

CASE REPORTS

Hyperostosis frontalis interna (HFI): A case report and review of literature

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ABSTRACT

Hyperostosis frontalis interna (HFI) is characterized by excess growth of the frontal bone. Although the exact cause of HFI is not known, it has been found to be associated with increased age, prolonged estrogen stimulation and elevated leptin. Literature shows its association with seizure, dementia, obesity, headache and endocrine abnormalities. HFI may be syndromic and non-syndromic. Timely identification of HFI can prevent unnecessary diagnostic tests. We report a case of non-syndromic HFI in an elderly female who presented with headache.

Key Words: Hyperostosis frontalis interna, Dementia, Headache

1. INTRODUCTION

Hyperostosis frontalis interna (HFI) is an uncommon condition characterized by excess bone growth with multiple nodules mostly on the inner table of the frontal bone but occasionally extending into other cranial bones. Although association has been postulated with increased longevity, prolonged estrogen stimulation and increased leptin levels, the etiology is mostly unknown.^[1] The syndromic forms of HFI like Morgagni's syndrome, Stewart-Morel syndrome and Troell-Junet syndrome are known to be associated with obesity and endocrine abnormalities. Non-syndromic HFI is often described as an incidental imaging finding in postmenopausal women. The disorder may be associated with a variety of conditions such as seizures, headaches, obesity, diabetes insipidus and excessive hair growth and sex gland disturbances.^[2] We report a case of non-syndromic HFI in an 85-year-old female presenting with headache.

2. THE CASE

Eighty-five-year-old Caucasian woman came to the clinic with the complaint of memory decline and chronic headache at frontal area for 2-3 years. Headache was moderate to severe in intensity, with no fever, nausea, vomiting, convulsions, visual disturbances, altered sensorium or focal weakness. Headache was constant throughout the day, relieved with sleep and was refractory to use of sertraline, levetiracetam or non-steroidal anti-inflammatory drugs. Examination showed an awake, alert elderly female with normal vital signs. Systemic exam was non-contributory except maxillary sinus tenderness. The Saint Louis University Mental Status Examination (SLUMS) was attempted for dementia but could not be completed as the patient was not willing to answer due to headache. However, family members reported she was having decline in the memory and cognitive functions for the past few years.

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Figure 1. X ray skull showing hyperostosis of frontal bone

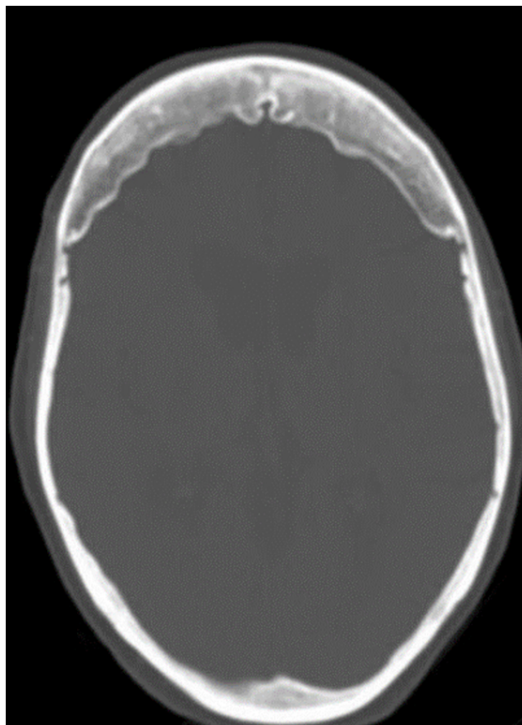


Figure 2. CT scan head showing hyperostosis of inner side of frontal bone

lipoprotein, high density lipoprotein and triglyceride levels were normal. Vitamin D level was low to 15 ng/ml. Sinus X-ray was positive for inflammation of the maxillary sinus as well as thickening of frontal bone consistent with HFI (see Figure 1). The patient was appropriately treated with antibiotics and nasal steroids but she reported that the headache did not resolve. Due to persistence of headache, computed tomography of head was done which revealed hyperostosis frontalis interna without other intracranial abnormalities (see Figure 2). After the diagnosis of HFI, supportive management for headache with analgesics was advised as there is no standard therapy for this rare condition. Subsequently, she was examined by our neurologist who was able to do a cognitive assessment and it was suggestive of dementia.

3. DISCUSSION

Hyperostosis frontalis interna (HFI) presents predominantly in postmenopausal women and is often associated with endocrine abnormalities, obesity, dementia and chronic headaches.^[3] It is typically an incidental finding in head imaging like X ray, CT scan or MRI. Although it has been reported in 5%-12% of population it is mostly seen in elderly obese females.^[4] In our case, the patient was an elderly woman with headache of long duration with no localizing signs in neurological exam except for cognitive impairment and finally was diagnosed to have HFI.

Studies have shown that HFI is more common in the modern population compared to historic populations for unknown reasons. Approximately 2,000 skulls from various geographic locations and ethnic groups as early as in the 7th century and a large group of European skulls prior to the 19th century were studied but none of these skulls displayed HFI. On the other hand, a study of around 1,700 skulls from the 20th century showed HFI in 5% males and 24% females.^[5] We can conclude from these studies that HFI is an underdiagnosed condition in elderly females. HFI is not very well studied and often not recognized. Its significance as an etiology to headache of the elderly is not well known.

HFI has been classified by the extent of bone overgrowth in frontal bone area of the endocranium. Type A is less than 1 cm beyond normal bone growth. Type B, C and D are bone overgrowth involving 25, 50 and more than 50 percentages respectively.^[6] As the HFI gets severe, significant compression of the meninges can lead to compromise in meningeal blood supply and specific regions of neural tissues in the frontal lobe including Broca's area, the premotor cortex, and the prefrontal association cortex resulting in headaches, impairments with cognition and memory.^[6] Our patient had headache refractory to analgesics because of hyperostosis frontalis externa. In the absence of any standard therapy for

Laboratory results showed normal complete blood count. Thyroid stimulating hormone was normal. Low density

this rare condition, supportive care with pain management is suggested. Our patient was counselled about the diagnosis with plan for management of headache with analgesics.

4. CONCLUSIONS

Hyperostosis frontalis interna is mostly an incidental finding in head imaging. It may present with symptoms of intracranial space occupying lesion like headache, seizure or

dementia. Treatment is mostly supportive with no definitive modality of treatment available. Prompt diagnosis helps to prevent unnecessary investigations and proceed with supportive care. More studies are needed to find out the appropriate therapy for this rare condition.

CONFLICTS OF INTEREST DISCLOSURE

The authors have no competing interests to declare.

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