Unilateral hyperostosis frontalis interna

Case report

TAKESHI HASEGAWA, M.D., HARUHIDE ITO, M.D., SHINJIRO YAMAMOTO, M.D.,
KATSUHIKO HABA, M.D., AND HIDEAKI MURATA, M.D.

Department of Neurosurgery, School of Medicine, Kanazawa University, Kanazawa, and Fukui
Prefectural Hospital, Fukui, Japan

The authors describe a case of atypical unilateral prominent hyperostosis frontalis interna in a 50-year-old
Japanese woman with psychiatric symptoms. Radiological, computerized tomographic, operative, and histo-
logical findings of this rare case are presented.

KEY WORDS • hyperostosis frontalis interna • Morgagni’s syndrome •
Morgagni-Stewart-Morel syndrome • computerized tomography

Hyperostosis frontalis interna (HFI) was first
described by the anatomist Morgagni more
than 200 years ago. His case was associated with virilism and obesity in a 75-year-old woman. The condition has often been reported in association with various metabolic, endocrinological, and neuropsychiatric manifestations. Its symptom complex has been referred to as the Morgagni’s syndrome, the Stewart-Morel syndrome, “metabolic craniopathy,” or the Morgagni-Stewart-Morel syndrome.

Hyperostosis frontalis interna is characterized by an irregular nodular thickening of the inner table of the skull and at times the diploë of the cranial bones. There is a predilection for the frontal squama, with bilateral symmetry and an unaffected midline cleft. Although HFI is a fairly common finding in elderly European and American women, in Japan it is extremely rare.

We report a case of atypical unilateral prominent HFI in a Japanese woman. The patient recovered after excision of the thickened bone.

Case Report

This 50-year-old Japanese woman came to the emergency room on August 22, 1979, with the chief complaint of a mild accidental burn about the size of a child’s palm on her back. She also complained of dizziness and cried out frequently. She sometimes spoke of “being blind” or “being possessed by a fox.” This was regarded by the psychiatrist as a hysterical manifestation associated with paranoia. Routine x-ray studies revealed an abnormal skull (Fig. 1), and she was referred to the neurosurgical department.

History. One year before admission, she had had another episode of hysteria. Her family history was negative, and she had three healthy children. Her menstrual periods were regular until 2 years before admission, and thereafter her menstrual periods became irregular.

Examination. The patient weighed 47 kg and was 156 cm tall. Physical examination was normal except for the burn. There was no neurological deficit. Laboratory studies revealed a serum alkaline phosphatase level of 118 mU/ml (normal range 30 to 85 mU/ml). Serum calcium, urinary 17-ketosteroids, 17-hydroxycorticosteroids, basal metabolic rate, and cerebrospinal fluid values were within normal limits.

Skull x-ray films showed increased thickness and density of the squamous portion of the left frontal bone, which projected inward, and absence of the frontal sinus (Fig. 1). The outer table and the midline of the squama frontalis were not involved.

Computerized tomography (CT) scanning demonstrated that the bone change was characterized by dilatation of the diploë space, with compression of the left frontal lobe. In the hyperostotic region, a high-density area indicating sclerotic bone between the skull tables was shown to be attached to the inner table (Fig. 2). A technetium-99m methylidiphosphonate bone scan re-
Unilateral hyperostosis frontalis interna

**Fig. 1.** Preoperative plain skull films, left lateral (**left**) and anteroposterior (**right**) views, demonstrating left-sided unilateral hyperostosis frontalis interna and absence of the frontal sinus.

revealed intense activity over the left squama frontalis (Fig. 3).

**Operation.** A left frontal craniotomy was carried out on September 17, 1979. The outer surface of the frontal bone was found to be intact. The dura was not adherent to the thickened squama frontalis, which caused marked depression of the left frontal lobe. The affected bone presented a bumpy appearance on its inner surface, and measured 4 cm at its thickest point. Both the diploic bone and the inner table were excised and sent *en bloc* for histological study. The outer table of the bone flap was utilized for cranioplasty. At the conclusion of the operation the brain expanded to its normal contours.

**Postoperative Course.** The postoperative course was uneventful. Postoperative CT scans demonstrated disappearance of the frontal lobe depression (Fig. 4). The patient was discharged on the 10th postoperative day, with the burn on her back also well healed. During the 3 years since the operation she has been well, without any neuropsychiatric disturbances.

**Histological Findings.** On gross examination, the thickening of the bone appeared to result from expansion of the diploë. Microscopically, the expanding diploë contained new bone which consisted of spongiose trabeculae of lamellar origin. Fatty marrow filled the intertrabecular spaces. The inner table was composed of lamellar compact bone with active-looking haversian systems. A spongy appearance was noted on its diploic side (Fig. 5).

**Fig. 2.** Computerized tomography scans before operation. **Left:** Plain scan showing hyperostotic bone change, which compresses the left frontal lobe. **Right:** Bone window scan showing bone change characterized by dilatation of the diploic space. There is a high-density area of sclerotic bone attached to the inner table.

**Fig. 3.** Preoperative bone scans, anterior (**left**) and left lateral (**right**) views, showing radionuclide uptake over the left squama frontalis.
FIG. 4. Postoperative plain computerized tomography scan showing reexpansion of the left frontal lobe.

FIG. 5. Photomicrographs of the decalcified specimen. Upper: The expanded diploë consists of spongy trabeculae. The diploë becomes thicker near the inner table. The outer table has been removed. H & E, × 3. Lower: The inner table shows compact lamellar bone with active-looking haversian systems. H & E, × 40.

Discussion

Moore\textsuperscript{14} was the first to classify fully the roentgenographic changes of cranial hyperostosis. His classification system included the following categories: 1) hyperostosis frontalis interna (HFI), in which the nodular or sessile thickening composed of newly formed cancellous bone is present on the squama frontalis; 2) nebula frontalis, in which the smooth thickening is seen at the upper portion of the squama frontalis; 3) hyperostosis calvaria diffusa, in which all the bones of the cranium are thickened; and 4) hyperostosis frontoparietalis, in which the thickening is localized to the frontal and parietal bones.

Histologically, as demonstrated in our case, the essential process of HFI is thought to be a deposition of new bone on the inner table and a progressive development of diploë in this table.\textsuperscript{10,11,14,17} In our case, the increased radionuclide uptake on bone scanning at the site of HFI and the elevated serum alkaline phosphatase level\textsuperscript{5} suggested that the bone changes were in an active phase.

Important characteristics of HFI are that it stops just before reaching the midline and that it tends to be bilateral.\textsuperscript{10,13,20} Our case represents a very rare instance of unilateral HFI. With this unusual variation, the differential diagnosis should include meningioma, calcified epidural hematoma, osteoma, or fibrous dysplasia.\textsuperscript{18,20}

Many different illnesses and symptoms have frequently been attributed to cranial hyperostosis.\textsuperscript{2-4,8,11,13,14} These include obesity, virilism, diabetes mellitus, hypertension, headache, cranial nerve defects, fatigability, convulsive seizures, mental dullness, dementia, irritability, depression, and hysteria. On the other hand, the existence of any clinicopathological significance of cranial hyperostosis has been questioned because the condition can occur without clinical symptoms.\textsuperscript{1,6,17} It still remains uncertain whether HFI represents a real clinical syndrome, and its etiology is as yet unknown.\textsuperscript{7,16}

From a clinical viewpoint, however, prominent HFI may compress soft tissues, with resultant dural irritation and pressure atrophy of the brain.\textsuperscript{10,13,14} Surgical decompression with removal of the thickened portion of bone has been suggested,\textsuperscript{2,11,18} but has not been reported previously. Some authors have stated that the densely adherent dura would make such a procedure extremely hazardous.\textsuperscript{2,18} However, our patient had no dural adhesion to the involved bone at operation.

Although our patient has done well for 3 years postoperatively, we are unable to conclude that the hysteria in our case originated from the HFI. Nevertheless, CT scanning did demonstrate compression of the left frontal lobe.

The incidence of HFI has been reported to be 1.44% by Moore,\textsuperscript{14} 4.79% by Salmi, \textit{et al.},\textsuperscript{17} and 5% by Schüller.\textsuperscript{18} An incidence of 15\%,\textsuperscript{20} 62\%,\textsuperscript{6} and 72\%\textsuperscript{4} has also been recorded in elderly women, and of 25\%\textsuperscript{4} and 42\%\textsuperscript{14} in emotionally disturbed women. Hyperostosis

T. Hasegawa, \textit{et al.}

\textit{J. Neurosurg. / Volume 59 / October, 1983}
Unilateral hyperostosis frontalis interna

frontalis interna is not uncommon among elderly European and American women, but is seldom found in men. Among Japanese, HFI is very rare; it is therefore possible that racial differences account for the lower incidence of HFI in Japan.

References


Manuscript received December 30, 1982.
Address reprint requests to: Takeshi Hasegawa, M.D., Department of Neurosurgery, School of Medicine, Kanazawa University, 13-1, Takaramachi, Kanazawa, 920 Japan.