Sinus Pericranii associated with Hypoplastic Straight Sinus and Persistent Falcine Sinus

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Case Report

Abstract

Sinus pericranii (SP) is a rare vascular abnormality of unknown origin that is defined as an aberrant anastomosis joining the intracranial and extracranial venous systems, usually near the midline. Sinus pericranii (SP) can be formed either by focal venous hypertension and abnormal development of diploic veins (congenital type) or by trauma (acquired type).

Keywords: Sinus Pericranii, falcine sinus, vascular abnormality

Case

A 16-month-old boy, full-term and normally delivered, was brought to the hospital for neurosurgery OPD follow-up, with midline scalp occipital painless swelling since birth. This swelling was increased by crying. However, there was no history of vacuum extraction, no history of discharge or bleeding from it, as well as no history of trauma. On physical examination, the swelling is soft, 2x2 cm in size, not compressible or reducible, and covered by normal skin. MRI showed an extracranial midline mass with a small bony defect, connected to the dural sinus, and associated with the hypoplastic straight sinus and persistent falcine sinus (Figures 1-4).

Discussion

SP is a rare vascular malformation of unknown etiology, defined as an aberrant anastomosis between the intracranial and extracranial venous systems, nearly in the midline. Congenital, syndrome-related, and acquired instances are all possibilities. The nature of SP is benign. Its venous abnormality consists of an emissary intradiploic vein originating from an intracranial sinus, as well as enhanced subgaleal drainage consisting of a network of thin-walled veins forming a varix on the external table of the skull. Furthermore, anastomotic connections can be made up of a single transosseous vessel or several venous systems, which can often reach several centimeters beneath the skull bones, causing severe bony erosion. As a result of contact between intracranial dural sinuses and dilated epicranial venous structures, blood flows into the sinus pericranii and drains into the intracranial venous sinuses. The size of it varies depending on the patient's body posture or, in this example, the Valsalva technique. It is unclear why this
is happening. However, as this case report shows, it appears to be primarily congenital in nature and is commonly associated with other venous anomalies and syndromes\(^6\). Sinus pericranii can be isolated (primary) or accompanied by other malformations (secondary), such as craniosynostosis or intracranial venous anomalies such as dural sinus hypoplasia, particularly straight sinus and persistent falcine sinus, as in this case\(^7,8\). The patient’s parents’ main concern is usually cosmetic, but the most common symptoms are headaches, pressure, or localized pain\(^9\). Furthermore, they may experience severe clinical symptoms such as bradycardia, bradypnea\(^10\), hearing loss, ataxia, or seizures in rare cases\(^5,11\).

![Figure 1: Photos 1 & 2 (T2-MRI), photos 3 & 4 (T1-MRI). All photos are in sagittal plan with post-contrast administration. They show extracranial midline mass with small bony defects (yellow arrows), connected to the dural sinus, and associated with hypoplastic straight sinus and persistent falcine sinus (red arrows).](image)

The lesion tends to grow in size over time, but it has also shown spontaneous remission in rare cases\(^12\). An abrupt change in the lesion’s appearance from soft and painless to firm and painful should cause alarm for both the patient and the clinician. In most situations, the management is surgical intervention. On the other hand, endovascular treatment and sclerotherapy are used sometimes\(^5,13\). CT or MRI can show a vascular lesion with a bone defect and its relationship to an intracranial vessel because of its association with significant diploic erosion. Doppler US is useful for the characterization of the nature of the mass lesion and differentiating different vascular anomalies\(^13\). Moreover, digital subtraction angiography (DSA) is the gold standard approach for diagnosis, it is, however, invasive, exposing children to greater radi-
Role of MRI in the diagnosis of Sinus Pericranii

Sinus pericranii (SP) is a rare vascular anomaly and should be considered a differential diagnosis in patients with a soft, midline subcutaneous scalp mass. It is the cutaneous sign of an underlying venous anomaly. For reaching such a diagnosis, clinical history is very significant, but imaging modalities such as CT and MRI are also used for diagnosis. Furthermore, digital subtraction angiography (DSA) is the gold standard for diagnosis, but it is invasive with more radiation exposure for children. It is used as an endovascular treatment. To confirm the diagnosis, pathological examination is still used. Although cosmetic considerations are the primary concern of the patient, the definitive therapy is surgical removal to avoid significant consequences. Finally, SP has distinct clinical and radiological features that can aid in differential diagnosis and treatment options.

References


