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Vascular Involvement in Cranial Hyperostosis

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Various skeletal dysplasias known to produce cranial hyperostosis include osteopetrosis, craniometaphyseal dysplasia, hereditary hyperphosphatasia, Van Buchem disease, and Engelmann disease [1]. Neurologic complications such as optic atrophy, facial paralysis, or deafness may result from progressive encroachment on the cranial foramina due to bony overgrowth at the base of the skull. Narrowing and compromise of the arteries and veins at the skull base by the same mechanism is not as well known or documented. This paper deals with the computed tomographic (CT) and angiographic findings in a child with hyperostosis of the skull base due to craniometaphyseal dysplasia.

Case Report

A 17-month-old girl was the product of a normal spontaneous vaginal delivery. At age 3 months the patient had been initially evaluated for rapidly increasing head growth. Three CT scans of the head obtained between age 3 months and 17 months demonstrated thickening of the skull base, prominent cortical sulci, and normal-sized ventricles. During this 14 month period, there was no evidence of progression of these CT findings except for minimally increased prominence of the cortical sulci. Skull films at age 3 months showed marked hyperostosis of the base of the skull, maxilla, and mandible (fig. 1).

The patient had a history of normal motor development. Medical history was remarkable for three episodes of otitis media at age 3 months, 10 months, and 16 months. There was no history of any visual or neurologic deficits. One week before the latest admission the neurosurgeon auscultated a bruit over the frontal region. The patient was admitted to evaluate the cause of the bruit in the presence of enlarging head circumference and to rule out the possibility of a dural arteriovenous malformation.

Physical examination revealed a 16½-month-old, well developed, alert female infant. Her height was 73 cm (less than 5th percentile); weight 9.5 kg (20th percentile); and head circumference, 54 cm (greater than 98th percentile). Her head was scaphocephalic with prominent veins over the forehead. A soft bruit was heard over the frontal region and there was evidence of hypertelorism. The anterior fontanelle was 2 cm in diameter, open and nonbulging; the posterior fontanelle was closed. Ophthalmologic examination was normal. The tympanic membranes were erythematous and exhibited decreased mobility. The patient breathed noisily through the mouth.

The cardiopulmonary and abdominal examination was normal, her arms and legs showed full ranges of motion, and the neurologic examination was also normal.

Skull radiography on this examination revealed frontal bossing with marked hyperostosis of the skull base, maxilla, and mandible. Selective internal carotid arteriography demonstrated narrowing of the petrous parts of the internal carotid arteries bilaterally (figs. 2A and 2B). The venous phases demonstrated tapering of the sigmoid sinus with narrowing of the jugular veins bilaterally at the origins (figs. 2C and 2D). There was enlargement of the left superior ophthalmic vein and angular facial vein. The intracranial venous sinuses were patent. CT examination revealed a thickened calvaria, most prominent at the skull base (fig. 3). The mastoids were not pneumatized. The frontal subarachnoid space and the cortical sulci were enlarged, although the ventricles were of normal size. Films of the spine and other bones revealed no evidence of sclerotic change. Routine laboratory values were normal, and there was no family history of skeletal or neurologic abnormalities.



Fig. 1.—Lateral skull, age 3 months. Hyperostosis of skull base, maxilla, and mandible.

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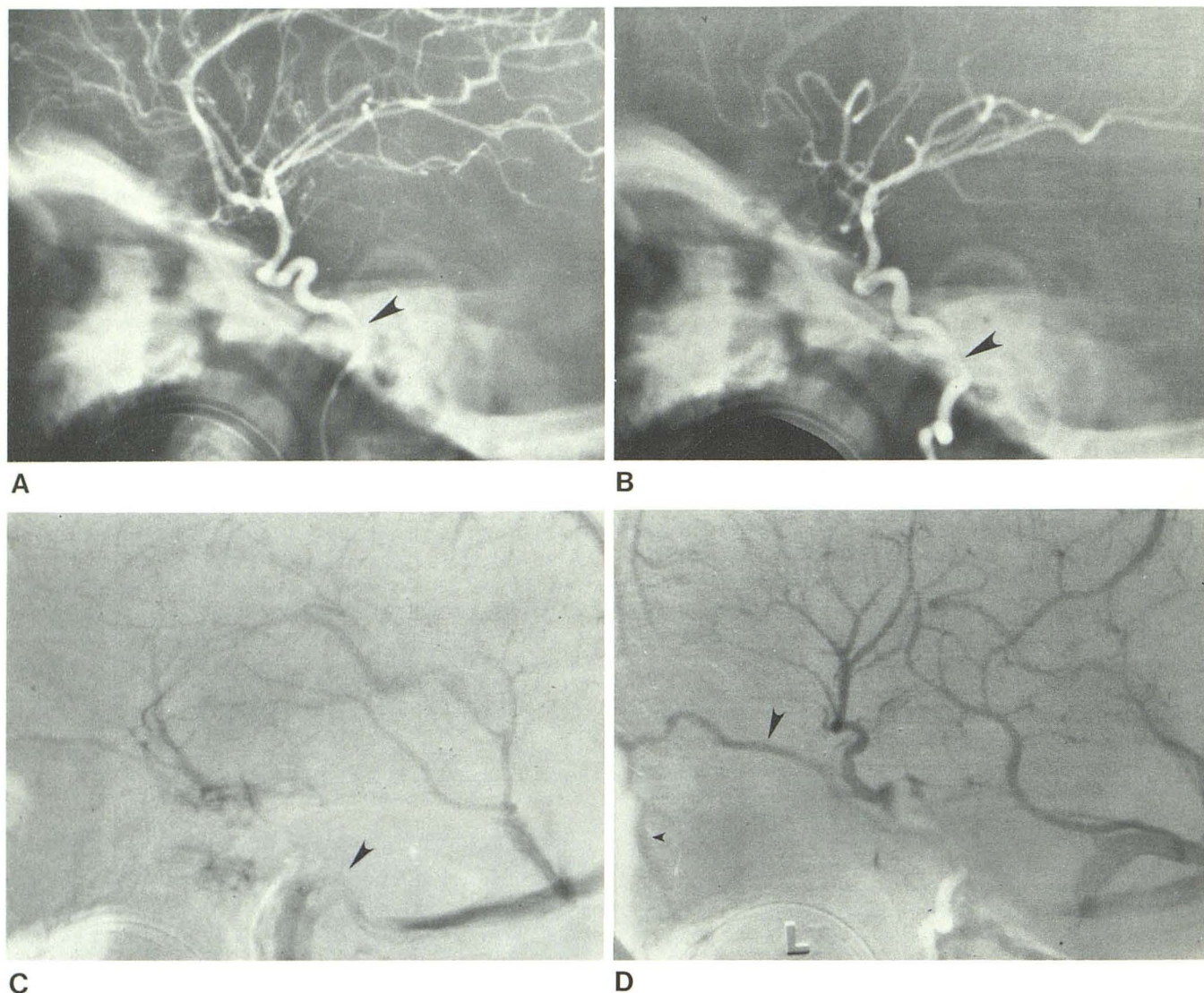


Fig. 2.—**A** and **B**, Bilateral internal carotid angiograms, age 17 months. **A**, Narrowing of petrous segment of right internal carotid artery (arrowhead). **B**, More severe narrowing on left (arrowhead). **C** and **D**, Venous phase after

internal carotid injections. **C**, Right side. Narrowing at origin of jugular vein (arrowhead). **D**, Left side. Enlarged superior ophthalmic vein (large arrowhead) and angular facial vein (small arrowhead).

Discussion

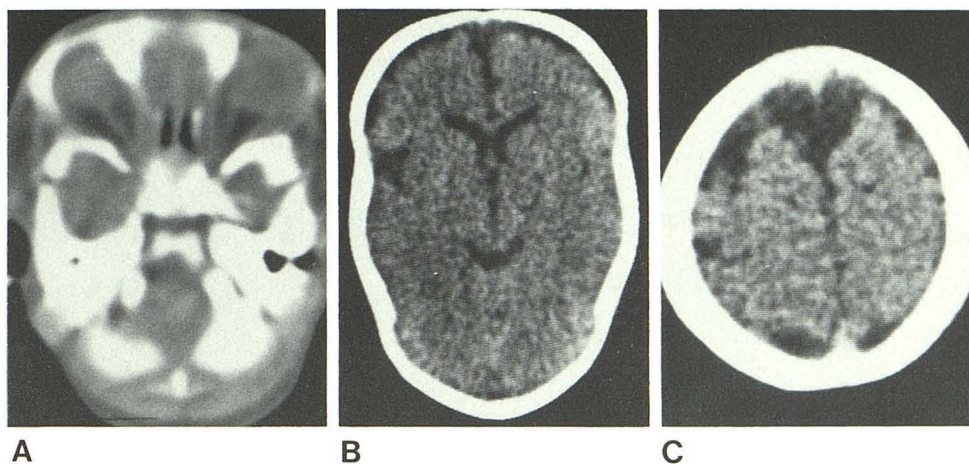
The findings on the skull radiographs in this patient are consistent with craniometaphyseal dysplasia, although virtually identical changes may be seen in osteopetrosis. Additional radiographic and clinical considerations favored the diagnosis of craniometaphyseal dysplasia. Reported cases of the infantile form of osteopetrosis usually have a systemic component manifested by anemia and hepatosplenomegaly. Neurologic manifestations such as facial palsy or deafness usually occur very early [2-4]. The "malignant" or infantile variety of osteopetrosis usually exhibits horizontal bands of hyperostosis throughout the spine. Although neurologic or vascular compromise resulting from cranial hyperostosis may be seen in either condition, none of these other features of osteopetrosis were present in this patient. Mandibular

hyperostosis, reportedly unusual in osteopetrosis, is a prominent feature of craniometaphyseal dysplasia [5] and is evident in this case.

The possibility of arterial narrowing secondary to osteopetrosis has been alluded to [6], but has not been well demonstrated. One angiographic study of a patient with osteopetrosis described an apparent arteritis that may have been associated with arterial stenosis [7]. Carotid artery involvement by craniometaphyseal dysplasia has not been previously reported. Although this patient did not show any evidence of arterial disease clinically, it is possible that further narrowing of the basilar foramina may produce ischemic symptoms later in life.

The basilar hyperostosis also produced narrowing of the veins in the jugular foramina. Such venous outflow obstruction results in a reversal of the normal flow pattern through

Fig. 3.—Cranial CT. **A**, Thickening of skull base. **B**, Normal ventricular size with prominent frontal subarachnoid space. Frontal bossing. **C**, Dilated cortical sulci.



the emissary veins [8]. Reversal of flow in the ophthalmic vein in our patient resulted in the unusual clinical presentation of craniometaphyseal dysplasia with a frontal bruit and prominent scalp veins. A similar form of venous compromise has been described as a consequence of the small jugular foramina of achondroplasia [8, 9].

CT demonstrated the expected dense, expanded bone of the calvaria and skull base. Dilatation of the cortical sulci, however, is not a well recognized feature of osteopetrosis or craniometaphyseal dysplasia. Although cerebral atrophy would be a likely etiology, the CT changes may also reflect gyral compression secondary to increased cerebrospinal fluid (CSF) volume. Recent reports suggest that wide cortical sulci may result from a disturbance of CSF absorption caused by elevated venous retrograde pressure [8, 9]. The increased pressure in the dural sinuses presumably impedes the flow of CSF across the arachnoid villi, thus expanding CSF volume [10–17]. This mechanism constitutes an attractive hypothesis to account for the association of jugular venous obstruction and dilated cortical sulci in this case, and has been suggested as a possible etiology for similar findings in achondroplasia [9]. The use of dynamic radio-nuclide and metrizamide CSF flow studies in such patients may help to elucidate the consequences of venous obstruction.

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