Case Report

Idiopathic focal calvarial thinning: A case report

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ABSTRACT

Background: Calvarial bone thinning is a rare clinical entity, with only several cases reported (including Gorham-Stout disease), but the cause is often unknown. Here, we report such a case of unilateral calvarial thinning with an unknown cause.

Case Description: A 77-year-old woman undergoing imaging examination for unruptured cerebral aneurysms for the past several years noticed a progressive cranial deformity. Computed tomography revealed progressive thinning of the right parietal bone and cranial deformity but laboratory tests showed no causative findings. A cranioplasty was performed to protect the brain and confirm the pathology. Grossly, pigmentation and deformity were observed on the outer plate of the bone but the inner plate was intact. Pathological examination revealed preserved bone cells and no necrosis. In addition, there were no findings of vascular hyperplasia or malignancy. It appeared that localized osteoporosis had occurred, mainly in the outer plate of the bone, but the cause was unclear.

Conclusion: Progressive focal calvarial thinning is rarely reported and the mechanism in this case was unknown. It is important to determine the cause of the bone thinning to evaluate the need for surgical intervention from the viewpoint of brain protection and prevention of cerebrospinal fluid leakage.

Keywords: Calvarial thinning, Idiopathic, Parietal bone

INTRODUCTION

Bone mass is tightly balanced between osteoblast-mediated formation and bone resorption by osteoclasts. When this balance is disrupted, bone thickening and thinning occur.[7] There are several known causes of calvarial thinning, including inflammation, trauma, tumors, hyperparathyroidism, granulomatosis, diabetes mellitus, osteomyelitis, systemic mastocytosis, sterile necrosis, long-term steroid therapy, Gorham-Stout disease (GSD), Winchester syndrome, and/or bone aneurysms, although some cases have unknown causes.[8] This report describes such a rare case of unilateral idiopathic calvarial thinning in which cranioplasty was performed.

CASE DESCRIPTION

A 77-year-old woman was referred to our hospital because of a recent and noticeable skull deformity. The patient also had a small and unruptured middle cerebral artery aneurysm
regularly examined by magnetic resonance imaging but no other intracranial lesions had since emerged. The patient had a history of well-controlled hypertension but no other significant medical history, including diabetes or head trauma. No relatives had skull deformities.

Physical examination on admission revealed a 6 × 10 cm depressed deformity on the right parietal region but the skin directly above was normal with no tenderness in the deformed area. Computed tomography (CT) showed that the skull of the right parietal bone was thin but no significant neoplastic lesions are shown in [Figures 1 and 2]. The patient underwent two CT scan examinations, one was 10 years ago and the other was 5 years ago, with a complaint of a slight headache, but was unaware of any skull deformity. Retrospective review showed no abnormalities on CT scan done 10 years earlier [Figure 2a] but slight thinning and deformation of the skull were visible on CT scan done 5 years earlier [Figure 2b]. This indicated gradual thinning and deformation over a period of years. Laboratory findings related to bone metabolism were normal: serum calcium (Ca) was 9.5 mg/dL (8.6~10.2 mg/dL), phosphorus (P) was 3.1 mg/dL (2.5~4.5 mg/dL), parathyroid hormone was 60 pg/mL (10~65 pg/mL), calcitonin was <0.5 pg/mL, alkaline phosphatase (ALP) was 97 U/L (38~113 U/L), interleukin 6 (IL-6) was 3.2 pg/mL (0~0.7 pg/mL), activated Vitamin D was 86 pg/dL (20~60 pg/mL), and rheumatoid factor was 5 IU/mL (0~15 IU/mL).

The patient was cosmetically concerned about the skull deformity. Furthermore, from the viewpoint of the need for pathological examination and since there was not enough bone to protect the brain, a cranioplasty was performed after excising the thinning bone. When the skin over the depressed area was turned over, there was an area of 6 × 10 cm in the right parietal bone that was thinned out and was creating a small crater in the calvaria [Figure 3a]. Bone removal was performed in a grossly normal area along with the depression, since it could not be determined that the lesion was definitely benign before and during surgery. The outer layer of bone was slightly darkened and thinned but the inner layer and dura mater were normal [Figures 3b and c]. A resinous compound was used for cranioplasty. The postoperative course was uneventful and the patient was discharged without sequelae.

Pathological examination showed a significant reduction in cancellous bone in the thinned area. Cortical bone volume was slightly decreased in the outer plate but was almost normal in the inner plate. No lymphatic vessel proliferation, malformation, or lymphocyte proliferation was observed, nor was there any obvious tumor, inflammation, osteolysis, or necrosis [Figure 4]. Thus, it was suggested that osteoporosis

![Figure 1: Computed tomograms on admission ((a), axial, (b) coronal, (c), and (d) three-dimensional imaging) showing thinning of the right parietal bone and cranial deformity.](image1)

![Figure 2: Computed tomograms showing temporal changes ((a) 10 years ago, (b) 5 years ago, and (c) on admission). A gradual thinning of the right parietal bone is observed (white arrows).](image2)
or bone atrophy had occurred idiopathically and locally, mainly in the outer platen.

DISCUSSION

Calvarial thinning can occur during various pathologies both as a primary manifestation in the skull and as a secondary involvement of trauma and various systemic diseases, such as primary and metastatic tumors inducing intracranial hypertension, hyperparathyroidism, inflammatory disease, diabetes mellitus, osteomyelitis, systemic mastocytosis, prolonged steroid therapy, bone aneurysm, or cystic angiomatosis of bone. Diseases that cause idiopathic and progressive osteolysis include GSD and Winchester syndrome.

In this case, with almost normal laboratory tests and no obvious genetic history, it was necessary to speculate on other causes, such as GSD and Winchester syndrome. GSD, also called “vanishing/phantom bone disease,” is characterized by spontaneous and progressive osteolytic lesions in a single bone or multiple bones. The most commonly involved sites are the mandible, followed by the scapula, ribs, humerus, pelvis, and femur while the bones of the skull are rarely affected. Clinical diagnosis of GSD can be difficult as it occurs widely in young and middle-aged individuals and symptoms may resemble and be misdiagnosed as rheumatoid arthritis, osteoarthritis, or osteomyelitis, among other painful conditions of the musculoskeletal system. Therefore, the diagnosis is often made by exclusion after inflammatory, infectious, metabolic, and neoplastic diseases are ruled out. In addition, there are no standardized guidelines for diagnosis. Histopathologic findings in GSD are characterized by presence of angiomatous tissue and absence of dystrophic calcifications, and evidence of local and progressive osseous resorption. GSD was initially suspected in this case, but the diagnosis was not made because histopathology did not find the characteristic angiomatous tissue. Winchester syndrome, conversely, is also a rare genetic disorder characterized by osteolysis but, in most cases, bone loss begins in the limbs, causing pain, and limiting movement. Therefore, we were able to definitively rule out Winchester syndrome.

Osteolysis is most likely due to the nonmalignant growth of hemangiomatous tissue. Bone resorption and destruction in this instance are thought to be caused by a pH decrease due to low hypoxia due to poor capillary circulation, increasing the activity of various hydrolytic enzymes, or by local hyperemia or mechanical forces that promote bone resorption. In this case, however, no obvious osteoblast or osteoclast proliferation was observed in the pathologic analysis and, in addition, the percentage of fatty marrow in the cancellous bone layer was low in the thinning area. Typically, differentiation into adipocytes occurs with aging but it was unclear whether this was directly related to calvarial thinning. Nonetheless, there were apparent changes in the outer plate of the bone, suggesting that chronic external forces to the area, of which the patient was unaware, may have contributed to the thinning. As an example of the forces, obstructive sleep apnea probably due to may result in calvarial thinning. However, in such cases, the external forces should have affected the inner cortex as well as the outer cortex and the cancellous bone, suggesting that the etiology of our case is incompatible with forces due to sleep disorders.
While, in cases of idiopathic bone loss, it is challenging to select the most appropriate treatment, surgery may be essential to prevent secondary disorders such as cerebrospinal fluid leakage\textsuperscript{[1,4,5]} and traumatic brain injury due to bone thinning.\textsuperscript{[1,4,5]} Such a decision is supported by a reported case of resection and cranioplasty in a patient with GSD and a subsequently good prognosis\textsuperscript{[5]} In the present case, the lesion was resected and cranioplasty was performed. One year after surgery, there was no recurrence and the prognosis was good. Although resection of the lesion and cranioplasty seems to be the optimal treatment, further long-term follow-up is necessary because the etiology of the disease is unknown. Therefore, further studies are needed to elucidate the disease’s pathogenesis and determine the optimal treatment strategy.

**CONCLUSION**

We report a case of idiopathic focal calvarial thinning with an unclear cause. Progressive calvarial thinning is rarely reported and the mechanism in this case was unknown. However, it is important to determine the cause of the bone thinning to evaluate the need for surgical intervention from the viewpoint of brain protection and prevention of cerebrospinal fluid leakage.

**Declaration of patient consent**

Patient’s consent not required as patient’s identity is not disclosed or compromised

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**Conflicts of interest**

There are no conflicts of interest.

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