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Case Report

Lateral sinus pericranii with internal jugular vein communication ☆

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ABSTRACT

An abnormal communication between the extra and intracranial venous structures in the head is called sinus pericranii. The condition usually involves the frontal superior sagittal sinus. Occurrence in the lateral scalp is an uncommon pattern. Its commonest manifestation is scalp swelling, which can be cosmetically unappealing. Radiologic findings play an important role in correctly diagnosing the condition and planning treatment. In this case report, we present the computed tomography findings of a 10-year-old male patient who presented with right scalp swelling. Postcontrast head and neck computerized tomography diagnosed a lateral sinus pericranii with an unusual location in the right scalp that also involves the ipsilateral internal jugular vein. Subsequently, a conservative treatment approach with close clinical follow-up was adopted.

Introduction

Sinus pericranii (SP) is a rare vascular anomaly in which aberrant communicating vascular structures of variable morphology and/or extent bridge the epicranial and intracranial dural venous sinuses [1–3]. Its commonest presentation is nontender scalp swelling, and treatment is often offered for cosmetic reasons [8–10]. It can be congenital or post-traumatic in etiology and commonly involves the superior sagittal sinus in the midline frontal convexity. Lateral location involving the temporal and occipital scalp epicranial scalp venous structures is uncommon [11–14]. But, depending on the patient's wishes or other troubling symptoms (eg, headaches, vomiting), surgical treatment might be pursued.

Imaging plays a pivotal role in correctly diagnosing the condition, separating it from other causes of scalp swellings, and for surgical planning [9,10]. Cross-sectional modalities, such as computed tomography (CT) and magnetic resonance imag-

Abbreviations: SP, sinus pericranii; IJV, internal jugular vein; CT, computed tomography; MRI, magnetic resonance imaging; DSA, digital subtraction angiograph.

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ing (MRI), are better at depicting skull lesions and the full extent of the vascular connections [18].

Case report

A 10-year-old male child from Borena in southern Ethiopia came to our tertiary referral hospital with a chief complaint of head swelling of 6 months duration that progressively increased to attain the size at presentation. He has no known past chronic illnesses or history of head trauma. He does not complain of any other symptoms like headaches, vertigo, or vomiting. He has comparable growth and development with his peers. On physical examination, he appeared comfortable with stable vitals. Anthropometric measurements, including head size, were unaffected. Examination of the head revealed a 10 × 3 cm (total length x width) inverted U-shaped soft, nontender, and non-erythematous right parietotemporal scalp swelling. He also had whitish plaques of tinea capitis on his scalp (already being treated at the time of the examination). The routine Complete blood count was normal. Based on the physical examination alone, cavernoma was the initial clinical impression. Afterward, a skull radiograph (Fig. 1) and post-contrast head and neck CT were done (Figs. 2, 3 and 4). The head and neck CT showed significantly dilated right parietal-temporal-occipital scalp vessels that communicate with the right transverse sinus through prominent emissary veins in the right temporal lobe. Some of the scalp veins also descend to communicate with the right internal jugular vein. Additional tortuous right posterior paravertebral vessels are also seen.

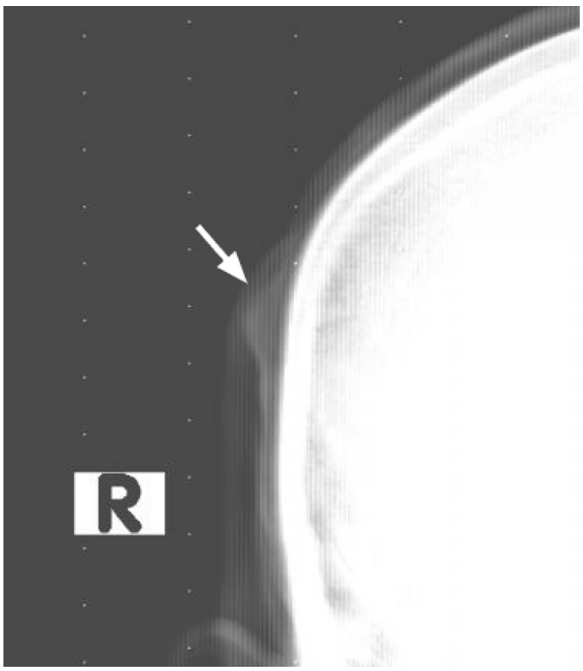

Fig. 1 – Zoomed and narrowly windowed frontal skull radiograph shows a noncalcified focal swelling in the right lateral scalp (white arrow).

Treatment

Given the absence of major neurologic symptoms and the child's relatively young age, it was decided to conservatively treat him with regular clinical follow-ups every 6 months to assess the size of the lesion and other potentially alarming or new clinical symptoms. The patients' parents have been informed of the treatment decision and have agreed to the treatment plan.

Discussion

Sinus pericrania (SP) is a relatively rare venous anomaly in which abnormal communication exists between the venous structures of the scalp and the intracranial compartment. Under normal circumstances, these 2 should never directly communicate [1–3]. The communication formed is responsible for intracranial pressure transmission to the scalp veins and the typical valsalva volume changes elicited on physical examination. Increased intracranial pressure with Valsalva is transmitted to scalp veins, which results in a size increment of scalp veins. The reverse will happen with movements that decrease intracranial pressure, such as sitting up. This abnormal communication can be congenital or acquired, and differentiation between these 2 can be established through histological examination, whereby the former will have endothelial linings and the latter, a collagen lining [4,5]. Both lack a muscular layer consistent with a venous malformation. When it is associated with other venous anomalies or vascular tumors such as systemic angiomas, cavernous hemangiomas, and other skeletal anomalies, a congenital cause can be suggested [1,3]. Trauma to the head, which affects the emissary veins with subsequent occurrence of SP, has also been well described and agrees with the pathophysiology of SP [4,6]. A third form called spontaneous SP has also been described following trivial incidences of increased intracranial pressure like vomiting or coughing [7]. Generally, most cases occur in patients less than age 30, and male predominance has been reported [8–10]. Other than head swelling, which is usually painless but aesthetically unpleasing and thus the major reason for presentation, headache, vertigo, nausea, and vomiting can also occur. Our patient's SP is unlikely to be congenital given the relatively late onset of the swelling, hence considered to be probably acquired. The absence of alarming neurologic symptoms fits the typical clinical scenario.

The morphology and size of the communicating vessels in SP vary and are the main points of interest from a radiologic and surgical point of view. The defining feature of sinus pericranii, as mentioned above, is communication between an extracranial blood-filled venous structure and intracranial venous sinuses. The communicating veins are usually single or multiple varicoid emissary veins or sometimes extensive intradiploic blood-filled sacs that form marked intraosseous connections (sometimes they can erode the calvarium, leaving a thin bony separation between intracranial venous sinuses and the subcutaneous tissue). SPs are usually located along the frontal-overlying superior sagittal si-

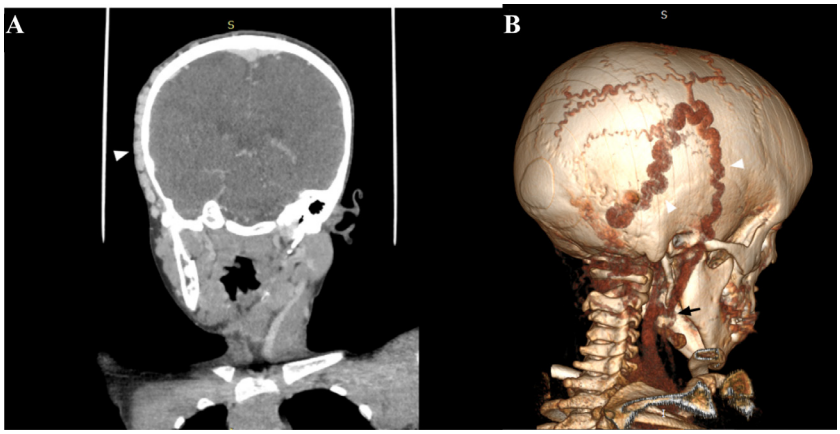

Fig. 2 – Coronal soft tissue window contrast enhanced head and neck CT (A) and angiographic 3D volume rendered images (B) - tortuous, homogeneously, and avidly enhancing right scalp vascular structures (white arrowhead). Black arrow in B shows communication with a right internal jugular vein.

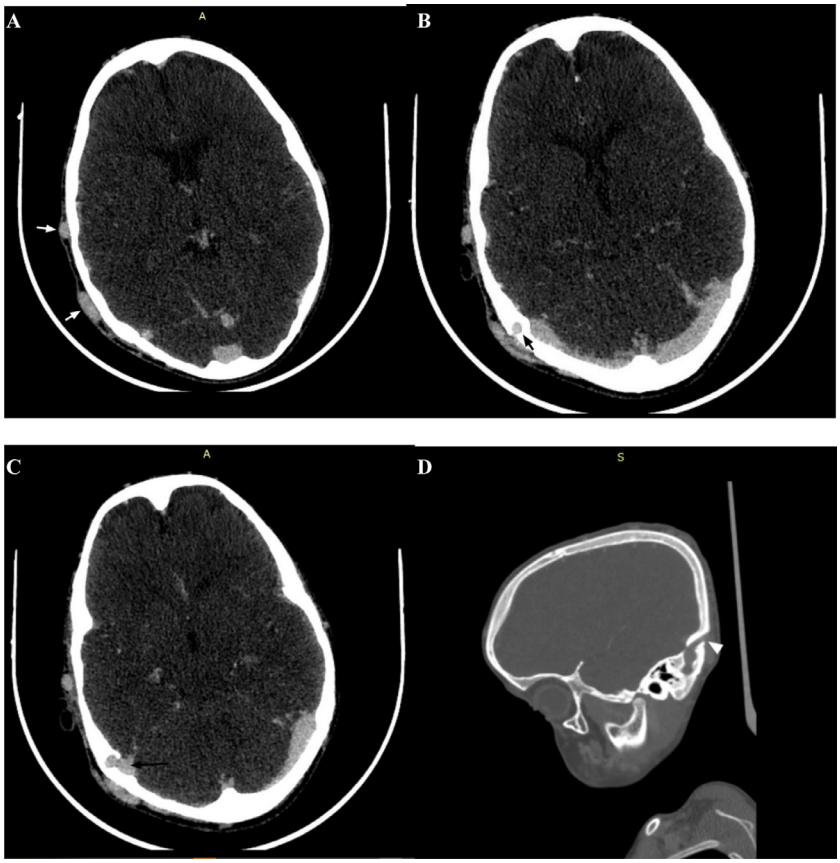

Fig. 3 – Axial angiographic (A–C) and sagittal bone window (D) postcontrast head and neck CT- Axial images (superior to inferior sections) show markedly dilated and tortuous right parito-temporo-occipital scalp vascular structures (white arrow) which communicate with the distal right transverse sinus through a prominent emissary vein within the right temporal bone (black arrows). No arterial feeder, nidus, or enhancing soft tissue lesion. No thrombosis of the dural venous sinuses or the dilated scalp venous channels. Sagittal bone windowed image better demonstrates the right temporal bone defect with sclerotic margin (white arrowhead).

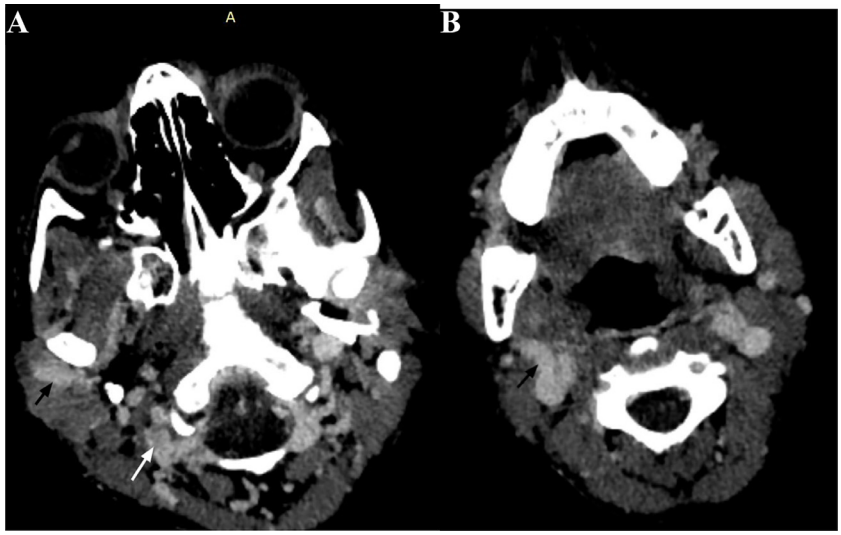

Fig. 4 – Axial (A, B) angiographic window postcontrast head and neck CT images (superior to inferior sections) - multiple prominent right posterior paravertebral vessels, (white arrow on a) some of them communicating with the prominent scalp vein (not visualized in these images). A large communicating scalp vein is also followed to drain directly to the right Internal jugular vein (black arrow on A and B).

nus (40%), parietal (34%), and occipital (23%) bones. Temporal bone (4%) is the least common distribution [7]. Nonmidline distribution is also referred to as lateral and has been reported as a highly unusual location [11–14]. Associated anomalous head and neck venous extensions can also occur. Our case is peculiar as no major superior sagittal sinus involvement was seen, and the main communications extend to the right temporal-occipital region (with the right distal sigmoid sinus) and the right internal jugular vein (Figs. 2, 3 and 4). Extensive paravertebral varicoid veins were also seen. To the best of our knowledge, we did not find any prior report of lateral SP with direct IJV communication and intracranial venous sinus involvement.

Imaging is recommended to differentiate the condition from other similar-looking mimickers like cavernomas, epidermoid or dermoid cysts, menigocele, etc. Radiography can show soft tissue swelling but is generally a nonspecific modality. Sonographic examination is a safe, noninvasive, and easily available modality that is especially advantageous for infants and adds to the diagnosis by elegantly showing the venous nature of the swelling and flow direction on color mode and the calvarial defect on gray scale imaging [15–17]. CT is better at showing the extent and location of the communicating vessels, involved scalp and intracranial venous structures, calvarial defect (bone window preferred), and associated intracranial venous anomalies like a developmental venous anomaly. MRI does well in demonstrating the large varicoid scalp veins and the differentiation of other anomalies, including complex noncommunicating subepicardial veinous malformations [18]. Unless flow is significant through the emissary veins, the communicating veins might not be seen clearly. In addition, MRI is not sufficient to show bony details. Digital subtraction angiography (DSA) is superior to both CT and MRI in the detection of the size, course, location, and flow characteristics of the lesion, all of which are crucial to surgical planning [9]. More importantly, it will show congenital normal anatomic variants related to the sagittal, transverse, or sigmoid sinuses or the dominance of the SP. There are 2 types of SPs, a dominant SP where it drains a major part of the brain, or an accessory SP, whose drainage is insignificant. This information is paramount to establish before resection, as the dominant type of SP will need to be preserved [9,10].

The natural course of SP is variable but generally tends to increase in size progressively. In some cases, though, it can stay stable or even regress. Treatment for SP is surgical, either by open or endovascular means, with the latter associated with fewer peri-procedural complications and a shorter hospital stay. But options are mainly dictated by patients or carers' wishes, concomitant neurological symptoms, or comorbid conditions. In the latter 2, surgical treatment is indicated. In our center, asymptomatic children are put on a periodic follow-up to observe the course of the condition before surgical treatment is undertaken. This is a justified approach, as the exact natural history of this rare condition is not fully known [9].

In conclusion, Sinus pericranii is a rare vascular anomaly of the head and neck region that requires cross-sectional imaging for confirmation and preoperative planning. The lateral location and associated major IJV involvement are highly unusual for the condition.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

Written informed consent was obtained from the patient's parents to publish this case report. Personal identifiers are not used in this paper.

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