Suez Canal University Medical Journal Vol. 25 (4), 2022
Pages 49-53

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[1]. 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[1-3]. 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[4]. The size of it varies depending on the patient's body posture or, in this example, the Valsalva technique[5]. It is unclear why this

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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].

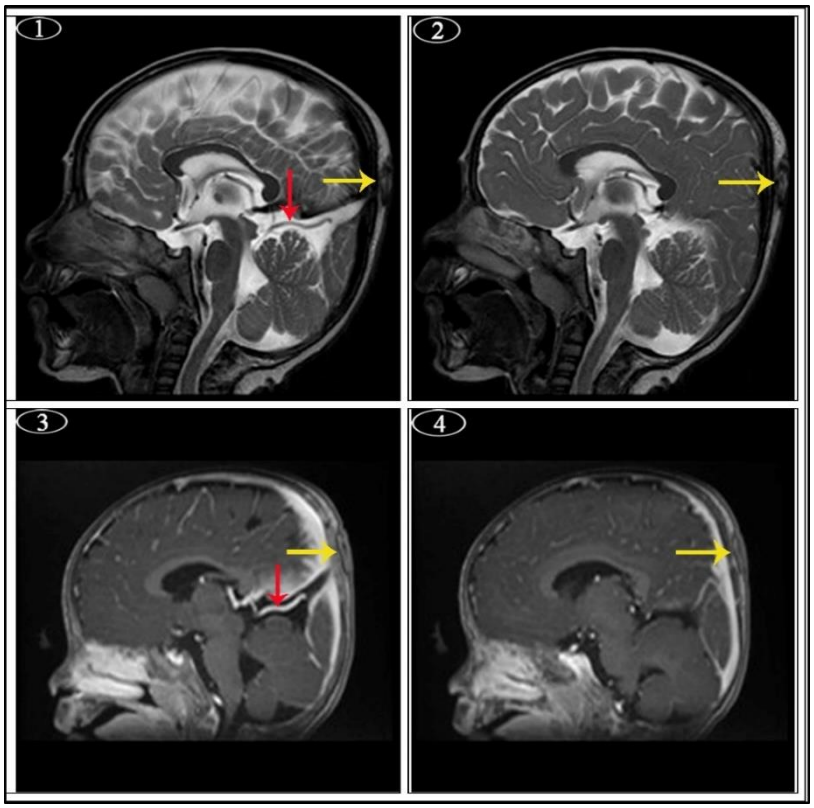

Figure 1: Photos 1 & 2 (T2-MRI), photos 3 & 4 (T1-MRI). All photos are in sagittal plan with post-contrast administration. They show extracranial midline mass with small bony defects (yellow arrows), connected to the dural sinus, and associated with hypoplastic straight sinus and persistent falcine sinus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

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-

ation, but in some cases, DSA is used as a method for treatment[14]. DSA is still important before surgical intervention as it can evaluate the intracranial venous dynamics and the effect of the SP on the venous drainage of the brain. However, nowadays, MR venography can replace conventional angiography for the diagnosis of SP. Based on DSA findings, SP is classified into two forms "dominant" if the primary blood flow is through the SP and bypasses the usual venous outlets, and "accessory" if only a small part of the venous outflow occurs through the extradiploic vessels. This distinction is critical in guiding therapy choices[15]. Sinus pericranii treatment includes conservative measures, endovascular embolization, and surgical removal[5]. However, surgical excision was the treatment of choice. Surgical removal[6,15-18] and direct venous or endovascular occlusion of the SP and its communicating veins[19,20] have been performed for cosmetic reasons or when increased intracranial pressure due to intracranial venous hypertension is a concern[10]. Other indications for SP treatment are to avoid some serious complications such as venous infarction, intracranial hemorrhage, bleeding, and air emboli, and to ameliorate cosmetic problems [21] and spontaneous thrombosis in an SP, which may occur [22]. However, spontaneous regression of sinus pericranii has been reported[23]. Active surgical management of sinus pericranii was justified by the prevention of infection, traumatic air embolism, or massive bleeding[9,24-26] or to decrease discomfort[15,25,27]. Different surgical procedures have been reported because significant hemorrhage may be encountered during the operation[25,26,28]. Such a procedure can control the bleeding from the communication easily with bone wax, gel foam packing, coagulation, and even by air-powered diamond drilling if it is not large enough[28]. This anomaly has a good prognosis. When the scalp flap is just lifted, however, the risk of SP recurrence by recanalization of the interosseous veins is substantially higher than when the skull bone housing the connecting venous channels are detached[29].

Conclusion

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.

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