Cranial asymmetry secondary to unilateral hemispheric damage during late childhood

Case report

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Skull abnormalities such as unilateral hypertrophy of skull thickness, enlarged sinuses, and elevated petrous ridge with contralateral body hemiatrophy are commonly associated with hemispheric damage that occurs during infancy. The present case emphasizes that such changes may be associated with cortical damage during late childhood.

KEY WORDS • skull • infantile hemiplegia • cerebral vascular accident • epilepsy

In 1868, Cotard reported that the skull and its sinuses may become hypertrophied on the side of an atrophic cerebral lesion. By 1889, the symptom complex of “infantile hemiplegia” had become established, and in 1897, there was enough literature amassed to allow Sigmund Freud to report a complete summary of the topic. In 1905, Taylor was the first to observe that in infantile hemiplegia there is frequently an acute onset of hemiplegia associated with an infectious episode in the first few months of life. The first comprehensive description of the radiological correlates was described by Dyke, et al., in 1933. They stated that in most cases the cerebral trauma that led to changes in the skull and central nervous system (CNS) occurred within the first 15 months of life. This syndrome of infantile hemiplegia has been referred to as the Dyke-Davidoff-Masson syndrome. Throughout the years, it has been generally accepted that patients with body hemiatrophy and contralateral skull hypertrophic changes have suffered damage to the sensorimotor cortex during infancy. To emphasize that such changes may occur as sequelae to CNS damage in later childhood, we present the following case.

Case Report

This 24-year-old, right-handed woman was a product of an uncomplicated pregnancy and delivery. Her childhood milestones were normal, as documented by regular examinations by a pediatrician. At the age of 9 years, 5½ months, she developed an infected appendix, which ruptured prior to surgery. She subsequently developed a p6stoperative peritonitis and subphrenic abscess. Before the abscess was drained, she became severely febrile and developed a tonic-clonic seizure followed immediately by left hemiplegia. Skull films at that time revealed a normally developing, symmetrical cranium, and an angiogram showed diminished flow through the middle cerebral vessels on the right side. She recovered from her abdominal problems, and was discharged from the hospital with a dense left hemiplegia and frequent focal motor and sensory seizures.

At the age of 24 years, the patient was referred to the University of Washington for surgical removal of her right parietal and frontal epileptogenic foci. Neurological examination at that time demonstrated no visual field defect, but marked hemiparesis of the left extremities (arm greater than leg) with only finger flexion and extension of the hand. She showed deficits in graphesthesia, stereognosis, and two-point discrimination on the left; however, she did not demonstrate signs of left-sided neglect. Her left arm and leg were 2 and 1 in. shorter, respectively, than the right. Radiological examination of the skull (Fig. 1) demonstrated massive hypertrophy of the right frontal and ethmoid sinuses, and hypertrophy of the right
temporal and parietal bones, with elevation of the greater wing of the sphenoid and petrous bones. These findings are typical of changes observed in patients with infantile hemiplegia.

**Discussion**

By the 7th year of life, the orbits in the child are almost as large as in the adult; the cribiform plates of the ethmoid, the body of the sphenoid, and the petrous parts of the temporal bones and the foramen magnum have reached their full size. At puberty, there is a rapid increase in the rate of growth in the frontal and facial regions owing to an increase in size of the air sinuses.\(^4,10\) There is a consensus in the literature that the skull changes normally associated with hemiatrophy are indicative of cerebral damage that occurred during infancy.\(^2,4,6,9,11\) However, in 1948, McRae\(^7\) reported that if lesions of the brain occurring within the first 2 years of life produce a difference in the volume between the two cerebral hemispheres, they will cause asymmetrical growth of the two sides of the skull, presumably because the brain is growing at its maximum rate at this time. Similar lesions occurring between the ages of 2 years and puberty, when brain growth is proceeding more slowly, may produce asymmetrical head growth if the cerebral lesion is of gross proportions. Lesions that occur after puberty (when the growth of the brain is fairly complete) will cause no asymmetry of the skull. Unfortunately, McRae's observations have been overlooked. Even Penfield and Jasper\(^8\) later implied that hypertrophic skull changes indicated cerebral trauma during infancy.

The patient who we are reporting had a normal infancy, and normal growth and development until the age of 9 years, at which time she suffered a cerebral vascular accident involving the middle cerebral artery on the right. Only later did she develop skull changes usually associated with earlier trauma. This has significance for the neurological surgeon, because of the importance in determining whether the patient's CNS trauma occurred during early infancy. At that age, the brain has much greater capability in using the contralateral hemisphere to compensate for functions lost in the injured hemisphere. Knowledge of the age at time of insult may modify the surgeon's approach to removing the epileptogenic focus. In addition, it underscores the need for careful evaluation of skull x-ray films when evaluating seizure disorders.

**References**

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5. Freud S: Die Infantile Cerebrallachmungen. Vienna: Holder, 1897

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