Xanthoma of the Occipital Bone and With Preserved Inner and Outer Bone Cortex: Case Report

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Abstract

Objective We present a unique case of a midline xanthoma of the occipital bone exhibiting atypical imaging characteristics with preserved bone cortex that has not previously been described.

Participant This man presented with refractory headaches and suboccipital pain and a mass within the diploe of the occipital bone but with preserved inner and outer cortex of the bone. Magnetic resonance imaging showed a midline, enhancing, and marrow-replacing process in the occipital bone measuring 1.5 cm in anteroposterior (AP) diameter, resulting in mild indentation of the dorsal aspect of the cerebellar vermis.

Results The patient underwent a suboccipital craniectomy. Tumor resection was from the foramen magnum to the inion and laterally until normal bone was encountered. The xanthoma was yellowish and bled a moderate amount upon resection.

Conclusion An isolated cranial xanthoma with preserved inner and outer bone cortex involving the occipital bone and of midline location has yet to be described. The differential diagnosis of osteoexpansile skull lesion with preserved bone cortex should now include xanthoma. Given the broad spectrum of imaging characteristics exhibited by this unusual diagnosis, surgical intervention is indicated from a diagnostic as well as a therapeutic standpoint.

Keywords
- xanthoma
- tumor
- occipital bone
- histology
- skull base
- neoplastic lesion

Introduction

Xanthoma of the cranium is an exceedingly rare diagnosis. Among these rare cases, there exists a predilection for xanthoma formation within the petrous temporal bone. Cranial osteolysis with destruction of bone cortex is considered an imaging characteristic of this lesion.[1–7] Additionally, there have been descriptions of xanthomas within the cerebral parenchyma proper and elsewhere along the neural axis.[8–16]

We herein report a unique case of a midline, occipital bone osteoexpansile skull base xanthoma with preserved bone cortex in a patient with hypercholesterolemia presenting with severe suboccipital pain and diffuse headaches.

Case Report

This 49-year-old man presented as a referral from his primary care physician after cranial imaging for severe suboccipital pain and headaches revealed a mass within the diploe of the occipital bone. The cranial computed tomographic (CT) scan had been obtained after the patient failed medical modalities for the treatment of his symptoms. The patient's medical history was significant for hyperlipidemia complicated by

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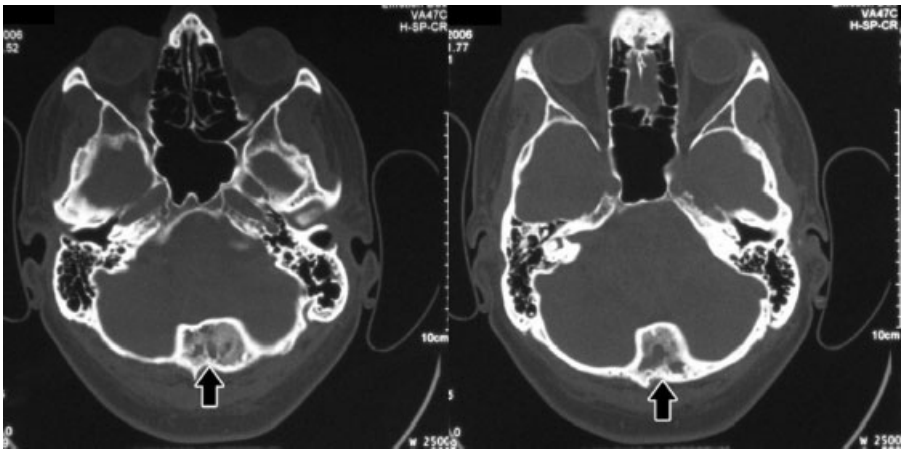

Fig. 1 Brain computed tomography scan ("bone windows") demonstrating an expansile, ground glass appearing lesion (arrows) of the occipital bone. Note the preserved and thickened inner and outer bone cortex.

coronary artery disease, which were being concomitantly treated with lipid-lowering and antiplatelet agents, respectively.

The patient's neurological examination was nonfocal. His cervical range of motion was full. No masses were palpable. There were no other stigmata of hyperlipidemia such as xanthomas involving the subcutaneous tissues. Hematologic analysis revealed a fasting total cholesterol level of 211 mg/dL, a high-density lipoprotein (HDL) of 54 mg/dL, and a low-density lipoprotein (LDL) of 119 mg/dL while on a lipid-lowering agent. Cranial CT scan (bone windows) revealed a mixed lytic and ground glass–appearing lesion within the diploe extending from the external occipital protuberance to the posterior margin of the foramen magnum. The outer and inner bone cortex were noted to be intact and mildly thickened (►**Fig. 1**). Magnetic resonance imaging (MRI) showed a mildly enhancing, marrow-replacing process in the occipital bone measuring 1.5 cm in anteroposterior (AP) diameter, resulting in mild indentation of the dorsal aspect of the cerebellar vermis (►**Figs. 2A–C**).

The lesion was approached via a standard midline incision extending from the external occipital protuberance to the C2 spinous process. A suboccipital craniectomy was performed and bony resection was performed from the foramen magnum to the inion and laterally until normal bone was encountered. There was obvious extradural compression of the cerebellum extending to the craniocervical junction. A cranioplasty was not performed. The xanthoma was yellowish and bled a moderate amount upon resection. Postoperative CT imaging revealed complete removal of the xanthoma, and no recurrence was found at 5-year follow-up. (►**Fig. 3**).

Pathological examination demonstrated histiocytes embedded within a loose, paucicellular fibrous stroma on low power (►**Fig. 4**). High-power examination revealed large tumor cells with foamy cytoplasm and uniform, centrally located nuclei, which were separated into small nests by delicate fibrous bands (►**Fig. 5**). The patient experienced an uneventful postoperative course. It has been 24 months since surgery, and his headaches and suboccipital pain have resolved.

Discussion

This is the first report of xanthoma presenting with osteoexpansile features but preserved inner and outer cortex of the bone. In addition, it is also the first xanthoma involving the

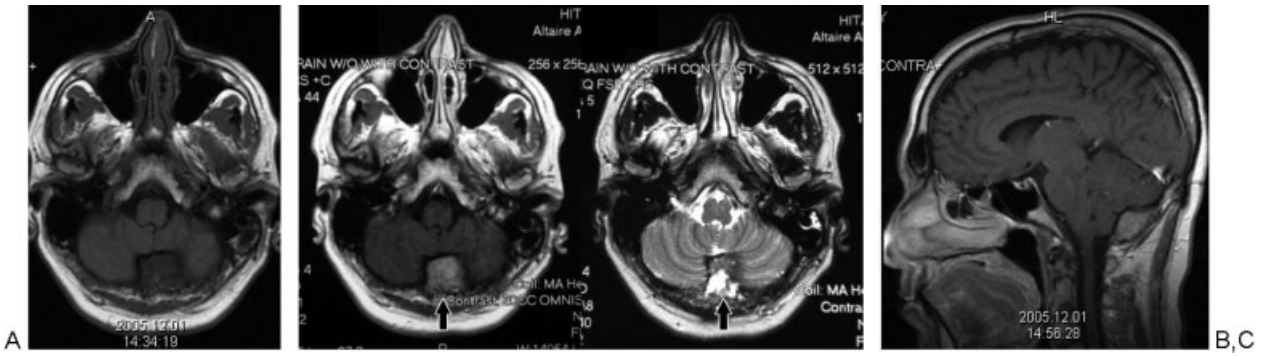

Fig. 2 (A) Axial precontrast T1 magnetic resonance imaging showing extra-axial lesion with mild indentation of the dorsal aspect of the cerebellar vermis. (B) Axial contrasted T1 (left) and T2 (right) magnetic resonance imaging showing an extra-axial lesion with mild indentation of the dorsal aspect of the cerebellar vermis. Note the enhancement of the lesion on the left image. (C) Sagittal postcontrast magnetic resonance imaging showing moderate enhancing extra-axial lesion with mild indentation of the dorsal aspect of the cerebellar vermis.

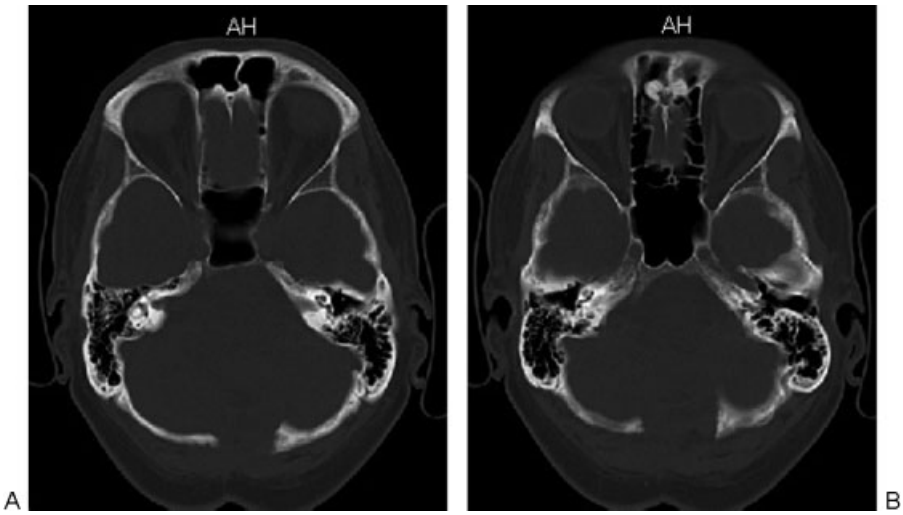

Fig. 3 (A, B) Postoperative computed tomography scan ("bone windows") showing occipital craniectomy with recurrence-free removal of xanthoma on 5-year follow-up image.

occipital bone. Additionally, the midline location has not been described previously.

As is frequently reported, hyperlipidemia is thought to be the predisposing condition leading to the development of this lesion. Elegant theories outlining the pathophysiology of xanthoma formation exists that logically postulate the origins of this non-neoplastic lesion, especially when contrasted with the theories of arterial atheroma formation on a smaller scale.[6,7,13,15] However, from a clinical neurosurgical standpoint, xanthoma of bone is best regarded as a benign tumor of histiocytic origin stemming from bone marrow, the progenitor of the reticuloendothelial system.[6] These lesions are, in fact, locally expansile, can progress, and may produce significant morbidity as exhibited by those patients with facial and/or vestibulocochlear nerve involvement. The diagnosis of an intracranial xanthoma cannot be made by radiographic methods alone. A well-circumscribed, often expansile cranial lesion that appears isodense or hypodense to brain is commonly exhibited on CT scanning, though variants certainly exist. Osteolysis with destruction of the bone cortex has been uniformly reported thus far and has aided radiological differential diagnosis.[2,3,5,8,15,17,18] However, our case presented as an osteoexpansile lesion but with preserved inner and outer

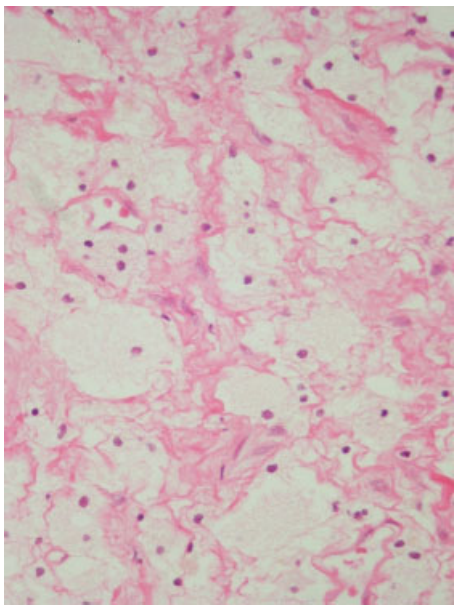

Fig. 4 Photomicrograph. Low-power view showing histiocytes embedded within a loose, paucicellular fibrous stroma, adjacent to a portion of bone. Original magnification 4 × . Hematoxylin and eosin stain.

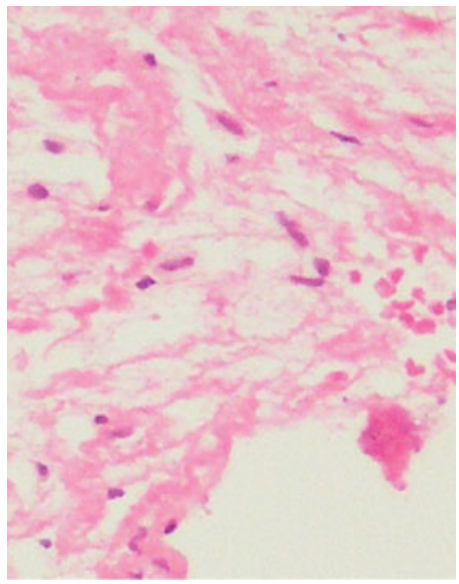

Fig. 5 Photomicrograph. High-power view showing large histiocytes with foamy cytoplasm and uniform, centrally located nuclei. Delicate fibrous bands separate the cells into small nests. Original magnification 20 × . Hematoxylin and eosin stain.

bone cortex, which was even somewhat thickened. The reason for bone cortex preservation remains unclear. Nonetheless, the differential diagnostics of the osteoexpansile skull lesion with preserved bone cortex on plain skull X-rays and/or cranial CT scans with bone reconstructions should now include xanthoma.

MRI reveals a broad spectrum of characteristics, influenced primarily by the intralesional lipid content and hemorrhagic breakdown products. The extent of contrast enhancement is also quite variable.[12,18] The differential diagnosis based on the aforementioned characteristics include but are not limited to epidermoid cyst, dermoid cyst, meningioma, cholesterol granuloma, benign fibro-osseous lesion such as fibrous dysplasia, metastatic tumor, and infectious or inflammatory processes[4,8,11,19] The microscopic demonstration of histiocytes with foamy cytoplasm embedded into a fibrous stroma along with macroscopic intraoperative observations aids in the diagnosis of xanthoma. Histologic appearance of cholesterol clefts and giant cells is helpful but not always present.

Complete resection of lesion will typically result in symptomatic improvement and should be attempted. Addressing hyperlipidemia or other secondary causes, such as diabetes mellitus, through lifestyle modifications and medical therapies is advised in all cases, especially those of subtotal resection to reduce the risk of recurrence or progression.

Conclusion

A unique case of isolated xanthoma with preservation of inner and outer bone cortex of the occipital bone and with midline location is described. The differential diagnosis of osteoexpansile skull lesion with preserved bone cortex should now include xanthoma.

Given the broad spectrum of imaging characteristics exhibited by this unusual diagnosis, surgical intervention with an attempt of radical resection is indicated from a diagnostic as well as a therapeutic standpoint. The likelihood of a coexisting endocrine or metabolic disorder is high, which should prompt an investigation into the patient's general state of health. Through proper management of comorbidities, recurrence and/or progression can often be avoided.

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