

Hyperostosis Frontalis Interna and Temporalis Interna: A Rare Case Report

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Abstract

Hyperostosis is a slow-growing benign bone tumour often seen in the bones of the cranial vault, more commonly found in elderly females. It is an incidental finding noted during radiological examination. The clinical manifestation of such tumour depends on its location inside the cranial cavity: the proximity to the paranasal sinuses, brain tissue, nerves, or blood vessels, etc. Its clinical findings may range from mild obstruction of the paranasal sinuses or blood vessels to severe compression of the surrounding cranial nerves. Here a rare case of hyperostosis of the frontal and temporal bones found during a routine cadaveric dissection in the Department of Anatomy is presented. Causes of the formation of such unusually enlarged bone masses inside the cranial cavity and their clinical presentation are discussed.

Keywords

Hyperostosis Cranialis Interna; Hyperostosis Frontalis Interna; Skull; Calvaria; Endocranium

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Introduction

Bone tumours are neoplastic growths that are seen in a population of diversified age groups. Benign tumours are non-metastatic growths. Based on their location, size, and extent of growth they can compress nearby tissues, imparting various patterns of clinical presentations. Conventional plain X-rays and advanced diagnostic scan aid are used to elucidate the involvement of anatomical structures [1]. Osteoma is a bone tumour that accounts for about 11% of benign bone growths. It is one of the most seen bone tumours, often incidentally detected during radiological investigations [1, 2].

Hyperostosis cranialis interna is a bone disorder characterized by endosteal hyperostosis, osteosclerosis of the skull base and the calvaria. It is associated with progressive bone overgrowth resulting in compression of cranial nerves I, II, V, VII, and VIII. Morgagni first described abnormal bone growths in the inner table of the cranium in 1765 [3]. They are abnormal sclerotic bone lesions seen in the inner cranial vault [4]. Abnormal bony growths associated with other clinical conditions are classified into the following clinical syndromes: Morgagni syndrome and Stewart-Morel syndrome. Morgagni-Stewart-Morel syndrome is another unique condition involving hyperostosis frontalis interna (HFI) associated with obesity, metabolic distur-

bances, and neuropsychiatric disorders [5]. Bone dysplasia can be caused by genetic or nongenetic factors, resulting in the disruption of the ossification process which can lead to hyperostosis; it can manifest itself in clinical or sub-clinical forms. It is a condition that may arise due to failure in bone resorption in prone individuals due to genetic mutations. An example of such conditions is hyperostosis corticalis generalisata (van Buchem disease) which is an extremely rare disorder showing uniform cortical thickening of the skull bones. It is a heterogeneous group of diseases involving deranged genetic pathways, showing abnormal proliferation of osteoblasts leading to excessive skull bone thickening [6]. The shape of hyperostosis deformities showed its genetic basis for its occurrence in the family [5].

Based on the incidental HFI findings in a cohort of study participants, patients with HFI were subsequently subjected to the additional radiological evaluation of their appendicular bone (femur) by means of computed tomography (CT) and DEXA scanning. The findings revealed increased cortical thickness and density of the femur bone. Subjects with HFI might, probably, indicate an underlying systemic metabolic bone derangement [7]. In case of HFI, the study found that the degrees of anisotropy differed between HFI subtypes in males. But there was no significant difference in the microarchitecture of the bone mass be-

tween males and females [8]. Fraction analytical findings of HFI bone fragments were categorized into the following types: A, B, C, and D, indicating bone density in an inclining order. Studies of HFI bone autopsy samples showed that type B HFI was found more commonly in males, and type D HFI was 4 times more common in females. In all the subjects, including males and females, increased thickness of the frontal and temporal bones was observed [9]. Hyperostosis of three cranial bones involving the frontal, temporal, and sphenoid bones was found in an elderly female cadaver during dissection. All these bones showed bone overgrowth characterized by smooth, ossified ridges and nodules protruding from the inner table of the cranium. Microscopically thickened compact bone was observed during haematoxylin and eosin (H & E) staining [10]. These syndromes with HFI were found clinically, as well as in post-mortem studies in the association of neuropsychiatric diseases with metabolic derangements [11, 12]. Based on some observations showing hyperostosis conditions involving other segments than the frontal bone might have been associated with other metabolic changes in the body, they were referred to as metabolic cardiopathy by Moore [13]. Temporal bone osteoma is another rare finding arising from the petrous part of the temporal bone that can cause progressive auditory impairment due to vestibulocochlear nerve compression [14]. However, the reliable data on hyperostosis involving several cranial bones are scarce. Here, a rare case of hyperostosis of the temporal and frontal bones is reported.

Case Report

During a routine dissection in the Department of Anatomy, JSS Medical College, Mysore, after removing the entire brain in a 75-year-old female cadaver, we observed two pairs of abnormal bone growths in the floor of the cranial fossa. Both bone masses were limited on either side of the cranial fossa, without crossing the sagittal plane. The first pair of bone masses was noticed in the anterior cranial fossa. They were located bilaterally in the inner cranial vault, arising from the internal surface of the squamous part of the frontal bone lying near the frontal sinus. In the same frontal bone, another tumour was seen at the superior surface of the orbital plate, which forms the floor of the anterior cranial fossa (Fig. 1). In the middle cranial fossa, a pair of bilateral bony masses was noticed at the cerebral surface of the squamous part of the temporal bone (Fig. 2). These masses were pale-coloured and hard in consistency. Histopathological examination showed the lamellar pattern of the trabeculae, the intervening spaces filled with hemopoietic tissues (Fig. 3).

Discussion

Benign skull base tumours are diversified in their origin; they arise from entities such as the brain parenchyma, the dura mater, the cranial vault, etc. Some tumours are commonly seen in the calvarium. They tend to show bone overgrowth of the outer (exostotic) or inner (enostotic) tables of the calvarium, resulting in a condition known

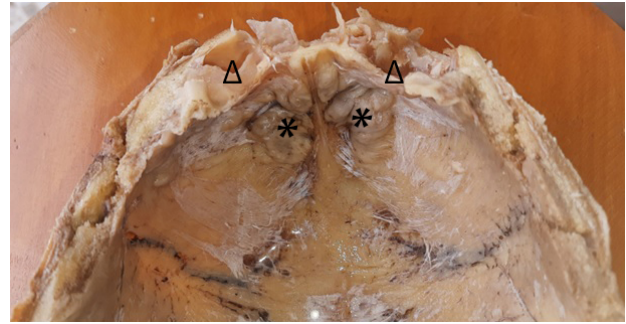


Figure 1. Calvaria: internal view. Δ – Frontal sinus lying close to the frontal bone mass in the cranial vault.
* – Frontal bone mass.

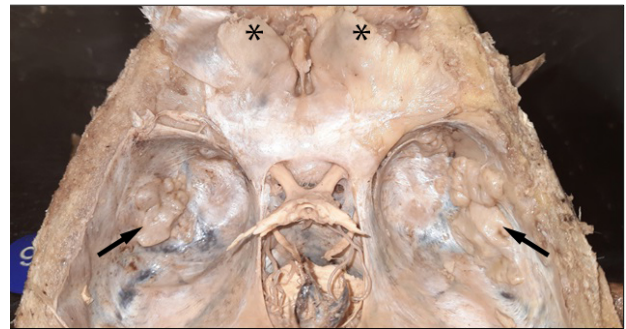


Figure 2. Base of skull: internal view. \rightarrow – Temporal bone mass. * – Frontal bone mass.

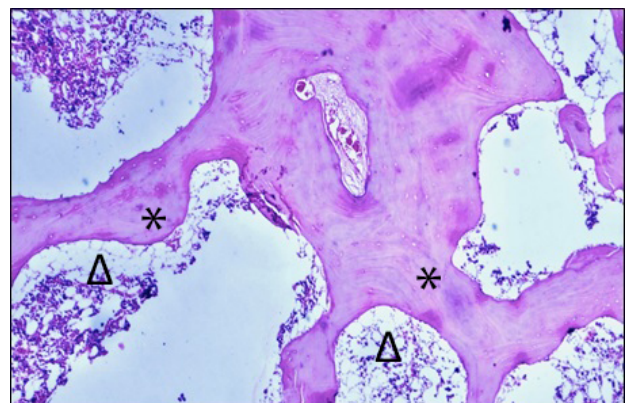


Figure 3. Histopathological findings. H&E staining. Magnification - 10x. Δ – the thick lamellar pattern of the trabeculae, the lacuna containing osteocytes.
* – the intervening space containing hemopoietic cells and fat.

as osteoma [15]. The clinical symptoms of intracranial space-occupying bone tumours may manifest themselves by insidious onset of a mere headache to severe disabilities. Such bone lesions affect the intracranial volume capacities leading to a change in the intracranial pressure, which, in turn, affects blood or cerebrospinal fluid (CSF) circulations or the brain tissue itself. Often, such lesions produce no obvious clinical symptoms and may be recognized incidentally during radiological investigations [16].

The Monro-Kellie Doctrine states that the three main contents of the cranium – the brain tissue, CSF, and blood,

exist in equilibrium. An increase in the volume of one component should cause a decrease in the volume of one or both remaining components [17]. In our case, tumours were located close to the frontal sinuses (Fig. 2); depending on the extent and size, frontal tumours growing into the wall of the frontal sinus can lead to sinus compression.

A very rare case of hyperostosis was reported in a 57-year-old female with a sudden episode of syncope. It was associated with progressive short-term memory decline. The temporal lobe of the cerebral hemisphere contains the hippocampus which plays an important role in turning short-term memories. Radiological findings showed cranial hyperostosis with circumferential brain compression leading to herniation of the hippocampus into the posterior fossa. In addition, it showed brain herniation because of cranial volume deprivation due to space-occupying lesions such as hyperostosis. Hence, the anatomical location of such an intracranial tumour is critical from the clinical standpoint. The clinical symptoms of such abnormal bone growths may be obvious or go unnoticed for a long time [18]. The number of foramina in the skull base plays an important role in transmitting various contents into and out of the cranium. The internal acoustic canal (IAC) of the petrous portion of the temporal bone provides a passage through which important structures such as the vestibulocochlear and facial nerves can pass through. A 67-year-old female presented with complaints of hearing loss, vitiligo, tinnitus, imbalance, etc. over the past two years, and was subsequently subjected to a clinical examination. During radiological examination, bone overgrowth in the inner wall of the IAC with canal narrowing was noticed, which led to vestibulocochlear nerve compression causing the above symptoms. Surgical decompression of the IAC wall provided significant relief to the patient. Such exostosis or bone hypergrowth at certain anatomical points is presented with an explicit manifestation of clinical symptoms [19].

A young 30-year-old woman with clinical findings of HFI presented with increased intracranial pressure of over 60 mm Hg associated with a history of intermittent attacks of headache and syncopal episodes. Increased intracranial pressure may lead to brain herniation and neurovascular compression which necessitates early measures such as surgical decompression. The histopathological findings of abnormal bone growth from this case have shown the lamellar bone patterns; these findings are consistent with our case of HFI [20]. HFI was detected as an incidental finding in a 53-year-old postmenopausal woman who presented with recent episodes of seizures. The women had no prior history of epileptic attacks or loss of consciousness. Her electrocardiogram (ECG) result was normal, but CT scan showed HFI [21]. In another case, a 27-year-old woman presented with complaints of treatment-resistant chronic headache. During radiological examination, HFI was incidentally found by means of CT scan. It was presumed that changes in sex steroid levels might impact skull bone growth in women. These observations showed the insidious nature of such HFI anomaly with prolonged mild symptoms, often treated symptomatically without little attention [22]. A subset of the postmenopausal female

population was analysed to find the prevalence of HFI. The majority of HFI cases were found in older females (50-59 years of age) that demonstrated HFI prevalence in a particular age group [23]. The above-mentioned studies have reported HFI cases more often seen in the female population, especially in women of postmenopausal age.

Deranged bone mineral metabolism in premenopausal women with an increase in dehydroepiandrosterone and testosterone levels showed the potential role of androgens in metabolic bone disorders [24]. Skeletal muscle tissue is in a relatively hypoxic environment which can induce subtle metabolic derangements that can manifest as tumours. For good bone health, there must be a functional balance between osteoclasts and osteoblasts. Bone formation is an energy-intensive and metabolically demanding process, where enhanced oxidative stress can impact osteoblast differentiation [25]. Metabolic bone disorders represent a wide range of heterogeneous disorders; they are highly influenced by the supply of micronutrients, including proteins and minerals. There is a tight metabolic regulation between bone formation and resorption which occur at discrete sites of the skeletal tissue distributed throughout the body. The asynchronous changes in osteoblast and osteoclast signalling pathway activity could result in various patterns of HFI [26].

Conclusions

Intracranial bone tumours may be asymptomatic, and their aetiology may remain undetermined. They are benign tumours seldom presented with obvious clinical manifestations. X-ray investigations are pivotal in their diagnosis, but they are most often incidental radiological findings. The clinical manifestations and severity of intracranial bone tumours depend on their location and size, which ultimately affects the nearby structures or intracranial pressure. Real anomalous cadaveric findings during dissections will certainly provide additional knowledge on clinical anatomy to preclinical medical students.

Ethical Statement

No approval of institutional review board was required.

Conflict of Interest

The authors declare that no conflicts exist.

Financial Disclosure

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