Unusual Aneurysmal Bone Cyst

A Case Report

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In 1942 Jaffe and Lichtenstein,11 closely examining two cases, consolidated a previously confused lesion of bone into an established benign entity of non-neoplastic disease. Later, Lichtenstein12–14 offered sound justification for this “new” entity and identified it beyond doubt by its distinct roentgen-ray appearance, its benign clinical behaviour and its peculiar histologic pattern. They called it “aneurysmal bone cyst”. Since Jaffe and Lichtenstein clarified this tumour numerous reports have appeared emphasising its prevalence in the second decade and its preferential location in the long bones and the spine, although virtually any bone may be involved. Location of the lesion in the calvarium has not been prominent.

We have encountered, in a Western Nigerian child, a case of chronic increased intracranial pressure caused by aneurysmal bone cyst of an unusually large size and in an uncommon site.

Case Report

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Clinical History. On Nov. 2, 1962 a formidable-looking occipital mass was presented at the Neurosurgery clinic of the University College Hospital, Ibadan. K.A., a 64-year-old right-handed Yoruba boy, noted a small mass in the cervico-occipital region some 4 months previously. He complained of fever, severe occipital headaches, and had transient vomiting. The mass rapidly increased in size and, simultaneously, his headaches improved for a while but recurred intermittently with dysarthria. There was no significant antecedent trauma to the head.

Examination. Vital signs were within normal limits. An initial temperature of 101.8°F. spontaneously subsided unexplained. No mass was seen or felt along the long bones or spine. A huge and hard nontender tumour deformed the cervico-occipital area, completely immobilizing the patient’s head and neck in a fixed torticollic attitude (Fig. 1). It measured 32 cm. horizontally, 21 cm. vertically and 8 cm. in its anteroposterior extent. The head itself measured 54 cm. in circumference, and Macawen’s sign was positive. No bruit could be elicited over the head or the lesion. Detailed neurologic evaluation revealed only engorged retinal veins with loss of spontaneous pulsations bilaterally, mild dysarthria with nasal-tone quality, and generalized deep tendon hyporeflexia. Sensorium was clear and cerebellar signs were absent.

Laboratory Studies. Hemogram and blood-chemistry profile were essentially normal (prothrombin index was 100 per cent). Ventricular cerebrospinal fluid gave an unreliably high protein of 250 mg. per cent.

Radiologic Findings. Plain films of the skull showed diastasis of coronal and sagittal suture. There was a large occipital bony tumour with well defined edges of thin bony shell and a multifoculated radiolucent “soap-bubble” pattern showing bony septa. The occipital-suboccipital segment of skull involved had been destroyed and replaced by a markedly indented wall of bone, ballooned intracranially at the anterior superior border of the tumour (Fig. 2A). Tomograms showed intact spinous processes and laminae of the cervical vertebrae. The outlines of the tumour extended down to the level of C4 vertebral body. In the vertebral angiograms (right femoral Seldinger technique of catheterisation) the posterior cerebral arteries were elevated significantly (Fig. 2B). On ventriculography (left Keen’s point trephine) the lateral and 3rd ventricles were found to be symmetrical but dilated (Fig. 2A), and upward displacement of the 4th ventricle was noted on brow-down view.

Operation. On Nov. 30, 1962, under endotracheal general anaesthesia and electrocardiographic monitor, the patient was placed in the prone cerebellar position and the lateral ventricle was tapped for decompression of the brain. A midline skin incision was made. On removing the thickened pericranium, the surface of the bony tumour was found to be indented in a spotty fashion by small areas of defects through some of which fragile greyish-black walls of cysts protruded. Nearly all of the mass was rongeured off in large and small fragments. The multifoculated tumour contained many large cavities lined by yellowish-grey membrane, 0.5 to 2 mm. thick. They contained either chocolate-coloured or yellowish turbid (motor oil-like) liquid or, as in the majority of them, dark sanguineous liquid constantly weeping into slow pools of blood. Maintaining adequate
hemostasis was a bit tedious; but reasonably effective control was secured from stage to stage chiefly with Surgicel and cotton-ball packing of the cavities. The ballooned shell of bone delineating the tumour anteriorly formed the suboccipital portion of the skull, indented inward, apparently exerting some chronic pressure on the brain contents of the posterior fossa. The posterior rim of the foramen magnum was nearly paper-thin. Before the indented suboccipital shell could be removed for desired decompression of the posterior fossa, the patient experienced a cardiac arrest. All resuscitative measures were without avail. A coroner's autopsy was obtained.

Pathologic Findings. Gross Appearance. The largest of the resected specimens removed at operation consisted typically of a bony mass, approximately 13×8×5 cm. (Fig. 3). The outer surface generally was smooth but some of several small translucent areas present bulged slightly. The cut surfaces were ragged and showed numerous dilated spaces of varying sizes and shapes, sometimes communicating with one another, but often separated by bony or fibrous septa. Most of these spaces contained clots of blood or greenish gelatinous material. Some were empty when received. Histologic Sections. Hematoxylin and eosin and van Gieson stains were used (Figs. 4, 5 and 6). There were numerous vascular spaces containing blood cells. When not in close proximity they were widely separated by fibrous connective tissue of variable cellularity. The walls of some spaces were lined by a single layer of flattened cells; others were devoid of lining cells. No elastin or muscle fibres were demonstrated in the dense supportive connective tissue adjacent to the blood-filled spaces. Multinucleated giant cells were prominent in some parts of the connective tissue. In haemorrhagic areas an occasional giant cell was seen amongst the extravasated red corpuscles. Iron-pigment granules were abundant, lying free in the connective tissue or engulfed within macrophages. Osteoid or formation of new bony tissue was evident in some septa.

Comment

Essential among the numerous reports on aneurysmal bone cyst are those of Jaffe and Lichtenstein,11 Lichtenstein,12–14 Jaffe,15 Dahlin et al.,9 Besse et al.,1 Cruz and Coley,6 and Dahlin.7 Recently Subramaniam and Mathias18 summarized experiences with this lesion in India.

It is unnecessary to recount extensively that the lesion now defined as aneurysmal bone cyst historically bore a less specific variety of names. Van Arsdale's18 and Cone's15 ossifying haematoma, Bloodgood's3 benign bone cysts and aneurysm of the long pipe bones, some of Coley's4 atypical giant-cell tumours and Ewing's9 aneurysmal giant-cell tumour are a few among the lot. Thompson,17 airing the residuum of the previous years, remained undecided with “a threefold title” of subperiosteal giant-cell tumour, ossifying subperiosteal haematoma and aneurysmal bone cyst. Taylor,16 in Britain, preferred the name
Fig. 4. Blood-filled spaces and cellular connective tissue in which area of haemorrhage, osteoid and giant-cell formation can be seen. Haematoxylin and eosin, ×120.

Fig. 5. Inset of Fig. 4 to show multinucleated giant cells, osteoid tissue and part of a vascular space below. Haematoxylin and eosin, ×320.
haemangiomaticus bone cyst (as a subtype of cavernous haemangioma).

We chose to evaluate the present tumour primarily in the light of Lichtenstein's simple criteria laid down in his 1950 differential consideration. The case being reported here is characteristically "a solitary, localized, and expanded fibrous lesion honey-combed by enormously dilated, plexiform vascular bed". It is circumscribed by "a thin shell of periosteal new bone." The "soap-bubble" radiologic appearance is obvious, and at operation the aneurysmal spaces "welled-up" disturbingly with blood. In the histologic examination giant cells are seen. There is no implication of trauma in the spontaneous and rapid development of the painful mass. The patient's age in the first decade fits into the established accounts of this tumour. The location in a flatbone of the calvarium is relatively uncommon although the occipital site has been encountered in single cases as exemplified by Lichtenstein and Besse et al. In the post-traumatic (4 month) case of Bhende and Kothare the lesion was present in the temporal bone in a 9-year-old boy, without increased intracranial pressure. The overwhelming majority of the cysts reported are outside the skull, being chiefly in the long bones and the spine. As in the spinal location, mechanical compression exerted upon the neural contents is of clinical significance in the present case in which partial obstruction of the 4th ventricle led to appreciable internal hydrocephalus with diastasis of sutures. The patient's demise at operation is indeed regrettable, particularly in the light of the benign nature of his disease, as emphasised by Lichtenstein. It was felt that simple curettage alone would be inadequate. The sub-occipital bony shell must be removed to decompress the posterior fossa and, in addition, radiotherapy is presently not available in our hospital to supplement a partial excision. Loss of blood was not to appear to be a problem since replacement was measured to be adequate. The possibility of a "sickle-cell" complication could not be substantiated or denied.

**Summary**

A case is presented of an occipital aneurysmal bone cyst in a 6½-year-old Western Nigerian boy. The large size, its unusual location and the rapidity of the growth are noted in addition to the extrinsic compression of the brain which resulted in appreciable internal hydrocephalus. The tumour in all its aspects satisfies Lichtenstein's criteria for its diagnosis.

We are grateful to Dr. A. B. Tompkins in whose Paediatric Service the patient first appeared at the University College Hospital, Ibadan; and also to Professor G. M. Edington for his experienced histologic diagnosis of the lesion.

**References**


