Angiography and computerized tomography in the diagnosis of aneurysmal bone cyst of the skull

Case report

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An aneurysmal bone cyst of the occipital bone, presenting as an intracranial space-occupying lesion, is reported. Clinical and neuroradiological findings are described in detail. The significance of angiographic and computerized tomographic findings is discussed.

KEY WORDS □ aneurysmal bone cyst □ benign skull tumor □ computerized tomography □ arteriography

ACCORDING to Jaffe and Lichtenstein, aneurysmal bone cysts are non-neoplastic benign lesions, with obscure pathogenesis. In 36% to 51% of cases, these bone cysts are found on the long tubular bones of the extremities, and in 20% to 26% they are associated with the vertebrae. They are rarely found in the skull (3% to 6% of cases), and only 22 cases have been reported to date. We have observed an additional case of aneurysmal bone cyst of the occipital bone, and, to our knowledge, this is the first case studied by cranial computerized tomography (CT).

Case Report

This 19-year-old woman was admitted to our institute in June, 1978. She denied any significant head trauma, but had noted, some 5 months previously, an occipital swelling that had rapidly enlarged during the last 2 months, associated with pulsating headache and vertigo that was increased by rapid movements of the head. During the last week before admission, she had suffered from nausea, vomiting, and double vision.

Examination. She had a marked swelling in the left occipital region, near the midline. The mass was hemispheric in shape, measuring 5 cm in diameter and protruding 2 cm from her head. It was firm, nonfluctuating, and painful to pressure, and was apparently attached to the occipital bone. The skin over it was normal. On neurological examination, mild bilateral papilledema and right sixth nerve palsy were found. She had an ataxic gait with a tendency to fall to the right.

Routine laboratory investigations were normal. Skull films revealed sellar changes consistent with increased intracranial pressure, and a wide area of radiolucency with fairly sharp margins located in the left portion of the occipital bone near the midline. Lateral and oblique views demonstrated that the lesion was delimited by an extremely thin, almost complete shell of bone, bulging both intracranially and extracranially (Fig. 1). Some osseous septa were seen within the lesion.

Computerized tomography confirmed the presence of a multiloculated lesion originating in the diploe of the left part of the occipital bone and expanding both intracranially and extracranially. The contents of the cavities within the lesion had different densities. After injection of contrast medium, the contents of the cavities with high density enhanced strongly, but that of the cavities with low density remained unchanged. The fourth ventricle was displaced anteriorly and contralaterally, and minimal enlargement of the third and
lateral ventricles was present (Fig. 2). A selective transfemoral angiographic study of the left vertebral artery and of both external carotid arteries was performed. Left vertebral angiography demonstrated displacement of several arteries of the posterior fossa, mainly of the vermian and hemispheric branches of the left posterior inferior cerebellar artery. These findings were consistent with an extra-axial lesion displacing the left cerebellar hemisphere upward and forward. Also, the left posterior meningeal artery was enlarged and supplied a faint abnormal circulation.

A left external carotid angiogram, with selective injection of the occipital artery, showed that this vessel supplied the extracranial portion of the lesion, with visualization persisting in the venous phase (Fig. 3). A minimal contribution to this pathological circulation from the right occipital artery was also observed.

**Operation.** On June 26, 1978, the lesion was exposed by a horseshoe occipital skin flap, and the whole lesion was removed en bloc, together with a narrow margin of the surrounding normal bone. Some detached fragments of the inner shell of bone, which were tightly adherent to the dura, were subsequently removed. The dura underlying the lesion re-expanded promptly as the lesion was removed. Intraoperative bleeding was minimal. The bone defect was not repaired.

**Pathological Examination.** The lesion was 5 cm in diameter. It involved the diploë of the occipital bone, and was delimited by a thin shell of bone that was continuous with the inner and outer tables of the surrounding normal bone. The cut surfaces of the surgical specimen showed a honeycomb of blood-filled spaces of different sizes separated by osseous septa.

Microscopic examination of the tissue from the central portion of the lesion disclosed several blood-filled channels of different sizes, bordered only by a thin layer of spindle-shaped endothelial-like cells with no elastica or smooth-muscle wall. The vascular spaces were surrounded by connective tissue, containing numerous multinucleated giant cells (Fig. 4).

**Postoperative Course.** The postoperative course was uneventful. Cerebellar signs, right sixth nerve palsy, and papilledema disappeared within 10 days. On consultation 1 year later, the patient was free of symptoms, and there was no evidence of recurrence.

**Discussion**

Only 23 cases of aneurysmal bone cyst of the skull have been reported, including this case. The location of the lesion was indicated in 17 of these. The occipital bone was involved in the present case and in five others,\(^5,13,18,19,23\) the frontal bone in five cases,\(^3,4,6,7,17,23\) the temporal bone in four cases,\(^1,6,7,12\) and the parietal bone in two cases.\(^3,6\)
Aneurysmal bone cyst of skull

Fig. 3. Upper Left: Left vertebral angiogram, lateral view, arterial phase. Arrowheads indicate the enlarged posterior meningeal artery, displaced forward. Upper Right: Left vertebral angiogram, late arterial-capillary phase. Arrowheads indicate the most lateral of the hemispheric branches of the left posterior inferior cerebellar artery, displaced forward and upward like the inferior vermian branch. Posteriorly, there are faint patchy areas of abnormal vascularity, possibly originating from the posterior meningeal artery. Lower: Selective injection of the left occipital artery, lateral view. Pathological circulation in the extracranial part of the aneurysmal bone cyst is indicated by arrows.

When considering only the 13 cases described in detail, it can be seen that aneurysmal bone cysts of the skull, like those located elsewhere in the skeleton, tend to affect patients less than 20 years old (nine cases), with no significant predilection for either sex. The duration of the clinical complaints is short, mostly less than 6 months (9 cases).

Aneurysmal bone cysts of the skull, like those affecting the flat bones, such as rib, clavicle, and scapula, show symmetrical expansion, involving both the inner and outer tables of the skull, and, therefore, always involve intracranial expansion. However, focal neurological deficits have been found in only five cases of 13; signs of intracranial hypertension were found in four cases of 13 (in three cases with an occipital lesion and in one patient with a frontal lesion). The diagnosis of aneurysmal bone cyst is based upon the roentgenographic and pathological findings that are considered almost typical, although bone tumors with cystic components, such as some osteogenic sarcomas and osteoblastomas, and most benign giant-cell tumors, should be included in the differential diagnosis.

As far as we know, the present case is the first to be studied by CT. Morphological density patterns, both before and after contrast medium injection, demonstrated very well the gross pathological architecture of the lesion. We think, therefore, that CT scanning is highly reliable for preoperative diagnosis of these lesions.

Angiography of aneurysmal bone cysts located anywhere in the skeleton is usually characterized by the demonstration of a pathological circulation with a patchy distribution, persistent in the venous phase, and occasionally with some arteriovenous shunts. It is interesting to note, however, that, when the lesion is located in the skull, demonstration of the pathological circulation is fairly uncommon. In four out of seven cases studied with angiography, the lesion was completely avascular. The angiographic
findings in our case are consistent with those most frequently described. 16,20 In all the cases reported, a full recovery was obtained by complete removal of the cyst. Either piecemeal or en bloc removal can be performed, but in our opinion the latter is preferred whenever possible, to reduce intraoperative bleeding.

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References


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Fig. 4. Photomicrograph of the central portion of the lesion showing a blood-filled channel, bordered by a thin layer of spindle-shaped endothelial-like cells, without elastica or smooth-muscle wall. Multinucleated giant cells are concentrated in the surrounding connective tissue. H & E, × 160.