

Hyperostosis cranii ex vacuo

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Section: Musculoskeletal system

Case Type: Clinical Cases

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Patient: 36 years, female

Clinical History:

A 36 year old female, born with a myelomeningocele, presented with a paralysis of the right facial nerve.

Imaging Findings:

The patient was born with a myelomeningocele. The patient has severe complications of the neural defect, including mental retardation, paraplegia and urinary incontinence. Because of a non-communicating hydrocephalus, she was treated with a ventriculoperitoneal shunt in her early childhood. Recently she developed paralysis of the right facial nerve. A CT examination of the brain was performed to exclude a central cause of the paralysis.

The ventriculoperitoneal shunt was correctly positioned within the right lateral ventricle. There was hydrocephalus, microcephaly and descent of the cerebellar tonsils within an enlarged foramen magnum (not shown). There were no imaging signs of a central paralysis of the right facial nerve (bleeding, ischemia, mass lesions). The most remarkable finding on the examination is the diffuse thickening of the calvarian bone. This thickening is most pronounced at the frontal bone (thickening up to 2.4 cm), but also affects the parietal, temporal and occipital bones. The other bones are not involved. The thickening itself consists of a concentrically layered expansion of the diploe. The pattern of calvarian thickening and the history of a ventriculoperitoneal shunt placed in the childhood suggest the diagnosis of hyperostosis cranii ex vacuo. This condition could not be linked with the development of a paralysis of the facial nerve.

Discussion:

Hyperostosis cranii ex vacuo can be defined as the diffuse thickening of the diploe of the calvarian bones. The calvarian bones consist of the frontal, parietal, temporal and occipital bones. The other skull bones are not affected. The thickening is contributed to a widening of the diploe, as in our case.

The disease was first described in 1965 by Emery in a case of a patient with ventricular shunting. Since then, a few dozen of cases have been published. In most of these cases, the patients were successfully treated for hydrocephalus in their childhood with a ventricular shunt. Another case is described after choroid plexus coagulation and one article suggests the phenomenon to be more general pointing to a relationship between dynamic changes in brain size and skull thickness.

In one series of 230 patients operated for hydrocephalus between 1938-1967, 7 cases were found after a follow up between 3 years and 2 months and 8 years and 6 months, thus leading to an incidence of 3% in patients treated for hydrocephalus.

A relationship with the severity of hydrocephalus, microcephaly, early closure of the sutures and intracranial hypotension has been suggested. In case of a normal development of the skull, outward pressure by the cranial content stimulates intramembranous bone growth along the cranial sutures. Due to chronic intracranial hypotension, the growth of the calvarian bone seems to occur at the inner table, leading to a widening of the diploe.

Differential diagnosis of thickening of the calvarian bones includes healed rickets, severe longstanding anaemia,

thalassemia, craniometaphyseal dysplasia, Paget's disease, fibrous dysplasia and hyperostosis frontalis interna.

Differential Diagnosis List: Hyperostosis cranii ex vacuo

Final Diagnosis: Hyperostosis cranii ex vacuo

References:

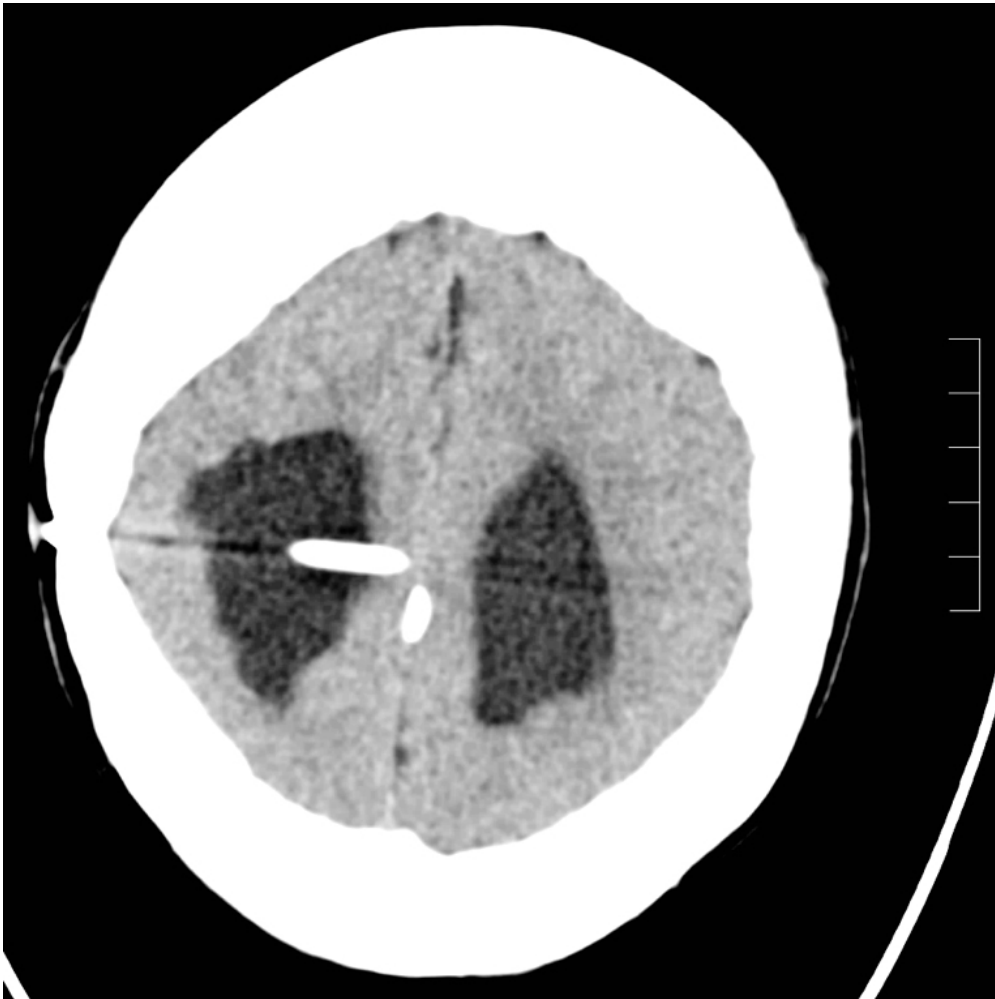
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Figure 1

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Description: Microcephaly, hydrocephalus, correct position of the tip of the ventriculoperitoneal shunt within the right lateral ventricle and the diffuse thickening of the parietal bones. **Origin:**

Figure 2

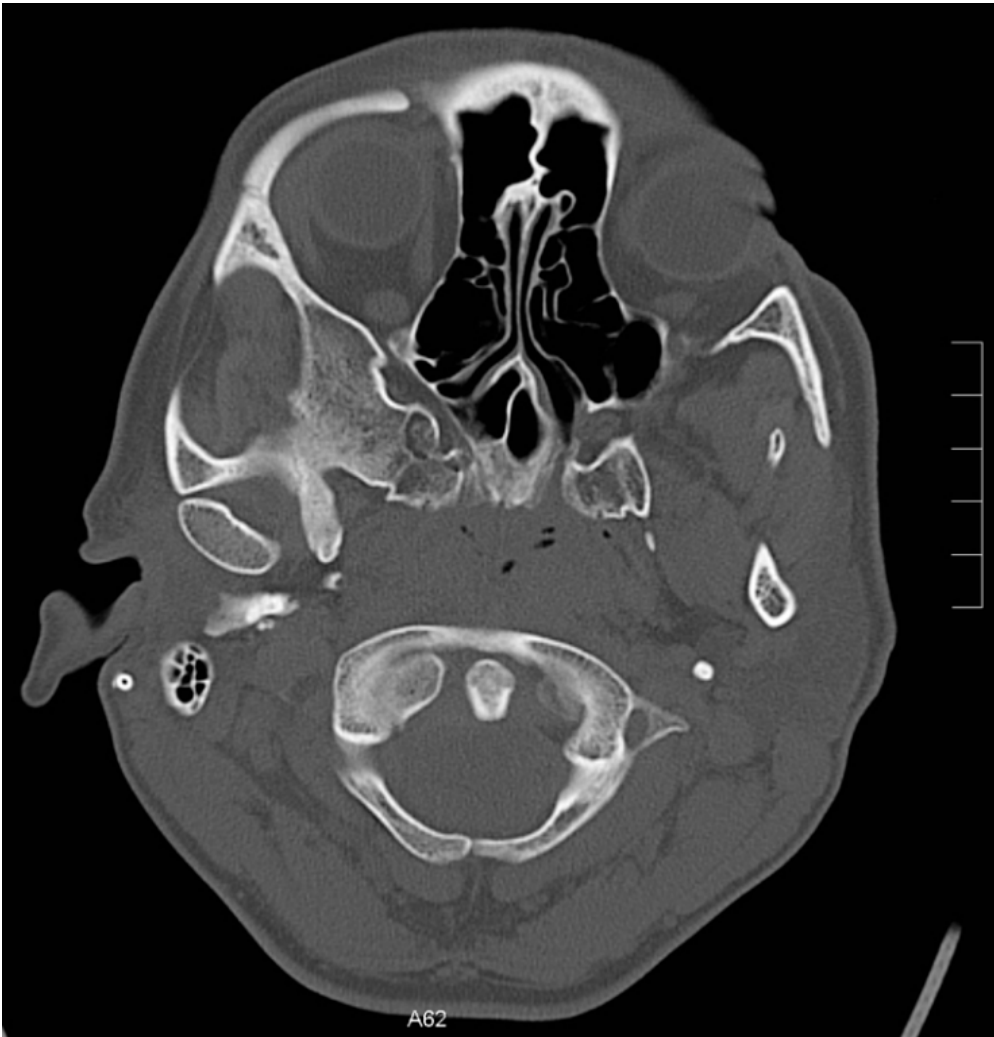
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Description: Notice the concentrically layered expansion of the diploe of the frontal, temporal and occipital bones. **Origin:**

Figure 3

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Description: The mandibular, zygomatic and sphenoidal bones show a normal thickness. Notice the fusion defect of the posterior arch of C1. **Origin:**

Figure 4

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Description: Absence of a mass lesion at the internal auditory canal (asterisk) or brain stem, thickening of the frontal and temporal bone. **Origin:**