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Hyperostosis Frontalis Interna in a Child With Severe Traumatic Brain Injury

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Abstract

Hyperostosis frontalis interna is an unexplained irregular thickening of the inner table of the frontal bone. Hyperostosis frontalis interna was first identified in 1719 by Morgagni as a symptom of a more generalized syndrome characterized by virilism and obesity. Most current studies have shown hyperostosis frontalis interna to be a sex- and age-dependent phenomenon, and females manifest a significantly higher prevalence of hyperostosis frontalis interna than males. In this article, the authors report the clinical case of hyperostosis frontalis interna in a 7-year-old child who had severe traumatic brain injury in the past; review the related literature; and discuss the clinical, radiological, and therapeutic features of this condition.

Keywords

hyperostosis, benign bone lesions, frontal bone, child, brain trauma

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Hyperostosis frontalis interna is a morphological feature of the frontal bone that usually presents as single or multiple bilateral nodules on the inner lamina, while most of the diploe and external lamina of the bone remain unaffected.[1] Hyperostosis frontalis interna is encountered more frequently in postmenopausal females and is seldom found in males.[2] Hyperostosis frontalis interna is usually asymptomatic; it is an incidental finding in X-ray, cranial computed tomography (CT), or magnetic resonance imaging (MRI) studies. Here, the authors report a surgically treated case of hyperostosis frontalis interna in a 7-year-old boy with severe traumatic brain injury, review the previously reported cases, and discuss the clinical management and prognosis of this disease.

Case Report

A 2-year-old boy was transferred to the emergency department approximately 4 hours after having severe head trauma with loss of consciousness. On physical examination, he had a right temporoparietal scalp laceration and subgaleal hematoma; the child had a status of decerebrate rigidity. His Glasgow Coma Scale score was 3. Brain CT showed intraventricular hemorrhage and subarachnoid hemorrhage (Figure 1A).

An external ventricular drain was performed. The patient's postoperative clinical course was uneventful. The boy was still unconscious, and a tracheotomy was intended. One week after surgery, cranial CT revealed that the ventricle hemorrhage had disappeared (Figure 1B), and the drain tube was removed. The patient remained unconscious.

One month later, repeat head CT showed dilation of the entire ventricular system associated with communicating hydrocephalus (Figure 1C). A ventriculoperitoneal shunt insertion was recommended (Figure 1D). His consciousness was gradually recovered, and he was awakened 2 months later. The patient was discharged.

At 2-year follow-up, he showed hypophrenia and dysphasia, which was diagnosed according to the diagnosis criterion of mental retardation established by the World Health Organization in 1985 and the "S-S checkup list of dysphasia." The neuroimaging findings revealed that the lateral ventricles were smaller than before (Figure 1E). The electroencephalogram revealed frequent, irregularly slow waves in the bilateral frontotemporal region.

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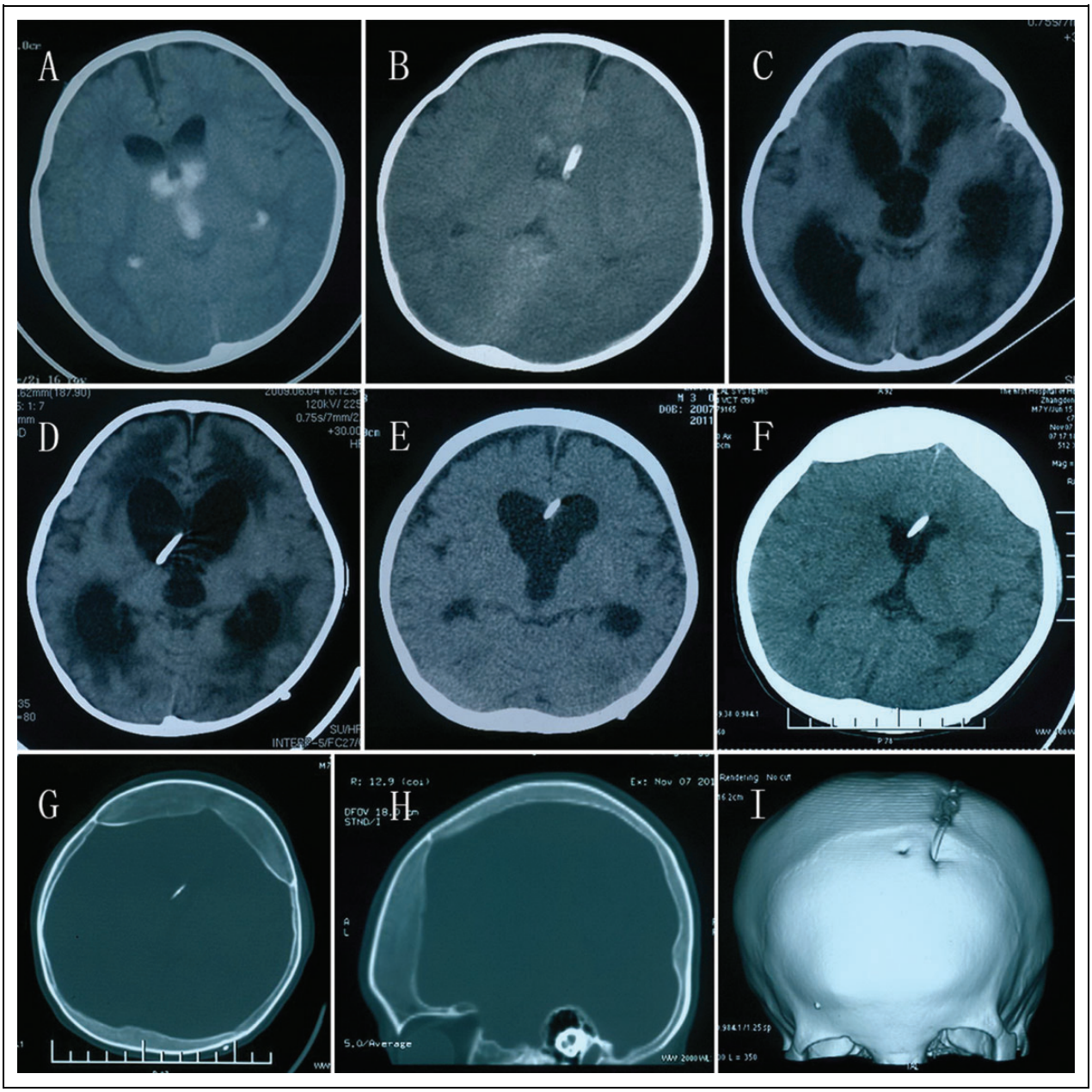

Figure 1. A, Head computed tomography (CT) illustrating the intraventricular hemorrhage. The hematoma completely filled the third ventricle and had produced hydrocephalus. B, Postoperative intraventricular external drainage resulted in a significant decrease in the intraventricular hemorrhage. C, Enlargement of both the lateral ventricles with surrounding white matter edema is consistent with communicating hydrocephalus. D, Postoperative ventricle peritoneal shunt. E, The ventricular system was shrinking, and the periventricular edema disappeared. F-I, Axial and sagittal CT images showing severe, bilateral thickening of the inner table of the frontal bone with cortex compression. I, The outer table of the frontal bone was intact.

Five years after surgery, the child was readmitted to our department due to headache, vomiting, imbalance, inappropriate behavior, and loss of interest. The symptoms had been observed over a period of 3 months. The patient had not developed a frontal extra-axial hemorrhage in the past 5 years. There was bilateral optic disc swelling, with hemorrhages of the optic nerve head and surrounding retina (Figure 2A). Routine investigations were normal. Serum cortisol, prolactin, progesterone, estradiol, T_3, T_4, thyroid-stimulating hormone, and glucose tolerance test were within the normal limits. The ventriculoperitoneal shunt catheter was blocked. Brain CT showed severe, bilateral thickening of the inner table of the frontal bone with cortex compression (Figure 1F-I).

During the operation, the dura was found to be thick, and the frontal bone was hyperostotic. The inner table was honeycombed and red (Figure 2B and C), and the intracranial pressure was elevated. To achieve the decompression, a complete resection of the mass was performed, and the dura was opened

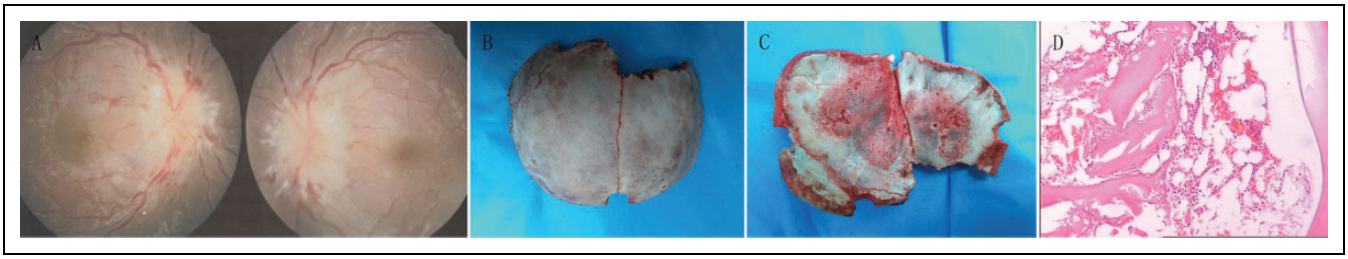

Figure 2. A, There were bilateral papilledema with hemorrhages. B and C, Intraoperative appearance of the frontal bone. D, Microscopic view of the histological specimen of hyperostosis frontalis interna. Hematoxylin and eosin staining; original magnification, ×100.

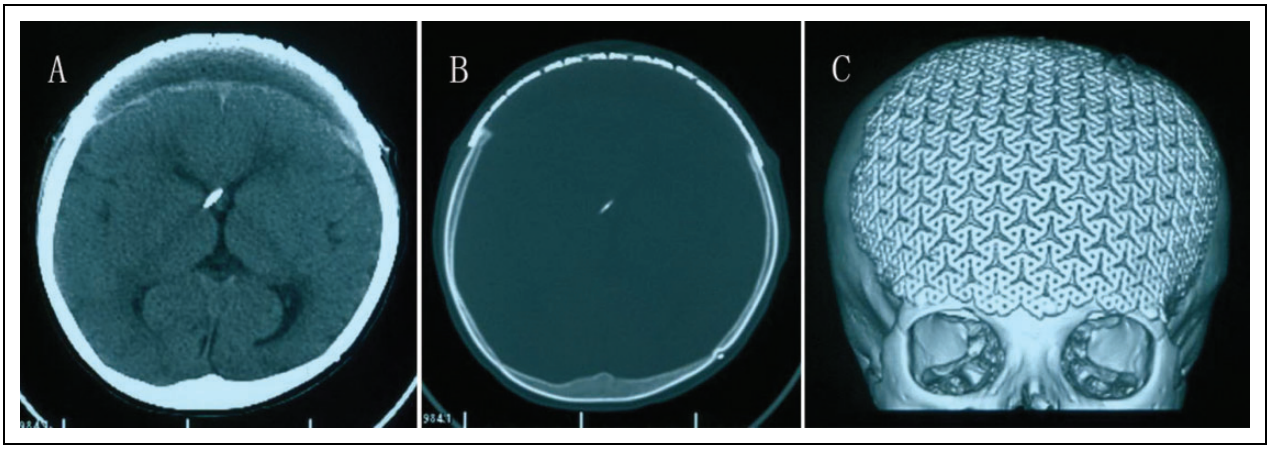

Figure 3. A-C, Postoperative brain computed tomography (CT) demonstrating that the frontal bone was removed and replaced with titanium mesh to relieve the brain compression.

in a star fashion. Three-dimensional image formation was used to shape the titanium mesh for the skull neoplasty.

The mass was immersed in formalin for study of the histological appearance of the mass with mineralized lamellar bone. Vascular channels were lined with osteoblasts. The pathological diagnosis revealed a hyperostosis frontalis interna (Figure 2D).

The child made a good recovery after the operation. The forehead had a good contour, and postoperative radiographs demonstrated that the bone grafts remained intact and no evidence of recurrence of the mass. Baseline radiography of the skeletal system was performed and showed no additional foci of hyperostosis (Figure 3A-C).

Discussion

Hyperostosis frontalis interna is an overgrowth of bony tissue in the inner plate of the frontal bone and has been documented in the medical literature for over 300 years.[3] It is currently considered an independent phenomenon in some studies. The estimated incidence of hyperostosis frontalis interna in the general population is 5% to 12%.[4] Hyperostosis frontalis interna has not been reported in an asymptomatic patient younger than 10 years of age; hence, the present patient is the first case in the Medline database.

The etiopathogenesis of hyperostosis frontalis interna remains a mystery. The most acceptable hypothesis regarding hyperostosis frontalis interna etiology is hormonal influence on bone growth. For example, estrogen stimulation could play a part in the emergence of hyperostosis frontalis interna and explain its predominance among females.[5] In our case, the hormone levels were normal, and interestingly, the hyperostosis frontalis interna was found after severe trauma. With the existing findings, it is difficult to address whether the trauma was related to hyperostosis frontalis interna. To the best of our knowledge, there have been no previous reports in the literature associating hyperostosis frontalis interna with brain trauma.

Hyperostosis frontalis interna usually does not induce any symptoms. The associated signs and symptoms are generally nonspecific and benign, but they can cluster together in some cases, giving rise to various syndromes. The most frequently presented complaints were Morgagni syndrome (headache, obesity, virilism, and hypertrichosis), Stewart-Morel syndrome (obesity and neuropsychiatric symptoms), and Troell-Junet syndrome (acromegaly, toxic goiter, and diabetes mellitus).[2,6] Our patient had imbalance which, to our knowledge, has not been mentioned previously.

In some severe cases, hyperostosis frontalis interna leads to compression of soft tissue, dural irritation, and brain atrophy because of the thickening of the skull and decrease in the intracranial volume. It has been established that hyperostosis frontalis interna can cause diverse psychiatric disturbances such as aggressiveness, paranoia, or depression.[7] Our case tends to confirm that due to extensive hyperostosis frontalis interna, frontal lobe compression can lead to cognitive

impairment and psychiatric disorders. The clinical symptoms and signs indicated the elevated intracranial pressure, which was not described in all the previous reports.

Hyperostosis frontalis interna is commonly an incidental finding in X-ray, CT, and MRI studies. Radiographically, hyperostosis frontalis interna is seen as a thickened inner table. Computed tomography scans, which provide the radiologist with enough information to distinguish hyperostosis frontalis interna from other bony growth, are better diagnostic tools for hyperostosis frontalis interna. In our case, the authors found bilateral frontal inner tables with diffuse uneven thickening. The intraoperative appearance of the frontal inner table was irregular nodular thickening.

Histologically, the process of hyperostosis frontalis interna is thought to be a deposition of new bone primarily on the inner table and a progressive development of diploe.[8] Hyperostosis frontalis interna is characterized by remodeling of the inner table of the frontal bone into a cancellous phenotype. The histology best represented a diagnosis of hyperostosis frontalis interna in our case.

There is no definite consensus on the treatment of hyperostosis frontalis interna. Hyperostosis frontalis interna is a benign process, and the majority of patients appear to be asymptomatic; thus, conservative observation can be used in these cases. If hyperostosis frontalis interna leads to headaches, neurologic symptoms, psychiatric disorders, and cognitive impairment, the surgical excision of the thickened portion of the bone is the only method of treatment to relieve the symptoms. Our patient underwent a complete resection of the mass. Postoperatively, the patient had an uneventful recovery, and the cognitive impairment was improved. A long-term follow-up was necessary.

Conclusion

Hyperostosis frontalis interna is currently regarded as an independent entity and starts to appear at a much younger age. The authors described a case in a 7-year-old boy, presenting imbalance, cognitive impairment, headache, vomiting, and papilledema with hemorrhages; this is the first such case reported in the literature. The etiology, however, remains unclear, and it is difficult to prove the correlation between hyperostosis frontalis interna and brain injury. If present, the surgical decompression can be an effective treatment method. Our case confirms that the clinical presentation was elicited due to compression of the involved cortex area. A further study of this disease should be performed in the future.

Authors' Contribution

Yaxiong Li contributed to conception and design, analysis and interpretation, and writing of the draft. Xin Wang contributed to acquisition and analysis. Yan Li contributed to acquisition of data and patient care.

Authors' Note

Patient consent to publish was obtained.

Declaration of Conflicting Interests

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Ethics approval

Ethics approval was provided by the ethics committee of Hebei Medical University.

Novel Insights

1. This is the first known report in the literature of a child with hyperostosis frontalis interna also having a history of severe brain trauma.
2. The clinical presentations, including imbalance, headache, vomiting, and papilledema with hemorrhages were not described in all the previous reports.
3. Surgical decompression can be an effective treatment for patients with hyperostosis frontalis interna who are having cognitive impairment, headache, and papilledema.

References

1. She R, Szakacs J. Hyperostosis frontalis interna: case report and review of literature. *Ann Clin Lab Sci*. 2004;34(2):206-208.
2. Raikos A, Paraskevas GK, Yusuf F, et al. Etiopathogenesis of hyperostosis frontalis interna: a mystery still. *Ann Anat*. 2011; 193(5):453-458.
3. May H, Peled N, Dar G, et al. Hyperostosis frontalis interna: criteria for sexing and aging a skeleton. *Int J Legal Med*. 2011;125(5): 669-673.
4. May H, Peled N, Dar G, et al. Hyperostosis frontalis interna: what does it tell us about our health? *Am J Hum Biol*. 2011;23(3): 392-397.
5. Attanasio F, Granziera S, Giantin V, et al. Full penetrance of Morgagni-Stewart-Morel syndrome in a 75-year-old woman: case report and review of the literature. *J Clin Endocrinol Metab*. 2013; 98(2):453-457.
6. Nikolić S, Djonić D, Zivković V, et al. Rate of occurrence, gross appearance, and age relation of hyperostosis frontalis interna in females: a prospective autopsy study. *Am J Forensic Med Pathol*. 2010;31(3):205-207.
7. Devriendt W, Piercecchi-Marti MD, Adalian P, et al. Hyperostosis frontalis interna: forensic issues. *J Forensic Sci*. 2005;50(1): 143-146.
8. Chen OI, Murthy AS. Benign hyperostotic mass of the frontal bone. *J Craniofac Surg*. 2012;23(1):e27-e30.