Hyperostosis Frontalis Interna and a Question on Its Pathology: A Case Report

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Financial support: None declared
Conflict of interest: None declared

Patient: Female, 90-year-old
Final Diagnosis: Cerebrovascular infarction
Symptoms: Unknown symptoms – post-mortem anatomical study
Medication: —
Clinical Procedure: —

Specialty: Anatomy • Neurology

Objective: Unknown etiology
Background: Hyperostosis frontalis interna is a boney overgrowth of the inner side of the frontal bone of the skull caused by overgrowth of the endocranial surface. It is most often found in women after menopause. It is also associated with hormonal imbalance, being overweight, history of headaches, and neurocognitive degenerative conditions. Female gender, advanced age, extended estrogen stimulation, and elevated leptin levels may also play a role. The thickening is usually confined to the frontal bone, but it can spread as far as the anterior parietal and temporal bones.

Case Report: During a medical school dissection course, an extensive boney overgrowth in the frontal regions covering the inside of the frontal bone of the skull of a 90-year-old female donor, who died of a cerebrovascular infarction, was identified. This boney overgrowth was mainly confined within the frontal region, but there was some boney overgrowth that extended to the temporal bones. The overgrowth in the endocranial of the temporal bone was not as severe as the overgrowth of the frontal bone. The morphology of the overgrowth was rigid, uneven, and bumpy. Based upon the physical characteristics, we concluded that this presentation was consistent with hyperostosis frontalis interna.

Conclusions: Our female donor was found to exhibit a phenomenon which could be clinically underdiagnosed due to its internal nature and asymptomatic presentation. Insight into the potential causes of HFI and its identification during clinical evaluation offers a path for future research to better identify and manage cases of HFI.

Keywords: Calvarial Hyperostosis • Endocrinology • Hyperostosis Frontalis Interna • Menopause • Pathology

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/937450
Background

Hyperostosis frontalis interna (HFI) normally presents with no symptoms and is often an incidental finding when obtaining imaging of the head such as MRI (magnetic resonance imaging), CT (computed tomography) scan, or X-ray [1]. HFI is described as a bony overgrowth of the frontal bone and can sometimes spread to the temporal or parietal bones. There are different classifications when diagnosing HFI and they are described as “types” dependent on the extent of involvement, appearance, shape, border, and involvement of other bones [2]. According to Hershkovitz et al, the 4 distinct types are classified as A, B, C, or D. They describe the morphology of the calvarium and what percent is covered with boney overgrowth; 25%, 50%, or more than 50% for types B, C, and D, respectively. Type A is just based on isolated small overgrowths covering less than 25% of the frontal bone [3]. This disorder presents most often among the middle-aged and elderly, especially females, and the severity has been shown to increase with age [2,4]. HFI has presented in many different cases of women who are post-menopausal, people who are obese, have endocrine abnormalities, hirsutism, chronic headaches, seizures, or dementia [1,5]. When this condition occurs in males, some cases had an increase in androgen stimulation [2]. HFI has also been linked to increased levels of leptin, which is higher in females more than males, and is associated with high body mass index and obesity [6]. As the severity of HFI increases, progressive compression of the dura mater can cause decreased blood supply to the meninges and certain regions of the frontal lobe, including Broca’s speech area, the premotor cortex, and the prefrontal association cortex, may further result in headaches and impairment of cognition and memory [1].

In the absence of any standard therapy for this rare condition, supportive care with pain management is suggested [1]. Best practice management of HFI begins with patient counseling about the diagnosis and outlining a plan of treatment of headache pain. Treatment is mainly supportive. Prompt diagnosis helps to prevent a misdiagnosis and prevent the patient from undergoing many unnecessary diagnostic tests [1]. When HFI is associated with secondary conditions (eg, seizure or Morgagni-Stewart-Morel syndrome), management should be focused on treatment of the secondary condition [7].

Case Report

This case comes from a 90-year-old woman who graciously donated her body to the University of Toledo College of Medicine and Life Sciences donor program. The patient died due to a cerebrovascular infarction. All other details of her past medical history were not available to us due to the anonymity of the donor program. When opening the cranium for dissection, our dissection team noticed that the surface of the endocranium was rough and bumpy (Figure 1). This growth of bone was not present throughout the skull and was mainly localized to the frontal bone with just some overgrowth in the temporal bones. The frontal bone was significantly thicker than the parietal, temporal, and occipital bones (Figure 2). This overgrowth of bone in the cranium is called hyperostosis frontalis interna. Using the classification system of Hershkovitz et al, it appears that most of the frontal bone and endocranium is involved, or more than 50%, making this type D [3]. It is also important to note that the overgrowth does not cross the calvaria midline, which is another defining feature of HFI [5].

Discussion

There are multiple case reports of hyperostosis frontalis interna (HFI). However, there are no concrete data that show why this occurs. Our goal here is to discuss possible avenues to open the doors for more research to discover the mechanisms involved in this clinical presentation.

Due to the asymptomatic nature of this disease, it is often undiscovered or discovered as an incidental finding [8]. Other than the fact that it can go undetected, it does share some...
Further investigation may establish a correlation between different pathologies and HFI, so physicians are more aware to make the diagnosis. There are many reports of different hormonal imbalances that also report instances of HFI in those patients. In one study (n=36), 72% of patients who had acromegaly had HFI, and of those, 19 had hyperprolactinemia [10]. The acromegalic patients who had hyperprolactinemia also had higher evidence of HPI than those in the normal control groups. The experimental results support that HPI can be a marker of pituitary disfunction [9]. Another case report, from Yamakawa et al [4], discusses a male with atrophied gonads who also had HPI, which further supports the idea that HPI could be due to gonadal imbalances. In vivo models could be used to directly control for androgen levels in mice or other vertebrates to scientifically test for a correlation. Lifestyle and diet were also found to have an impact on HFI prevalence from different archeologic periods [11]. Different populations were analyzed for the occurrence of HFI and how active those populations were. In the study, they confirmed that longer exposure to estrogens increased the occurrence of HFI in females. Based on the activity levels of the different populations, this article found that groups of farmers who lived a mobile pastoral lifestyle ate more meat than carbohydrates. They also ate more milk and dairy. It was hypothesized that this group of people had lower fasting glucose levels and elevated fasting insulin levels due to the physically challenging work requirements. If elevated insulin levels suppress sex-hormone-binding protein and the insulin-like growth factor-binding protein 1, then there could be an increase in free androgens eventually leading to HFI [11]. In the United States we have only seen an increase in obesity since 1999. The obesity prevalence in the U.S. from 2017 to 2020 is 41.9% [12].

We hypothesize that with the increase in obesity there will also be an increase in HFI. A recent case report from 2021 presented a young woman who had head trauma that led to development of HFI. The patient’s symptoms of schizophrenia began afterwards and when treatment did not help, they imaged her head, leading to the finding of HFI. This is a rare case where trauma could be the cause of HFI [13]. This case report illustrates the need of all specialties, including psychiatry, to be aware of HFI. Clinical presentation of HFI as stated above is similar to many others, which furthermore increases the need for knowledge of why this occurs, how to diagnose and treat it, and what other causes HFI has. Lastly, a case report from Ramchandren and Liebeskind [14] presented a patient with Klinefelter’s syndrome who was also documented to have HPI. The patient presented with chronic headaches and, principal findings with other illnesses, giving it the possibility to be underdiagnosed or misdiagnosed [8]. One disease, for example, is osteitis deformans (Paget’s disease), which is a pathologic thickening of bones such as the skull, spine, and pelvis. This disease occurs mainly in the middle-age to elderly population and accounts for 1% of the population. This classically presents with a thickening of the skull and often does not have symptoms. Most people notice a change in hat size and if it does progress it can eventually cause hearing loss if the petrous part of the temporal bone impinges the vestibulocochlear nerve [8]. The major difference of this presentation is the location being in the temporal bone. Also, in Paget’s disease, the bone is brittle due to the pathology of the osteoblasts and osteoclasts at work. In HFI, the bone is not brittle and is exceptionally durable.

Figure 2. The donor’s calvarium illustrating the degree of thickening of the frontal bone as well as the minor overgrowths of the temporal bone.

A second condition – osteomatosis (Leontiasis Ossea or Virchow’s Disease) – can also mimic HFI. Osteomatosis is a hereditary disorder that involves the membranous bone of the skull. This disease involves significant thickening of the cranial bones, especially the maxilla, mandible, and frontal and temporal bones. A case study by Plenk and Garner [9] of a family with a high incidence of colonic polyposis identified a connection between the large-bowel lesions and bony tumors of the face and scalp in the same individuals. While the process of bony growth can begin in early adolescence, the growth is slow and may go unnoticed for many years. Knowledge of these conditions could be critical when diagnosing a patient so that it is not misdiagnosed.
after negative tests for central nervous system abnormalities, an MRI was done, which resulted in the diagnosis of HFI. The hormonal imbalances in people with Klinefelter’s syndrome could be at the root of the cause. This may explain why HFI develops and should be a target of further research. There are many routes that could be explored with regards to discovering a cause of HFI. Further case studies and experimental research looking into lifestyle, insulin levels, and androgen levels could aid in that discovery. More established correlations with HFI may allow physicians to perform correct diagnosis and prompt treatment.

Conclusions

Due to the lack of consensus for the cause of hyperostosis frontalis interna, we discussed different avenues that could be addressed in future research on this rare finding. There are many endocrine-driven pathways that could be explored to assess impact on patients with HFI, such as case reports on people with Klinefelter’s syndrome, acromegaly, and trauma, and those with atrophied gonads. Furthermore, if insulin and lifestyle are correlated with HFI in anheologic findings, then it could be useful to investigate current populations and conduct case studies to see if those with type 2 diabetes mellitus are developing HFI later in life. HFI is not routinely considered as a cause of patient symptoms [11]. If HFI is better known in the world of clinical medicine, this diagnosis can be made more effectively, and patient education and treatment would be timelier and focused. Lastly, if insulin resistance is correlated with HFI, then we should see an increase in cases of HFI in the future with regards to obesity prevalence in the United States, which makes it even more important for those to become aware of this diagnosis.

Acknowledgements

The authors would like to express their gratitude to: The donor who graciously donated their body for the medical students at UTCOMLS; The donor’s family members, Elizabeth Dalzell, the anatomy lab supervisor, and embalmer; and Margaret Hoogland for her assistance with the preparation of the final manuscript.

Declaration of Figures’ Authenticity

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References: