Intraosseous cystic cavernous angioma with occipital skull osteolysis

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ABSTRACT

Intraosseous cavernous angiomas (CAs) of the skull are rare, and those cases that appear are commonly localized in the frontal bone. Computed tomography (CT) and Magnetic resonance imaging (MRI) typically show a well-defined intradiploic lytic mass with homogeneous enhancement. We describe an intraosseous cystic CA of the occipital skull in a 46-year-old man who presented with transient right-sided deafness and posterior cervical pain. MRI revealed a large (3.7 cm × 3.2 cm × 4.1 cm) extra-axial tumor, compressing the right cerebellar hemisphere, with heterogeneous peripheral enhancement. A CT scan showed osteolytic change of the occipital skull. The tumor was totally resected via a suboccipital approach. Intraoperatively, we found a mainly cystic tumor containing xanthochromic fluid. Histologically, the tumor was diagnosed as a cavernous angioma. This is the first reported case of an intraosseous CA of the skull with cyst formation. The characteristic radiological imaging of the presented case mimicked a malignant tumor with peripheral enhancement and prominent osteolytic change.

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Introduction

Intraosseous cavernous angioma (CA) of the skull is a rare and benign vascular tumor. CA usually appears in the frontal bone, and rarely in the occipital bone [1]. Typical radiological findings show a well-defined intradiploic lytic mass with homogeneous enhancement. Intraosseous CA of the skull with prominent osteolysis is extremely rare, as the tumor usually grows slowly, with intradiploic expansion [1]. Some investigators have reported CAs with cyst formation, but none before now have described intraosseous CAs with cysts [2].

We present a case of CA of the occipital skull with cyst formation and osteolysis. A neuro-radiological study showed an extra-axial tumor with heterogeneous peripheral enhancement and prominent osteolytic change, that is, the same findings typically manifested by a malignant tumor.

Case report

A 42-year-old man with no previous medical history presented with right-sided transit deafness and a 3-month history of posterior cervical pain. His neurological examination revealed nothing abnormal. Computed tomography (CT) showed an extra-axial, slightly high-dense mass compressing the right cerebellar hemisphere, with osteolytic changes of the occipital bone and the posterior part of the mastoid cells (Fig. 1A, B). Magnetic resonance imaging (MRI) demonstrated a large (3.7 cm × 3.2 cm × 4.1 cm) extra-axial mass. The lesion was rendered with iso-signal intensity on T1-weighted images, hyper-signal intensity on T2-weighted images, and slightly hyper-signal intensity on diffusion-weighted images, with heterogeneous peripheral enhancement (Fig. 1C–F). Cerebral angiography revealed occlusion of the right sigmoid sinus and no abnormal vascular staining. No uptake of 18 F-FDG was found anywhere in the body on 18 F-fluorodeoxyglucose positron emission tomography combined with CT (FDG-PET/CT). Our first diagnostic guess was a skull bone tumor such as a giant cell tumor, osteoma, chordoma, dermoid cyst, aneurysmal bone cyst, fibrous dysplasia, or osteolytic intraosseous meningioma.

Surgery and post-operative course

The tumor was surgically resected via a right suboccipital approach for a definitive diagnosis. A bone defect was identified during dissection of a deep layer of suboccipital muscles, and a xanthochromic fluid spilled out. The lesion was cystic, with portions of the septal wall containing a xanthochromic fluid. The cyst and septal wall were both composed of pinkish soft tissue with some vascularity (Fig. 2A). The tumor was located in the extradural space and the cyst wall adhering to the dura mater was sharply dissected with clear margins. The dural attachment was intact. Some parts of the mastoid air sinus were opened during the dissection. The tumor was totally resected (Fig. 2B). Autologous fat grafts were placed in the skull base defect to close the opened mastoid air sinus.

The patient complained of right-sided conductive hearing loss postoperatively because of fluid collection in the opened mastoid air sinus, but recovered completely after 1 week. Postoperative CT and MRI confirmed total tumor removal, and no evidence of recurrence was found at a 1 year follow-up.

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Histopathology

Histopathological examination revealed compact vessels that varied considerably in caliber and were composed of a lined single layer of endothelial cells without muscle or elastic fibers. Few interposed bony trabeculae were noted. Immunohistochemical studies revealed a positive reaction for CD31 marker in lined vascular endothelium (Fig. 3A, B). The surgical specimen was histopathologically diagnosed as a cavernous angioma of the occipital skull.

Discussion

Intraosseous CA is a rare and benign vascular tumor usually found in the vertebra, and sometimes in the skull. CAs reportedly account for only 0.7% of all osseous tumors, while intraosseous CAs of the skull are rarer still, accounting for only 0.2%. The frontal bone was the most common site of involvement among 125 intraosseous CAs of the skull reviewed by Heckl et al., followed by the temporal and parietal bone [1]. The occipital bone is very rarely affected.
Intraosseous CA of the skull usually grows slowly, with expansion of the outer table [1]. Intracranial expansion and destruction of the internal table always lag far behind the changes in the external table, which leaves little time for the development of neurological symptoms or signs of mass effect in most cases. Intraosseous CA of the skull has a variable clinical presentation predominantly based on the tumor location. The tumor in the present case was a relatively large cystic CA accompanied by prominent osteolysis. The biological behavior and growth mechanism of the tumor may have differed from that of a general intraosseous CA of the skull. We hypothesize that the slow growth of the cyst prolongs the time required for an intraosseous CA to destroy all layers of the skull. Tumors of this type are also likely to remain asymptomatic for long periods because of their anatomical distribution.

The radiological characteristics of intraosseous CAs of the skull have been well described. CTs reveal an intradiploic lytic mass with rarefaction and a honeycomb or polka-dotted pattern [1]. CT in bone window is useful in planning surgical resection, as it depicts both the site and extent of the region. The signal characteristics on MRI are variable. Intraosseous CAs generally have a heterogeneous signal in both T1- and T2-weighted images, because the signal intensity depends on the quantity of slow-moving venous blood and the ratio of red marrow to fatty marrow within the lesion [1,3]. The lesion is typically enhanced after an administration of gadolinium [1].

The imaging and clinical characteristics of the present case differ from the typical findings of intraosseous CAs of the skull in two important ways. First, MRI revealed an extra-axial mass with heterogeneous peripheral enhancement. The CA was a cystic lesion containing xanthochromic fluids, and the peripheral enhancement was intraoperatively confirmed to be the wall of the cyst. Second, the lesion manifested prominent accompanying osteolytic change on CT. This CA was difficult to diagnose appropriately without histopathological findings, as the radiological findings mimicked the features typically manifested by malignancies such as osteoma, osteosarcoma, giant cell tumors, and metastatic tumors.

The biological progression of cerebral CA is well recognized. The tumor growth has been divided into four stages: 1) progressive ectasia of the vascular channels; 2) thrombosis of the contiguous vascular channels with fibrosis; 3) connective organization of collected peripheral blood; 4) peripheral cyst formation resulting from internal hemorrhage [4]. CA growth is also reported to be perpetuated by the tumor’s own proliferative mechanism [5]. We speculated that the cyst formation predominately explained the larger size and osteolytic feature of the present CA compared to the general size of previously reported cases of intraosseous CA. Cerebral CAs with cyst formation have been reported on several occasions [2]. Cystic CAs are thought to form and grow via histological mechanisms similar to those of chronic subdural hematomas [2]. In our case, we believe that the formation and growth of the cystic CA were induced by repeated hemorrhage from the sinusoids of the CA from the neocapillaries of the cyst wall. The cyst walls of CAs sometimes show radiographic enhancement due to neo-angiogenesis [2]. The present case was the first reported intraosseous CA of the skull with cyst formation, in addition, the cyst wall was enhanced on MRI. The existence of hemorrhage from sinusoids or neocapillaries of the cyst wall might be visible in histologic specimens, especially in Prussian-blue-staining, but the histopathological findings of this case, without iron staining, could not prove the existence of repeated intra-lesional hemorrhage. However, the cyst fluid obtained during surgery was confirmed to be similar to the xanthochromia of cerebrospinal fluid. These findings suggest that recurrent hemorrhage from sinusoid or neocapillaries of the CA were responsible for the formation and growth of the cyst.

Conclusion

Intraosseous CA of the skull is rare, but it should be considered in the preoperative differential diagnosis of skull tumors, even if the characteristic radiological appearances are not evident. Three features of the present case are distinct: the lesion was cystic, the wall of the cyst was enhanced on MRI, and the lesion was accompanied by prominent osteolysis. These findings differ from a typical intraosseous CA and mimic malignant skull tumors. This is the first reported intraosseous CA of the skull with cyst formation. Though still unclear etiologically, we presume that the cyst of the intraosseous CA formed by a mechanism similar to that of a cerebral CA. Specifically, we believe that repeated intra-lesional hemorrhage similar to the common clinical presentation of cerebral CA induced the cyst to form and grow, and that the slow growth of the cyst with intradiploic expansion induced the osteolytic change. CAs with osteolysis and cyst formation can only be definitively diagnosed by surgical resection, as CAs with osteolysis and cyst formation are very difficult to distinguish from malignant skull tumors based on neuro-radiological findings alone.

References