Aneurysmal bone cyst of the sphenoid with orbital involvement

Jill V Hunter, Cheryl Yokoyama, Ivan F Moseley, John E Wright

Abstract
We present a case of aneurysmal bone cyst involving the roof of the orbit and sphenoid bone, with plain film, computed tomography, and magnetic resonance imaging findings. The natural history and treatment depend on the presence of associated abnormalities such as fibrous dysplasia or a giant cell tumour. In this case the lesion was solitary and was successfully removed, so that possible complications from radiotherapy were avoided.

Aneurysmal bone cyst (ABC) is uncommon. The commonest site in 40–50% of cases is the metaphysis of a long bone. In the skull ABC is rare, representing less than 1% of cases: of 59 in the literature 15 involved the orbit,1–15 of which four affected the sphenoid bone1–4,12,13 and one the sphenoid sinus.10
There is an association between peripheral ABC and other bony abnormalities; an associated lesion has implications for the choice of treatment. We describe a boy with a lone ABC involving the greater and lesser wings of the sphenoid.

Case report
A 7-year-old white boy presented with a history of one month's variable, painless, periorbital swelling, without visual disturbance. There was no relevant past history, in particular no head injury. There was right sided periorbital swelling, with 4 mm axial proptosis. Visual acuity was 6/9 in both eyes; examination otherwise gave normal results. Plain skull radiographs showed a small mixed density lesion posteriorly in the roof of the orbit (Fig 1). Six days later there was no palpable mass or sign of inflammation. The right eye still showed 3 mm axial proptosis, unchanged with head positioning or the valsalva manoeuvre. The physical findings were thought to represent a small orbital haemorrhage.

The findings on examination remained unchanged despite further episodes of intermittent proptosis and lid swelling. Fourteen weeks later, however, the right eye was displaced 6 mm forwards, with limitation of upgaze; horizontal movement was less affected (Fig 2). Retinal striae were noted, though the optic disc appeared normal. Visual acuity was now 6/12 in the right eye. There was no afferent pupillary defect.
X-ray computed tomography (CT) (Fig 3) demonstrated a very large destructive lesion in the sphenoid bone with layering of high density material suggestive of a cavity containing blood.
Biopsy was performed via a lateral canthotomy (Mr J E Wright). A bluish-purple mass, displacing the peristemeum away from the bone towards the orbit, was incised and a cavity containing old blood was entered. Deep to this lay highly vascular, friable tissue, biopsy revealed multinucleate giant cells and osteoid consistent with ABC.
Magnetic resonance imaging (MRI) (Fig 4) showed a large, rounded, well defined mass apparently extending from the right sphenoid into the orbit. T1-weighted (IR 1500/500/40)

Figure 1: Postero-anterior (upper) and lateral (lower) radiographs at the time of presentation (November 1988). The right orbit is marginally larger than the left, and there is a small area of osteolysis with surrounding sclerosis (solid arrow) in its roof. The sphenoid ridge is slightly less distinct (open arrow) than its fellow.

Figure 2: Preoperative appearance with right proptosis.
images indicated the presence of clotted blood.

At frontal craniotomy (Professor D N Harrison) a large, vascular, mass was found occupying the orbital roof from 1 cm behind the orbital rim to the middle cranial fossa. Most of the roof and the posterolateral wall of the orbit was absent. The mass adhered to the optic nerve anteriorly. It was separated from the frontal lobe and orbital periosteum and removed piecemeal. The cavity was curetted and right

**TABLE 1 Clinical features of orbital ABC**

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<thead>
<tr>
<th>Reference number</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Side</th>
<th>Proposis</th>
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<th>Visual acuity</th>
<th>History (months)</th>
<th>Trauma</th>
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<td>22</td>
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<td>R</td>
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<td>Medial</td>
<td>R</td>
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Aneurysmal bone cyst is a benign, multicystic, vascular lesion of young people. Up to 85% of cases occur before 20 years of age, with one documented case in a 72-year-old. There is a slight female preponderance. Half of ABCs involve the metaphyses of long bones and one-fifth the spine. The skull and orbit are affected in about 1% of 59 cases reported in the skull 15 involved the orbit.

Other bony lesions are present in up to one-third of cases. In 57 extracranial ABCs the most common associated lesions were: solitary bone cyst (18), osteoclastoma (14), and osteosarcoma (12). Other associations include fibrous dysplasia, non-ossifying fibroma, chondroblastoma, fibromyxoma, and giant cell reparative granuloma. At presentation ABC may be large, with a history of insidious onset of pain (in more than half) and swelling (in one-quarter). Pathological fracture may occur in the long bones or spine. Symptoms have typically been present for less than three months, though 6% of patients do not present for one year or more. The recurrence rate for extracranial ABCs is 21–44%, higher rates occur with incomplete excision and in children.

In the orbit ABC has been reported in nine females and six males aged from 14 months to 42 years. One case had bilateral orbital involvement from a sphenoid sinus ABC. Presenting symptoms included painless proptosis, diplopia, ptosis, headache, visual deterioration, and nasal congestion. The average duration of symptoms was two months (Table I and Table II).

Aneurysmal bone cyst may arise in the roof and/or walls of the orbit. One proposed cause is trauma, but in none of the orbital tumours was injury noted.

Sudden proptosis secondary to rapid expansion of the lesion — presumably haemorrhagic — is common. The rapidity of progression between the slightly abnormal plain film and the grossly pathological CT only 14 weeks later was striking and raised the possibility or malignancy, as in other cases. However, Dabska and Buraczewski described four radiological phases: osteolysis; rapid growth; stabilisation; and healing, with progressive ossification. The short history suggests that many patients present during the phase of rapid growth. As in our case, signs of globe compression such as retinal striae may be seen.

None of the reported orbital cases was secondary to another lesion. In keeping with the benign nature of ABCs, all remained extradural.

Accepted methods of treating peripheral ABCs include excision, curettage (sometimes with bone grafting), cryotherapy, and radiotherapy. Some of these are not applicable to the orbit. Review of the literature reveals two cases of orbital recurrence, following needling and partial excision, within two years. Two patients received radiotherapy after surgery. One orbital ABC, mistaken for a giant cell tumour and treated with high-dose radiation, presented again as a heavily calcified mass. No cases of osteogenic sarcoma complicating radiotherapy have been reported in the orbit, though this has occurred with extracranial ABCs. Our patient remains well one year after operation.
CONCLUSION
Orbital aneurysmal bone cysts appear to form a well defined subgroup of ABC of the skull. Although rare, these potentially curable, benign lesions should be considered in young patients with painless proptosis of rapid onset.

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