CLINICAL STUDY

Aneurysmal bone cyst of the orbit: a case report and review of literature

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Abstract

Aneurysmal bone cyst is a benign fibro-osseous lesion which rarely occurs in the orbit. We report on a 7-year-old girl with aneurysmal bone cyst of the orbit who presented with painless proptosis and diplopia. Optic nerve compression resulted in field loss and delayed visual evoked potentials. Radiological and histological features are discussed. The lesion was excised via a frontal craniotomy and the orbital roof reconstructed with a prefabricated titanium plate. Post-operatively a rapid resolution of the proptosis and diplopia followed. Previous reported cases of this rare entity in the orbit are also reviewed.

Key words Aneurysmal bone cyst, Orbital, Optic nerve compression, Proptosis, Prefabricated titanium plate

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Aneurysmal bone cyst (ABC) is a rare non-neoplastic lesion of unknown aetiology. The metaphyses of long bones (60%) and the vertebrae (20%) are the most frequent sites for this tumour.[1] Lesions arising in the orbit represent less than 0.25% of reported cases.[2] In a third of cases ABC is associated with an underlying bone disorder. It is usually found in the first two decades of life with some authors reporting a female preponderance.[3,4] Since 1953 when the first report of orbital aneurysmal bone cyst appeared in the literature, 21 cases have been described.

Case report

A 7-year-old girl was referred to the eye department with a 2 week history of right painless proptosis and diplopia in upgaze. Her parents reported a minor accident 2 weeks prior to the appearance of the proptosis when she fell off her bicycle and hit the right side of her head. On examination her vision was 6/5 in the right eye and 6/4 in the left with normal pupillary reactions. A 3 mm right proptosis (Fig. 1) was present with restriction of upgaze in the right eye (Fig. 2). Pupillary reactions were normal and both optic discs were healthy. No retinal folds were seen. Goldman test showed constriction of visual fields in the right eye. Colour vision was normal when tested with Ishihara colour plates. CT and MR scans showed a complex multiloculated mass (Figs. 4, 5) in the roof of the right orbit displacing the superior rectus on to the optic nerve and compressing the superior oblique. The medial orbital wall was deficient at this site. The mass contained posterior solid enhancing components and a larger cystic component anteriorly with what appeared to be blood degradation products. A clearly defined fluid level on T2- and T1-weighted MR images was seen.

A month later the patient presented to the eye clinic complaining of blurred vision in her right eye. The visual acuity was 6/6 in the right, 6/4 in the left. A subtle relative afferent pupillary defect was present. Proptosis as measured with Hertel's exophthalmometer had increased to 5 mm. The right optic disc appeared swollen. Goldman fields showed an enlarged blind spot on the right side in addition to the previously noted constricted field. Visual evoked potential (VEP) demonstrated delayed responses from the right nasal retina suggesting

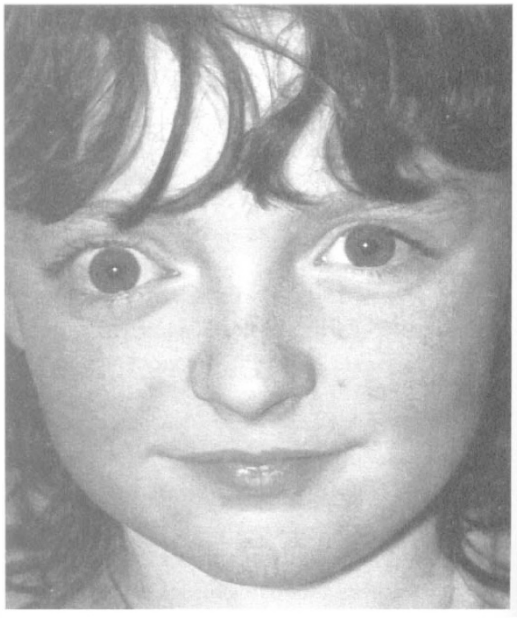

Fig. 1. *Photograph showing right proptosis.*

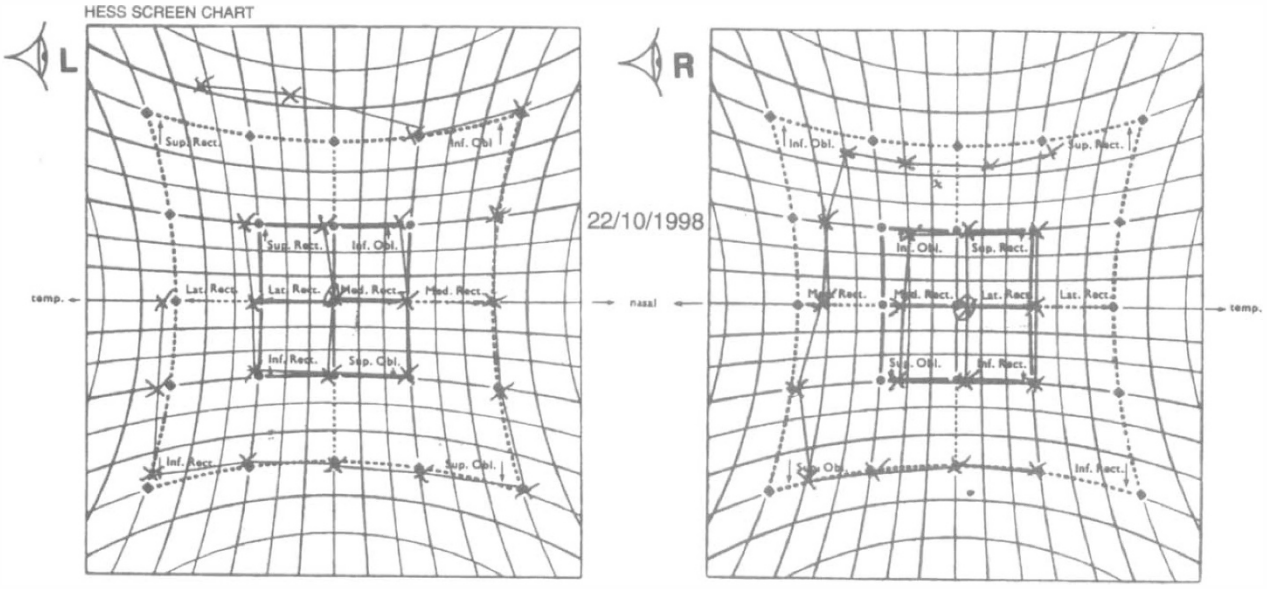

Fig. 2. *Pre-operative Hess chart. There is restriction of upgaze.*

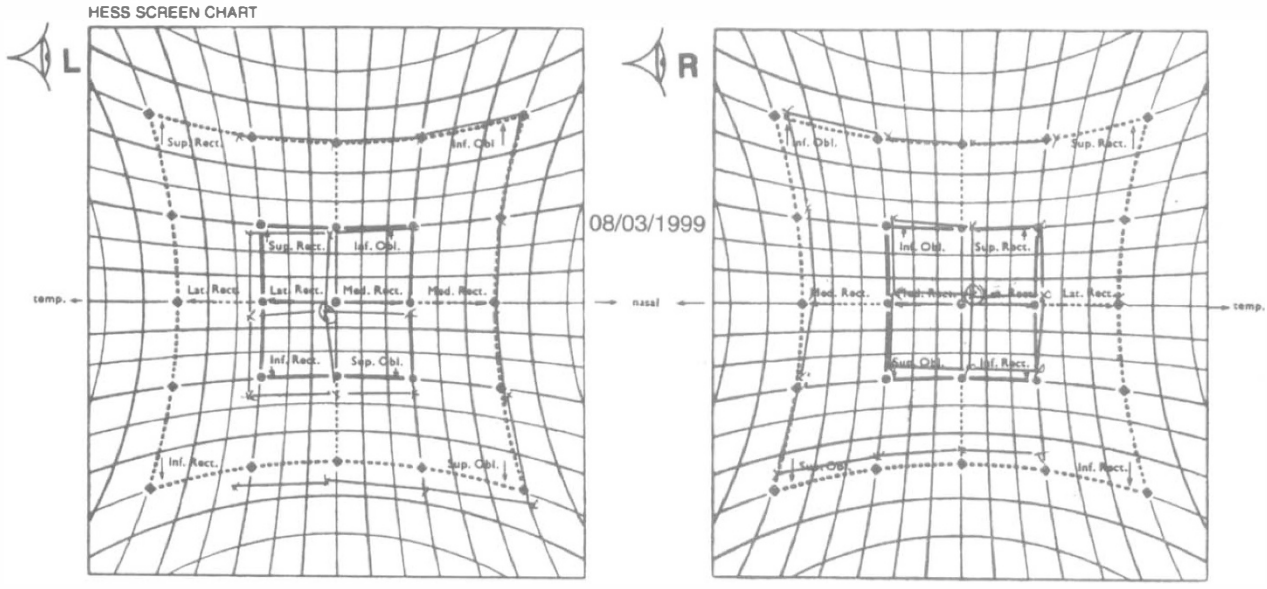

Fig. 3. *Post-operative Hess chart.*

optic nerve compression. In light of the evolving picture of optic nerve compression, she underwent a frontal craniotomy to decompress the orbital roof lesion. Surgical findings were of a multicompartmental cystic mass that had eroded through the orbital roof. The mass was dissected free from the dura and orbital fascia and total macroscopic removal was obtained. The orbital roof was reconstructed with a purpose-made titanium plate (Fig. 6) modelled from three-dimensional reconstructions of the pre-operative CT scan.

Multiple fragments of haemorrhagic and membranous tissue were obtained on excision of the lesion. Histopathological examination revealed thin vascular spaces of varying sizes surrounded by multinucleate giant cells and cellular fibrous tissue containing some bony trabeculae. This appearance confirmed the diagnosis of an aneurysmal bone cyst. No secondary lesion was found.

On the third post-operative day the patient's vision was recorded as 6/6 in each eye with normal pupillary reactions. The patient reported a subjective improvement in the vision as well as diplopia. No proptosis was detected and extraocular movements appeared full on testing. The right optic nerve swelling had resolved. Goldman fields test performed at 1 and 3 months post-operatively demonstrated progressive improvement. A Hess chart recorded 3 months post-operatively showed complete restoration of her ocular movements (Fig. 3). A repeat VEP, however, remained unchanged. The patient is being kept under review in the eye clinic as this lesion is known to have a recurrence rate of 10–33%.

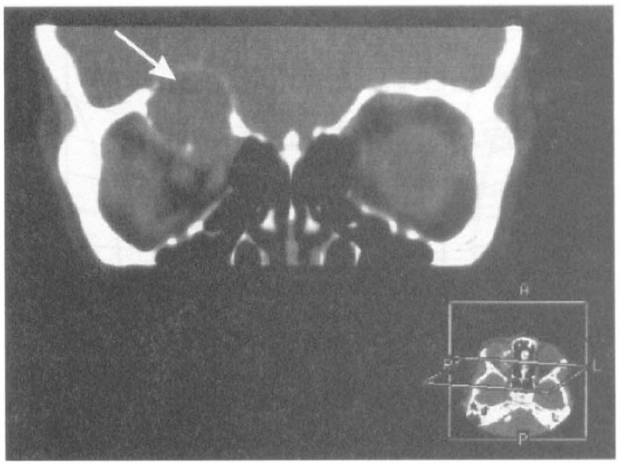

Fig. 4. *CT scan indicating a cystic lesion in the orbital roof (arrow).*

Discussion

ABC is a rare benign lesion of the bone arising most frequently in the metadiaphysis of long bones (e.g. humerus, femur, tibia), small tubular bones (e.g. clavicle, phalanges) and vertebrae.[1] ABC arising in the orbit has been described in less than 0.25% of cases. The lesion expands rapidly and destroys the surrounding bone.

Orbital aneurysmal bone cyst commonly presents with painless proptosis which progresses rapidly. Other signs include diplopia, ptosis, reduced vision, visual field changes, nasal congestion and headache. Visual field changes seen in our case were an enlarged blind spot and field constriction. A central scotoma secondary to orbital ABC was documented by Yee *et al.*[16] in a 10-year-old boy (Table 1, no. 10). Other features of optic nerve compression, i.e. mild afferent pupillary defect and delayed VEP, as seen in our patient have not been reported previously. The progression of these signs indicated an increase in the size of the lesion, prompting surgical intervention. Where electrophysiological studies are available, VEP is a reliable and sensitive test of optic nerve function. As the lesion expands optic disc swelling and retinal folds[24] may also be observed.

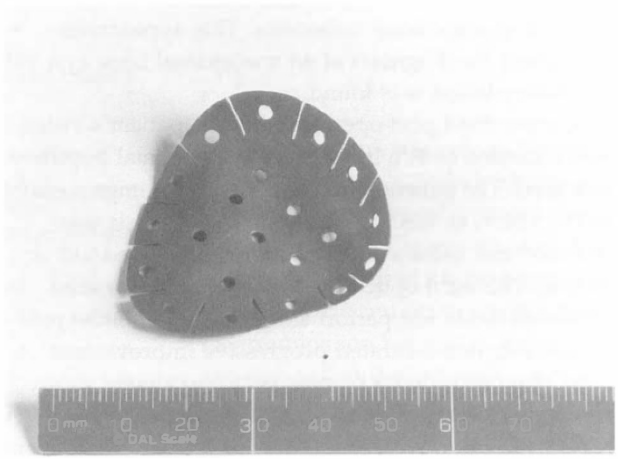

Fig. 6. *Prefabricated titanium plate used to repair the orbital roof defect.*

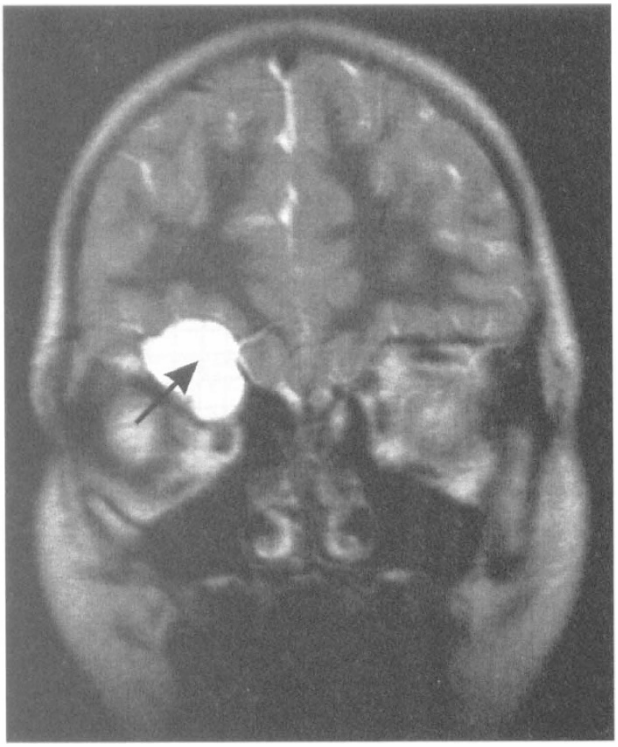

Fig. 5. *MR scan following gadolinium administration showing enhancement of the lesion (arrow).*

A history of trauma, often trivial, is present in nearly half the ABCs occurring at extracranial sites. This association is less frequent with orbital ABC, as ours is only the second case reported with a preceding history of trauma. The roof of the orbit is the most frequent site for an orbital ABC (Table 1). Presentation is most common in the first two decades.[26]

Several hypotheses have been put forth to explain the aetiology of ABC. Jaffe[27] and Lichtenstein[28] postulated that a local circulatory disturbance such as a thrombosis or an arteriovenous malformation in the bone leads to markedly increased venous pressure and development of a dilated and engorged vascular bed within the affected area. Thompson[29] and Dabezies[30] studied cases of ABC secondary to trauma and concluded that it represented an osseous manifestation of a post-traumatic arteriovenous fistula.

Extracranial ABCs are associated with secondary lesions in a third of cases. The associations reported include fibrous dysplasia,[24,31] von Recklinghausen's diesase, osteosarcoma, non-osteogenic fibroma, osteoclastoma, osteoblastoma, haemangioendothelioma and haemangioma of the bone.[32] It is therefore vital that an evaluation of any aneurysmal bone cyst must include a careful examination to rule out any associated lesion. Beisecker *et al.*[3] suggested that an osseous arteriovenous malformation initiated by a primary bone lesion creates haemodynamic disturbances giving rise to a second bony lesion. This hypothesis is supported by manometric studies demonstrating elevated pressures within these lesions. The expansile haemodynamics of the

Table 1. *Reported cases of orbital aneurysmal bone cysts*

Case no.	Reference	Age and Sex	Site	Proptosis	VA	Diplopia	Other features	Trauma	Secondary lesion
1	Siedenbiedl *et al.*[7]	22M	Roof	+	N	+	–		–
2	Arnould *et al.*[8]	31M	Roof	+	N	–	–	–	–
3	Kubicz *et al.*[9]	8M	Medial	+	NA	–	–	–	–
4	Constantini *et al.*[10]	14F	Roof	–	NA	–	Headache	–	–
5	Fite *et al.*[11]	8F	Roof	+	N	–	–	–	–
6	Offret *et al.*[12]	10F	Roof	+	N	+	–	–	–
7	Komorn[13]	26F	Medial	+	NA	+	–	–	–
8	Powell *et al.*[14]	16F	Lateral	+	N	+	–	–	–
9	O'Gorman *et al.*[15]	1M	Roof	+	NA	NA	–	–	–
10	Yee *et al.*[16]	10M	Sphenoid & ethmoid sinus	–	Red	–	Central scotoma	–	–
11	Flament *et al.*[17]	11F	Roof	+	NA	NA	–	–	–
12	Iraci *et al.*[18]	1M	Roof	+	N	+	–	–	–
13	Ronner *et al.*[1]	12F	Roof	+	Red	–	–	–	–
14	Calliauw *et al.*[19]	19F	Lateral	+	N	+	Pain	–	–
15	Johnson *et al.*[20]	42F	Roof	+	N	+	Ptosis; headache	–	–
16	Hunter *et al.*[2]	7M	Roof	+	Red	–	–	–	–
17	Patel *et al.*[21]	11F	Medial	+	NA	+	EOM restriction; medial canthal mass; epistaxis	–	–
18	Bealer *et al.*[22]	2F	Lateral	–	NA	–	Infraorbital, swelling and temporal mass	–	–
19	Dailey *et al.*[23]	43M	Zygoma	–	–	–	Haematuria	–	Renal Ca
20	Lucarelli *et al.*[24]	19M	Zygoma, sphenoid and roof	–	Red	+	EOM restriction; ptosis; optic nerve compression; retinal folds	+	Fibrous dysplasia
21	Hino *et al.*[25]	1M	Roof	+	NA	NA	–	–	–
22	Menon *et al.* (this study)	7F	Roof	+	Red	+	Signs of optic nerve compression	+	–

VA, visual acuity; +, present; –, absent; N, normal; NA, not available; Red, reduced; EOM, extraocular movements; Ca, carcinoma.

arteriovenous malformation create bone erosion and resorption producing the vascular channels lined by bony trabeculae that are thought to be typical of ABC.

Histological features are consistent with a reactive proliferation of bony tissues characterised by a fibrous matrix of spindle-shaped cells with a variable amount of extracellular collagen formation. Within the fibrous matrix small and large channels are found, some of which are filled with blood. Giant cells, haemosiderin-laden histiocytes and areas of osteoid formation are seen within the stroma.[16]

Radiological appearances of ABC are suggestive but not diagnostic of the entity. It is seen as a osteolytic lesion with bone enlargement, cortical thinning, geographic destruction, well-defined contours, no periosteal reaction and no visible matrix.[33] Fine trabecular images forming incomplete partitions may occasionally be identified. Four radiological phases – osteolysis, rapid growth, stabilisation and healing with progressive ossification – were described by Dabska and Buraczewski.[31] It is thought that most patients present during the phase of rapid growth.

CT scans show a multilocular mass confined by a peripheral cortical layer. Infusion of contrast enhances the septa between the blood lakes. A characteristic feature of ABC is the appearance of fluid–fluid levels, which may be demonstrated on CT, MRI and ultrasound. The levels correspond to the varying densities of the supernatant fluid and sediment secondary to haemorrhage within the bony cavities. These levels are best observed by keeping the patient immobile for 3–10 min and then changing the position to confirm the shifting fluid levels. The average attenuation of the sediment is 60 Hounsfield units (range 38–90 HU) while that of the supernatant fluid is 30 HU (range 9–62 HU). The difference in the relaxation time of the degradation byproducts of haemoglobin enable fluid levels to be seen clearly on T2-weighted MR images.[35] Ultrasound can detect these levels in a plane perpendicular to the sonic beam.[36] It is an inexpensive, safe and reliable method, particularly useful in young children who may otherwise require sedation or anaesthesia for CT or MRI.[36]

Various treatment modalities have been used in the management of extracranial ABCs. These include curettage with or without bone graft, surgical resection, irradiation and cryotherapy. Irradiation carries with it the potential risk of osteosarcoma.[37] Orbital ABCs have largely been treated by surgical resection alone, although radiotherapy (Table 1, nos. 11 and 12), and systemic steroids (Table 1, no. 10) have been used as adjuvant treatment. Bone grafts have been used in previous cases to repair the bony defects resulting from surgical resection. In our patient the defect was repaired using a prefabricated titanium plate. The titanium plate was fashioned with the help of pre-operative CT scans.

The post-operative period was marked by rapid resolution of diplopia and subjective improvement in the visual acuity. At last follow-up, the visual field recorded a steady improvement but the VEP continued to show a delayed amplitude from the nasal retina. A similar pattern of recovery was documented by Yee *et al.*[16] and Lucarelli *et al*.[24]

A recurrence rate of 10–30% has been reported most of which have been detected within 2 years of the initial treatment.[5] This underlines the need to review these patients at regular intervals playing careful attention to the optic nerve function and ocular motility.

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