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CASE REPORT Open Access

A rare case report of dominant paediatric sinus pericranii: food for thought!

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Abstract

Background: Sinus pericranii (SP) is a rare venous anomaly abnormally connecting the intracranial dural sinuses with the epicranial venous channels. Various authors have attempted to classify this entity to decide management for this potentially life-threatening condition. With approximately 200 cases reported in world literature till date, no definitive guidelines for management have emerged.

Case presentation: A female child aged 7 years presented to us with complaints of occipital swelling since birth. This swelling was gradually increasing in size as reported by the parents. On examination, the swelling was located in the right parieto-occipital region, size 10 cm diameter in lying position, soft, non-tender, fluid-like diffuse swelling with underlying areas of palpable bony defects, partially reducible in sitting up position, and cough impulse was present. On evaluation, a case of dominant SP was established.

Conclusion: Traditionally, SP has been reported mostly in paediatric population and managed surgically or endovascularly for selected cases, while the remaining cases have been observed. Profuse haemorrhage has been a major deterrent to manage these lesions surgically. Long-term complications like trauma, haemorrhage, infection and thrombosis have been reported in conservatively managed cases. The neurosurgical community has thus been left searching for the optimal management of SP. The authors report this case attempting to classify this case according to the available classification systems and hence decide the best possible management.

Keywords: Paediatric, Dominant, Sinus pericranii

Background

Sinus pericranii (SP) is a rare venous anomaly abnormally connecting the intracranial dural sinuses with the epicranial veins. SP was first described by Hecker in 1845 as a "varix spurious circumscriptus venae diploicae frontalis" [1]. This rare clinical entity has been reported scarcely in the literature, and approximately 200 cases were reported till June 2014 [1]. Largest case series which has been published was of 21 cases which threw light upon the classification and management of SP [2]. However, no clear guidelines have been established regarding the management of such cases. The authors report here a case of dominant SP [2] in a child, thus highlighting the difficulties in management of such cases.

Case presentation

A female child aged 7 years presented to us with complaints of occipital swelling since birth. The size of the swelling had been gradually increasing over the years as reported by mother. There was no other significant birth or perinatal history. On examination, the swelling was located in the right parieto-occipital region, size 10 cm diameter in lying position, soft, non-tender, fluid-like diffuse swelling with underlying areas of palpable (skull) bony defects, partially reducible in sitting up position (6 cm diameter), and cough impulse was present. (Fig. 1). Rest of the systemic examination was normal, and there were no associated anomalies.

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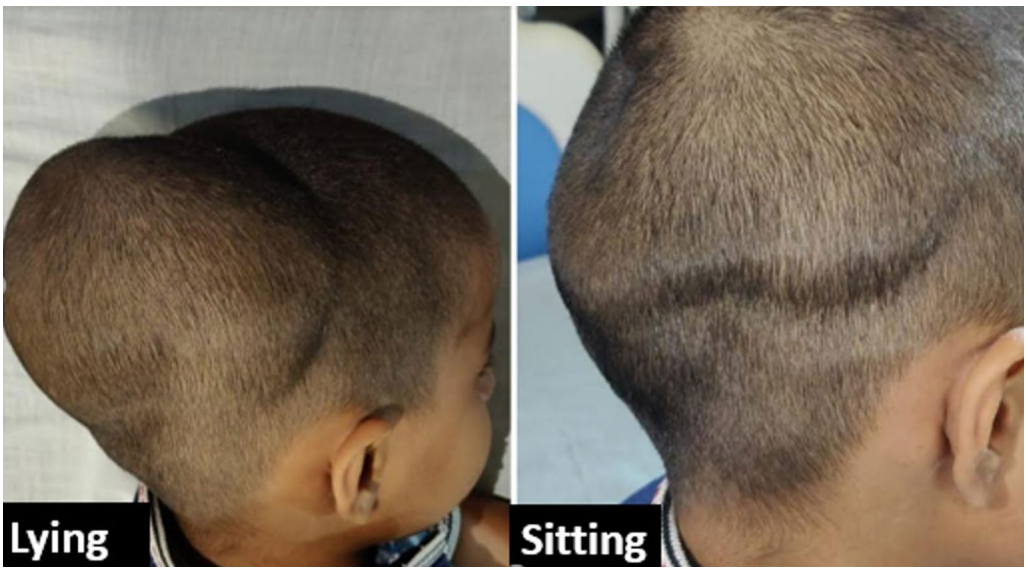

Fig. 1 Swelling reduces partially in sitting up position and reappears on lying down

Evaluation

MRI brain showed large mixed heterogeneously hyperintense lesion in bilateral parieto-occipital scalp. Initial suspicion was of haemangiolymphangioma (Fig. 2). 3D contrast-enhanced computed tomography (CECT) head was shown with multiple foci of nodular irregular enhancement within it and causing marked scalloping and thinning of adjacent calvarium with near complete

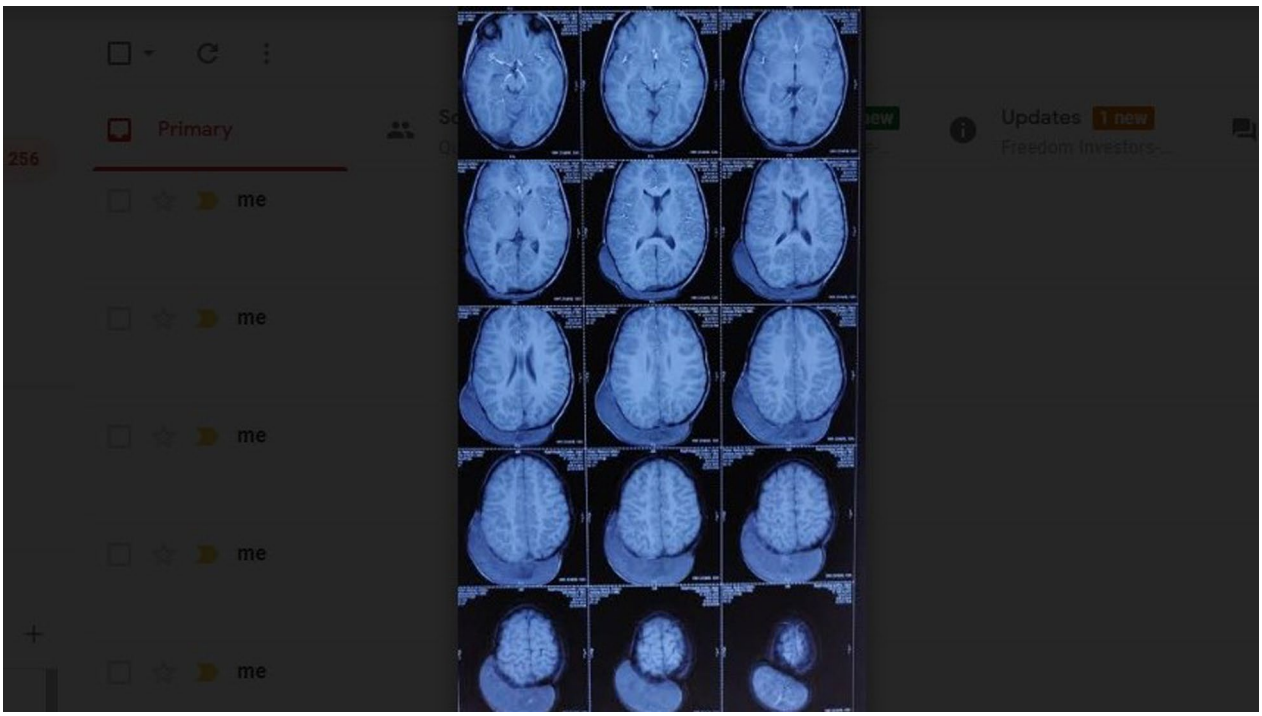

Fig. 2 T1-weighted axial non-contrast images show a large heterogeneously hyperintense swelling over bilateral parieto-occipital region of the scalp suggesting haemangiolymphoma

paper thinning at places; however, no evident intracranial extension was seen (Fig. 3). Computed tomography angiography and venogram was confirmatory of multiple large abnormal communicating veins (>3 mm diameter) (Fig. 4).

Discussion

Stromeyer coined the name "*Sinus pericranii*" in 1995 describing the lesion as "a blood bag on the skull which stands in connection with the veins of the diploe and through these with the sinuses of the brain, whereby an imperfect formation of the outer osseous lamella is easily palpable" [1]. The first significant case series reported significant associations of SP with scaphocephaly, systemic angiomas, multi-sutural synostosis, Crouzon and oxycephaly. These authors evaluated the surgical risk based on anatomical characteristics of the malformation, stating that multiplicity or size (>6 cm) of SP ($p=0.036$) and multiplicity (>3) or size (>3 mm) of transcranial channels ($p=0.004$) was associated with more severe haemorrhage grade [3].

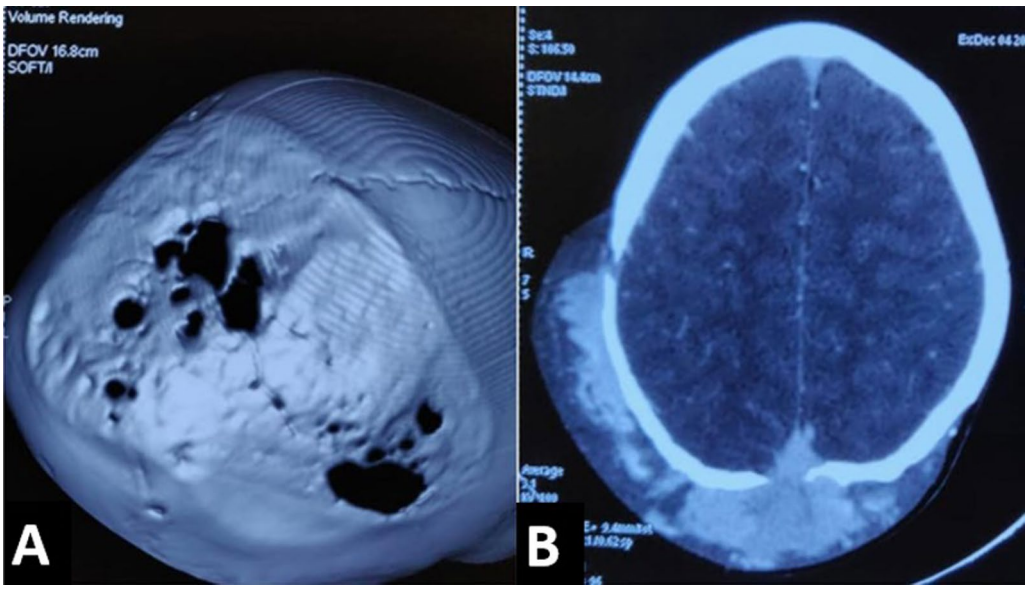

Fig. 3 **A** Multiple bony defects in the parieto-occipital skull bone (>3 mm diameter). **B** Sagittal contrast-enhanced image showing heterogeneously enhancing scalp swelling

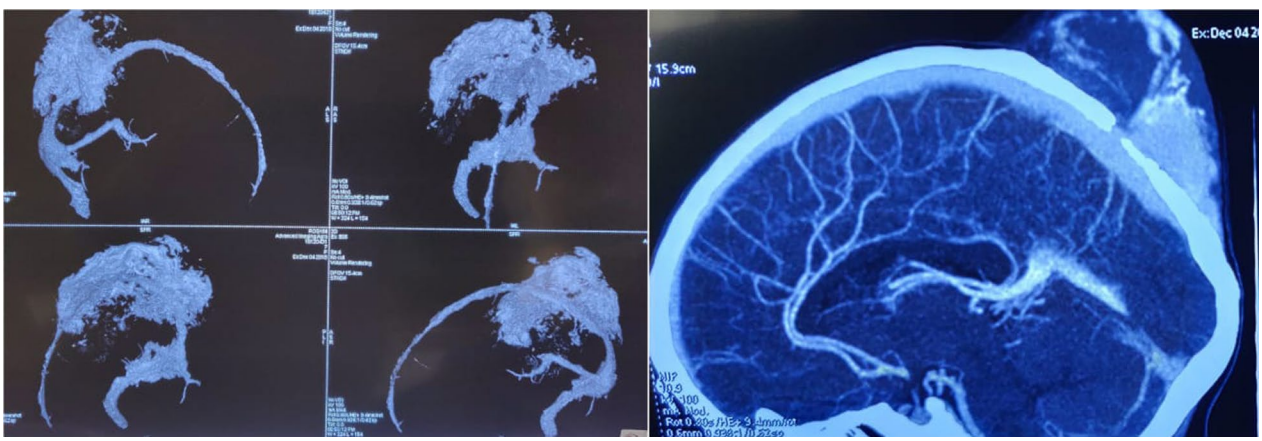

Fig. 4 MR venogram showing multiple large abnormal venous communications between superior sagittal sinus and the scalp swelling confirming dominant SP

The main risk of the surgery was profuse haemorrhage. So, the question now arises whether we need to treat this entity at all? Many case reports have described spontaneous thrombosis in a case of SP which can be fatal [4]. Others have described infection or haemorrhage due to trauma.

In the quest for better understanding and management of this entity, the first attempt at classifying SP was made as late as 2015. A landmark case series of 21 paediatric cases for this condition classifies SP into dominant (draining the majority of the intracranial venous outflow) and accessory (draining only a minority of the intracranial venous outflow) types. The dominant SPs were not treated. Among the patients with accessory SP, 4 were not treated, 2 underwent surgical ligature, and 8 were treated endovascularly. This paper concluded that dominant SP be managed conservatively [2].

Considering the lack of management protocols for this rare condition and after reviewing the literature, the authors decided to manage their case conservatively owing to large size of SP (>10 cm), multiplicity (>3 venous channels) and large diameter of venous channels (>3 mm) [5]. No clinical or radiological evidence of associated syndromes like von Hippel–Lindau syndrome, internal cerebral vein aneurysm or blue rubber syndrome was found in our patient [6]. The patient has been advised 6-monthly follow-up or reporting to us in case of fever, pain in swelling or head trauma.

Follow-up

The patient reported no fresh complaints on 6-monthly follow-up.

Conclusion

This unique case of dominant SP was managed successfully in a conservative manner and can be considered as a stepping stone to help set protocols for the management of such rare cases in the future.

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Author contributions
AKA conceptualised the idea of the case report, was the chief neurosurgeon managing the case and critically reviewed the manuscript. NB was the major contributor in script writing, organisation of data and pictures and corresponding author. Both authors read and approved the final manuscript.

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Availability of data and material
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Declarations

Ethics approval and consent to participate
Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images.

Consent for publication
The parents of the patient included in this case report gave written informed consent to publish the data contained within this study.

Competing interests
The authors declare that they have no competing interests.

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