Idiopathic Bilateral Sigmoid Sinus Occlusion in a Child

Case Report

KENNETH R. SMITH, JR., M.D.
Section of Neurological Surgery, Department of Surgery, St. Louis University School of Medicine, St. Louis, Missouri

Thrombosis of major dural sinuses has often been associated with hemorrhagic cerebral infarction, intracranial hypertension, and death. In one series before the antibiotic era, sigmoid sinus thrombosis was a significant factor in one of every 300 deaths (German autopsy material 1900–1925), and approximately one patient of 200 with otitis media developed thrombosis of the sigmoid sinus. In the past decade, otogenic sigmoid sinus thrombosis has become a rarity, although still associated with a mortality of about 25%. Cavernous sinus thrombosis secondary to infection remains an occasional neurosurgical problem, but in the majority of sinus thromboses today the cause is obscure or is related to trauma, pregnancy, or the puerperium.

Bilateral, idiopathic, essentially asymptomatic occlusion of the sigmoid sinuses is extremely rare. Angiographic studies of a patient with such a condition have been done and are the basis of this report.

Case Report

A 4½-year-old boy was admitted to Cardinal Glennon Memorial Hospital on October 5, 1967, because of an enlarged head since his first year and increasingly dilated veins of the scalp since his second year.

He had been born after an uneventful pregnancy, but was hospitalized for 2 weeks after birth because of jaundice, fever, and an enlarged liver; at that time a cardiac murmur was noted. He grew and developed somewhat slowly, although he sat alone at 8 months and walked at 17 months. At the age of 26 months, cardiac catheterization demonstrated aortic stenosis, mitral insufficiency, and pulmonary hypertension, with normal systemic arterial O₂ saturation and no polycythemia. Because the patient was able to function fairly well, it was thought that cardiac surgery should be delayed.

General Examination. When examined in October, 1967, the patient was alert and in no distress. His speech and intelligence were slightly below average. His head measured 58 cm in circumference, and his chest, 55 cm. Veins of the forehead and on the bridge of the nose were larger than normal (Fig. 1 left). An even more striking vascular feature was a large, tortuous vein in the scalp above the left ear going from the mastoid to the posterior zygomatic area (Fig. 1 right). There was a thrill and a loud bruit over the vessel; auscultation elsewhere showed only soft carotid bruits and a loud holosystolic cardiac murmur.

The patient's general examination was otherwise unremarkable except for a height of 42 inches and a weight of 34 pounds. The cranial nerves and funduscopic examinations were normal, as were motor, sensory, and cerebellar functions. Laboratory examinations of the blood and urine were normal. Lumbar puncture was not done.

X-ray Examination. A chest x-ray showed enlargement of the left and right ventricles with pulmonary congestion. X-rays of the head disclosed mild cranial enlargement with no evidence of increased intracranial pressure and no abnormal calcification. The bony vascular channels, including the grooves for both transverse sinuses, appeared normal except for a large channel in the left posterior temporoparietal area.

On October 6, 1967, right brachial and left carotid arteriograms showed normal arterial and deep venous structures with some dilatation of the ventricles (Fig. 2). There was no abnormal arteriovenous anastomosis. In films taken 6 and 7 sec after injection of contrast material, all dural sinuses were well-defined except for the sigmoid si-
nuses and the internal jugular veins, which did not opacify. The transverse sinuses were of normal size and configuration. A Towne projection (Fig. 3 left) demonstrated an abrupt ending of these sinuses. The left transverse sinus was drained by a huge emissary vein, which was the one visible and palpable on physical examination and which produced the abnormal vascular channel on the routine x-rays. This vein coursed superiorly, anteriorly, and inferiorly to drain into the left jugular veins (Fig. 3 right). Other smaller parietooccipital emissary veins were seen to drain the transverse sinuses caudally into the neck (arrow, Fig. 3 right). Anteriorly, several prominent veins drained into

FIG. 1. Infrared photographs demonstrated the prominent veins of the forehead and the upper nasal region (left) and the massive emissary vein that drains the left transverse sinus (in the scalp above the ear, right).

FIG. 2. Left: Left carotid arteriogram outlining normal arteries, with no arteriovenous shunting. The broad sweep of the pericallosal artery indicates mild ventricular enlargement. At the tempo-parieto-occipital junction, a cranial defect is created by the emissary vein (see Fig. 1) which fills with contrast material in the venous phase of the angiogram. Right: X-ray 4 sec later showing normal cortical and deep venous patterns. The left occipito-parietal-emissary vein is just beginning to opacify. There is a prominent venous pattern in the region of the cavernous sinus (arrow).