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Dr. Reber Saeed Yousif
Neurosurgeon Specialist, M.B. Ch. B, F.I.C.M., M.D., Department of Neurosurgery Azadi Teaching Hospital, Duhok, Iraq

Dr. Salman Ahmed Kheder
Specialist Neurosurgery, Ibn Sina Teaching Hospital, Mosul, Iraq

Karam Fawaz Aldarzi
Specialist Neurosurgeon, Ibn Sina Teaching Hospital, Nineveh Health Directorate, Ministry of Health, Nineveh, Iraq

Corresponding Author:
Dr. Reber Saeed Yousif
Neurosurgeon Specialist, M.B. Ch. B, F.I.C.M., M.D., Department of Neurosurgery Azadi Teaching Hospital, Duhok, Iraq

Rare cranial vascular anomaly, sinus pericranii, three cases report

Dr. Reber Saeed Yousif, Dr. Salman Ahmed Kheder and Karam Fawaz Aldarzi

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Abstract
Background: Sinus pericranii (SP) is a rare vascular (venous) anomaly that connects the intracranial dural sinuses to the extracranial venous channels via emissary trans-osseous veins. It is a collection of non-muscular venous blood vessels that adhere tightly to the cranium and directly communicate with an intracranial venous sinus through diploic veins. About 200 cases have been reported worldwide, however numerous writers have tried to classify and treat this potentially fatal illness. Three unique cases of cranial abnormalities arrived. Different clinical presentations and initial patient care affected prognosis.
Methods: A retrospective study of three cases of congenital vascular SP was recorded in our department.
Results: All three cases were young, male, and had tumor-like masses in the left parieto-occipital venous abnormality improperly linking the intracranial dural sinuses with the epicranial veins. The tumours were entirely eliminated by surgery. By histopathology, vascular endothelium in the pathologic material supports congenital or spontaneous origin.
Conclusion: Paediatric sinus pericranii is uncommon and may be congenital or acquired. Incomplete sutural fusion or in-utero dural sinus thrombosis can induce congenital malformations. Usually involves the SSS and possibly TS, connected by diploic veins. All of our patients were congenital transverse sinus. Plain X-rays, CT, MR, and DSA can diagnose SP. Bony flaws, cortical thinning, and isolated erosions are shown on plain skull radiographs. Clinical examination and brain CT angiography with or without contrast are the most reliable diagnosis methods, and surgery is the best therapy. Resecting the extracranial venous package and ligating the emissary communication is crucial.

Keywords: Sinus pericranii, vascular anomaly, CT angiography, operation

Introduction

Case report 1

History and presentation

A three-year-old boy was brought to medical attention due to a swelling on the back of his head, present since birth and gradually increasing in size. This swelling was notably observed by his parents. Upon examination, the swelling was found in the left parieto-occipital region, resembling a tumor. It measured 15 cm in diameter when the child was lying down. The swelling was soft, not painful to touch, and had a fluid-like quality. It was partially reducible when the child sat up. Additionally, there were detectable bony defects under the swelling, and a cough impulse was present, indicating a change in size with different head positions. Other systemic examinations were normal, with no associated anomalies. CT angiography revealed an extracranial vein passing through a skull defect and connecting to the upper part of the transverse sinus. As show in fig 1, 2.

During surgery, the decision was made to operate on the mass, which was found beneath the cranial vault's periosteum, adjacent to the lambdoid suture, and draining into the transverse sinus. The mass appeared reddish and was connected to multiple scalp veins. The surgeons dissected the area between the soft tissues and veins, maintaining hemostasis with bipolar coagulation. The mass was then carefully lifted from the skull. After identifying the extracranial veins penetrating through the bone defect, the proximal part of these veins was ligated. Following this, the distal portion of the tied area was dissected, with bleeding controlled using bipolar coagulation. Finally, the emissary vein, which facilitated communication between the extracranial and intracranial structures, was coagulated using bipolar coagulation.

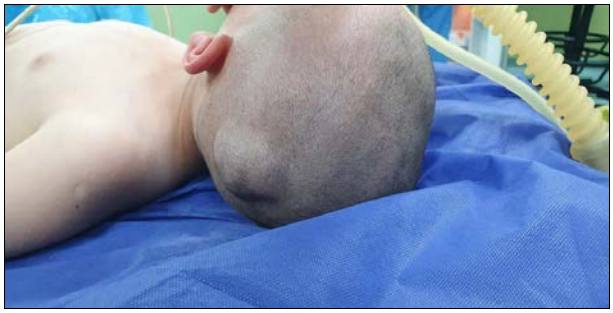


Fig 1: Young child with large, round left occipital mass of (15 cm) diameter

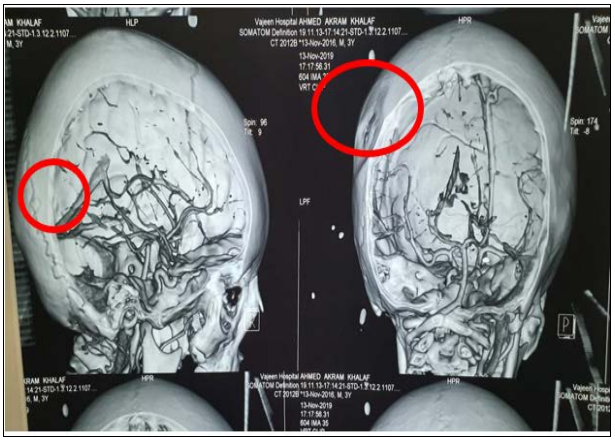


Fig 2: CT angiography of the mass showed the site of connection extracranial with intracranial

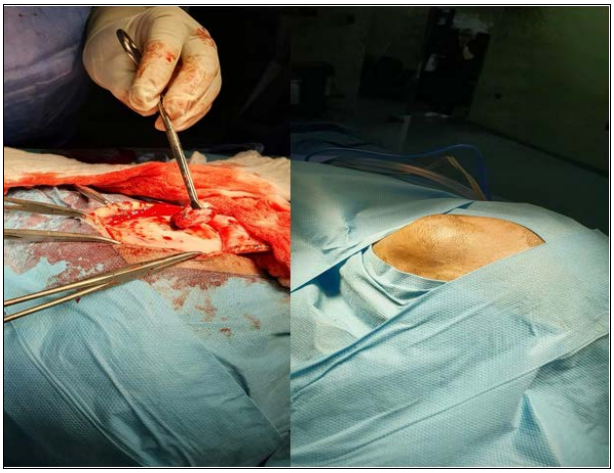


Fig 3: Surgery of mass showed the site of connection extracranial with intracranial

Case report 2

History and presentation

A nine-year-old child came to a clinical consultation exhibiting a soft, non-pulsatile, worm-like, and fluctuant mass. This mass was easily compressible and collapsible, situated along the left lateral side near the Transverse sinus. Notably, the mass vanished when the child sat up but reappeared upon performing the Valsalva maneuver. The lesion was prominently visible when the patient was lying down. CT angiography revealed the presence of emissary vessels traversing the skull bone, indicating a connection between them. As shown in fig 4, 5.

Operation

The child was thoroughly prepared for surgery, including necessary laboratory tests and arranging fresh blood for transfusion. In the operating theater, the surgical team pinpointed the site of the inter-connection between the emissary vessels and the skull. The surgical objectives included identifying the lesion's location, disconnecting the emissary vessels using bipolar coagulation, and cleaning and debriding the soft tissue wound. After the operation, the patient was moved to the Intensive Care Unit (ICU), where he showed daily improvements in consciousness. He was later transferred to the inpatient ward for further observation. During his hospital stay, there were no signs of meningitis or infection at the surgical site. The patient was treated with broad-spectrum antibiotics and discharged home after three days. Follow-up visits over the next six months showed that he was clinically stable, with no signs of the lesion recurring. As shown in fig 6, 7.

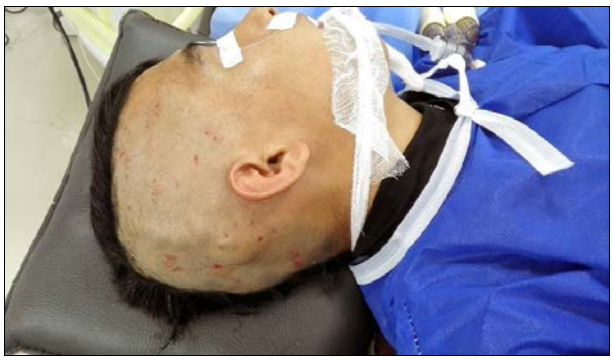


Fig 4: Young child with elongated worm, tortuous right occipital mass

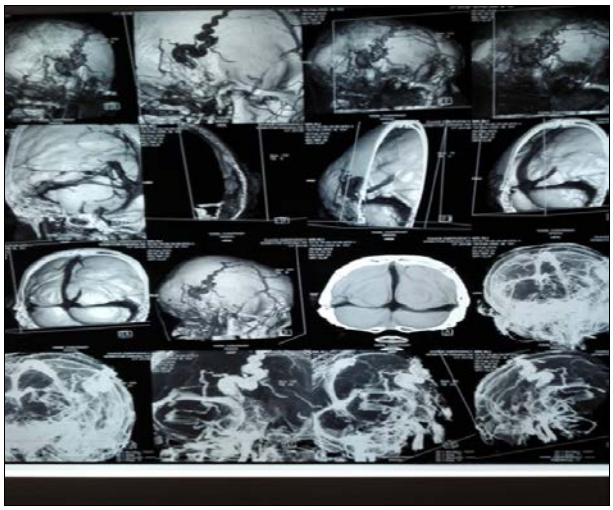


Fig 5: CT angiography digital subtraction angiography (DSA).

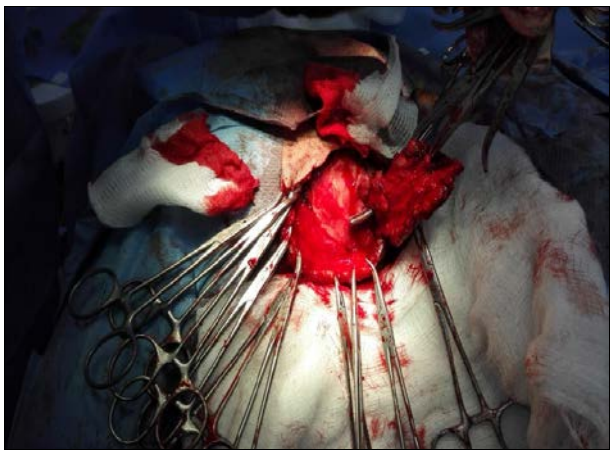


Fig 6: Intra-operative (Black – arrow) site of the connection

Fig 7: (Black circle) skull opening the connection site

Case report 3

History and presentation

A seven-year-old boy was brought in by his parents due to pain in his scalp experienced during hair combing. Upon examination, he reported pain specifically in the right temporal region. Palpation revealed a round, soft, tumor-like lesion in the left lateral area along the transverse sinus. This mass was soft, fluctuant, non-pulsatile, easily compressible, and collapsible. Interestingly, the mass disappeared when the child sat up, but reappeared upon bilateral compression of the jugular veins or performing the Valsalva maneuver. The lesion was most noticeable when the patient was lying down. CT angiography confirmed the presence of emissary vessels crossing the skull bone, indicating a connection. As shown in fig 8, 9.

Operation

Prior to surgery, the child was properly prepared. During the operation, surgeons dissected the area between the soft tissues and the veins, ensuring hemostasis with bipolar coagulation. After the surgery, the patient was initially moved to the Intensive Care Unit (ICU) and later transferred to the inpatient ward for further observation. Throughout his stay in the hospital, there were no indications of meningitis or infection at the surgical site. He was treated with broad-spectrum antibiotics and discharged home after four days. As shown in fig 10.

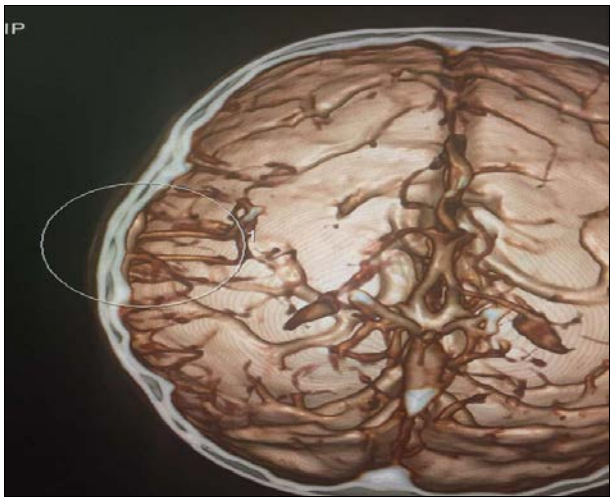


Fig 8: CT angiography digital subtraction angiography (DSA). show the connection transverse sinus

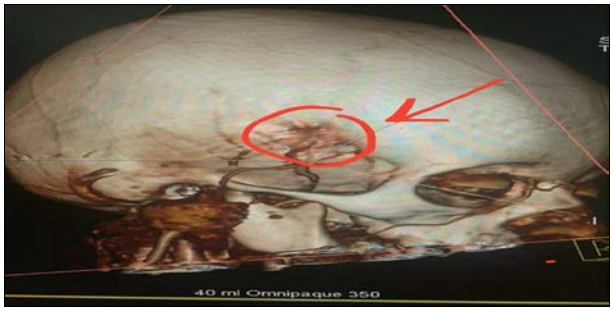
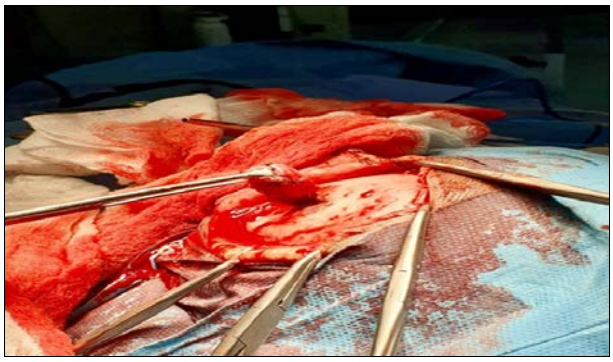


Fig 9: CT angiography show the connection [site of entry] intra cranial and extra cranial vessels.



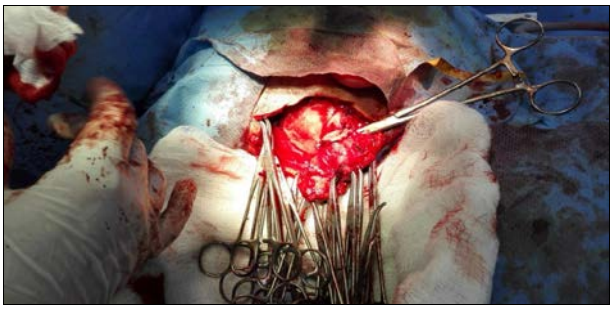


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Fig 10: Intra-operative (Black – circle) site of the connection

Discussiona

Sinus pericranii (SP) is a rare vascular anomaly that connects the intracranial dural sinuses with the epicranial veins through the cranial diploe and emissary veins [1]. This condition was first described by Hecker in 1845 as a specific type of venous malformation [2]. The largest published case series, which includes 21 cases, has shed light on the classification and management of SP [3]. SP is mostly asymptomatic and is often addressed for aesthetic concerns, with approximately 80% of cases presenting as a non-symptomatic, palpable mass [2]. Headache is a common symptom in symptomatic cases of SP, which typically involves the Superior Sagittal Sinus (SSS) and occasionally the transverse sinus, communicating through diploic veins [4, 5]. SP usually manifests in childhood, tends to grow over time, and can be congenital or acquired. Congenital lesions might result from incomplete sutural fusion or in-utero dural sinus thrombosis. Acquired lesions are categorized as either spontaneous or traumatic, the latter often due to tearing of emissary veins. The presence of endothelial lining suggests a congenital or spontaneous origin, while connective tissue indicates a traumatic origin [6-9]. Poppel *et al*. [8] consider SP as a subtype of cavernous hemangioma involving the pericranium. Changes in the skull associated with SP might be due to vascular pressure [10]. Diagnostic investigations include plain X-rays, CT, MRI, and digital subtraction angiography (DSA) [11, 12]. For surgical treatment, the main objective is to resect the

extracranial venous package and ligate the emissary communicating vein. Materials such as bone wax or gelfoam are used to obliterate the communication [13, 14].

Table 1: Data from various Case Reports

Cases	Age	Sex	Site	Shape	Sinus connection	Opereation	Complication
Case one	3 Years	Male	Left occipital	Round	Left transverse sinus	Done	Non
Case two	9 Years	Male	Right occipital	Elongated Worm -shape	Right transverse sinus	Done	Non
Case three	8 Years	Male	Right temporal	Small cystic	Right transverse sinus	Done	Non

Conclusion

In these three pediatric cases, all male and congenital in nature, each involved a connection to the transverse sinus through diploic veins. The diagnosis of Sinus pericranii (SP) was primarily based on distinctive clinical features. Given the potential for life-threatening complications and cosmetic concerns associated with SP, comprehensive diagnostic tests were conducted, including plain X-rays, CT scans, MRI, and digital subtraction angiography (DSA). These cases necessitated early surgical or endovascular intervention. The report highlights a successful surgical treatment of a patient with spontaneous SP, where the mass was situated beneath the periosteum of the cranial vault, near the lambdoid suture alongside the transverse sinus. Interestingly, despite radiological abnormalities, the involved bone was found to be histologically normal, indicating that the lesion primarily affected the dura and the space between the pericranium and the outer table of the skull. During their hospital stay, there were no instances of meningitis or infection. The patients were administered broad-spectrum antibiotics and prophylactic antiepileptic drugs before being discharged home.

Conflict of Interest

Not available

Financial Support

Not available

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