showing mixed density mass in the squamous and petrous part of the left temporal bone. (D) Axial CT image in bone window showing thinning of left temporal bone and erosion of petrous bone.

thickening of the scalp tissue on left temporal region was also seen.

The patient underwent MRI of brain. On T1-weighted MRI, the above-described mass was of mixed signal intensity with internal septations and hyperintense areas within, suggestive of haemorrhage (figure 4A). On T2-weighted MRI, the mass showed increased signal intensity with fluid–fluid levels within, multiple internal septations and hypointense rim around the mass (suggesting capsule; figure 4B,C). All the components of the mass gave a ‘bubbly’ appearance. On the basis of CT and MRI, diagnosis of ABC was made.

TREATMENT

Complete surgical excision of the cyst was performed, and on surgery the mass was surrounded by fibrous capsule. Radiotherapy was administered to the patient. Histopathological examination revealed blood filled cavernous spaces, separated by collagenous tissue containing fibroblasts and multinucleated giant cells along with extravasated red blood cells (RBC), hemosiderin, granulation tissue and inflammatory cells in the stroma. Features were suggestive of ABC.

OUTCOME AND FOLLOW-UP

Postoperative period was uneventful and the patient was symptom free on 3 months of follow-up.

DISCUSSION

ABCs are uncommon, rapidly growing benign bone tumours that account for 1–2% of all primary bone tumours.⁴ Eighty per cent of ABCs are seen within the first two decades of life, with a mean age at presentation of 16.1 years.⁵ ABC has a slight female preponderance, with a male-to-female ratio of 0.84:1.⁶ These lesions typically arise from the long tubular bones of the extremities, such as the femur, humerus, tibia and fibula. They are also commonly found in the vertebrae. Skull bones are a very rare site with an incidence of 3–6% of all ABCs. Most ABCs are primary that is, they occurs de novo. Secondary ABCs arise with a coexisting bone lesion and are reported to occur 30% of the time.⁷ An ABC can arise from a pre-existing chondroblastoma, osteoblastoma, chondromyxoid fibroma, giant cell tumour, fibrous dysplasia and rarely from a malignant bone tumour such as an osteosarcoma. Pathogenesis of ABC is not very clear, but according to some researchers it develops

Figure 4 (A) Axial section T1-weighted MRI showing mixed signal intensity mass with hyperintense areas suggestive of haemorrhage (black arrow). (B) Axial section T2-weighted MRI showing mixed signal intensity mass with fluid–fluid level and multiple internal septations (black arrows). Peripheral hypointense rim seen around the mass (black arrowheads). (C) Coronal section T2-weighted MRI showing increased signal intensity of the mass with multiple internal septations seen within the mass (black arrows). Peripheral hypointense rim seen around the mass (black arrowheads). The mass is compressing the brain parenchyma and ipsilateral lateral ventricle with mild midline shift.

following trauma or may be congenital or acquired lesion of intraosseous blood vessel walls or results from a chromosomal translocation $t(16;17)(q22;p13)$.^{3 8 9}

The clinical presentation of an ABC is related to the location and size of the lesion. ABC located in the temporal bone have manifested in the past as swelling with or without pain in the temporal region, decreased hearing, headache, trigeminal nerve palsy or facial nerve palsy.⁶ The previously reported cases presented with symptoms of raised intracranial pressure, dysphagia and decreased hearing, severe endolabyrinth-type perception deafness and hearing loss with recurrent meningitis. But in our patient there was no hearing loss and facial nerve palsy.

Different types of radiographic imaging modalities have been employed in the diagnosis of ABC. On standard skull radiographs, an ABC is characteristically described as a fusiform,

expansile lesion located eccentrically, and is often referred to as having a ‘soap bubble’ or ‘blowout’ appearance.^{2 6 8} CT findings show a multiloculated lesion originating in the diploe and expanding both intracranially and extracranially. The lesion may have both solid and cystic components with fluid–fluid level within and the solid components usually shows enhancement on postcontrast study.^{10 11} MRI is a valuable tool for aiding in diagnosis of ABC and it characteristically reveals an expansile lesion surrounded by a hypointense fibrous capsule on all sequences.¹² Hypointense internal septations may be present within the lesion, separating it into multicystic compartments with internal fluid–fluid layering derived from the layering of uncoagulated blood. The lesions usually impart intense contrast enhancement after intravenous gadolinium injection.^{7 10} Angiography often shows abnormal circulation with patchy areas of delayed venous

phase contrast and occasional arteriovenous shunts. Interestingly, unlike ABC in the skeleton, lesions in the skull are less likely to demonstrate pathological circulation.⁶

The definite diagnosis of an ABC is carried out by biopsy from the lesion with histopathological examination. Gross pathological examination shows honeycomb-appearing cyst comprising multiple sinusoidal spaces filled with unclotted blood and blood-tinged serous fluid.¹³ Histologically, these honeycomb-like spaces lack endothelial linings and are filled with haemorrhagic fluid. The thin septa within the cysts are composed of proliferated, spindle-shaped fibroblasts with scattered multinucleated giant cells of the osteoclast type, granulation tissue, hemosiderin-laden macrophages and extravasated red blood cells.

Complete surgical removal is the treatment of choice for an ABC, and it is curative. Other treatment modalities include en-bloc resection, curettage and bone grafting, endovascular embolisation, cryosurgery and radiotherapy.⁹ It has been noted that recurrence of ABC of the calvaria is rare, unlike ABCs in other locations.⁵ Radiotherapy is a possible consideration for treatment of recurrent ABC of the temporal bone.¹⁴ In our patient, there was no recurrence noted radiographically at the 3 months of follow-up.

Multimodality imaging usually predicts the diagnosis, but definitive diagnosis can only be made by histopathological examination. The treatment of choice is complete surgical excision. Megavoltage radiotherapy may be considered an option in inoperable cases or recurrent ABC.

Learning points

- ▶ Aneurysmal bone cyst of the temporal bone is a rare manifestation.
- ▶ On imaging, aneurysmal bone cyst appears as an expansile, well-defined, bone lesion with internal septations, low signal intensity rim of capsule, multiple cystic lobulations of varying signal intensities, fluid–fluid levels within some of the cysts and ‘Bubbly’ appearance.
- ▶ CT and MRI are the imaging modalities for evaluation of aneurysmal bone cyst, aiding in diagnosis and helpful in treatment planning.

Contributors All authors substantially contributed to the concept and design of the paper. SA and KA acquisition the data and drafted the article. SA, MKG and RKR revised the article for important intellectual content. All authors gave final approval of the article.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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