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# Safety of surgical management of accessory sinus pericranii in infants

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## ABSTRACT

**Purpose**: to better understand the pathological process of sinus pericranii and the safety of the surgical intervention.

**Methods**: patients' archive review of patients who underwent surgical management for sinus pericranii with a confirmed diagnosis and with follow up period greater than 6 months.

**Results**: 6 infants were included; all underwent disconnection surgically with good outcome in all cases.

**Conclusion**: disconnection surgery for sinus pericranii is a safe procedure with a good prognosis.

Keywords
sinus pericranii, scalp swelling, developmental venous anomaly

## INTRODUCTION

Sinus pericranii (SP) is first described in 1850, definition evolved over time with better understanding of the pathology, firstly described by Stromeyer as a bag of blood [1], later it was described as an outpouch of the dural sinus till 1936 when Fevre and Modec outlined the communication between intracranial and extra-cranial venous systems.

Sinus pericranii is a rare extracranial developmental venous anomaly characterized by emissionary vein connecting dural venous sinus to a subgaleal venous varix which may cause skull erosion [2]. Though the exact pathogenesis is not well understood, a number of post- traumatic cases were reported even if the trauma passed unnoticed at the time [1,3]. On the other hand, SP has been associated with congenital conditions like esophygeal atresia, meningocele, craniosinostenosis and other intracranial venous anomalies [4,5,6].

Another theory adopted by Renier and Marche proposed SP is linked to increased intracranial pressure, based upon observing SP in patients with hydrocephalus, macrocephaly and craniosinostenosis [7]. SP is presented solely as "primary" pathology, or "secondary" to an intracranial vascular malformation.

The rarity of the pathology and paucity of observational studies make it challenging to plan for treatment and/or expect the course of the disease, however the natural course of the disease is benign, hemorrhage could be fatal in case of accidental injury.

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In this study, we report our experience with six cases of SP who were managed surgically, all of them were of infantile group age.

## Patients and methods

Patients' archive was reviewed to include patients who were admitted to neurosurgery ward for scalp lump during the years 2018 to 2020 and whose follow up continued for at least 6 months. Patients with other extracranial vascular malformations other than SP e.g scalp AVM were excluded as well as patients with non-vascular scalp lesions. Only patients younger than 2 years were included.

Medical records were searched for clinical presentation, radiology, associated pathology and management. Follow up records in outpatient clinic were obtained for the mentioned period.

## Results

Study included 6 infants ,4 of them were males. Age ranged from 3 to 8 months, with average 4.3 months. Four families (67 %) gave history of head trauma, patients were followed up for at least 6 months with average 11.8 months follow up period (Table 1).

| Case | Age (months) | Sex | Head Trauma | Radiology | Surgical maneuver | Follow up (months) |
|---|---|---|---|---|---|---|
| 1 | 3 | m | yes | CT, MRI, MRV | Disconnection, mass excision | 6 |
| 2 | 4 | m | yes | MRI, MRV | Disconnection | 17 |
| 3 | 8 | f | yes | MRI, MRV | Disconnection, mass excision | 15 |
| 4 | 5 | f | no | MRI, MRV | Disconnection, mass excision | 13 |
| 5 | 3 | m | no | CT, MRI, MRV | Disconnection | 11 |
| 6 | 3 | m | yes | CT, MRI, MRV | Disconnection | 9 |

**Table 1.**

Five lesions were midline and one lesion was parasagittal, while all were related to Superior Sagittal Sinus with various patterns of venous drainage but no cases showed significant parenchymal drainage into the varix. Pathological examination showed endothelial lining of the varix in five cases, and one case's pathology report was irrelevant. One case was associated with esophageal atresia, otherwise neither associated pathologies nor intracranial vascular anomalies were reported.

**Surgical technique:** all surgeries were performed under general anesthesia, scalp incision directly over the lesion with surgical target to identify the emerging vein to disconnect and excise the subgaleal mass when applicable.

**Outcome**: All patients were discharged fully conscious with no neurological deficit either on discharge or during follow up visits. One patient showed superficial wound infection in the first follow up visit (one-week post-operative), which responded to conservative management.

## Discussion

SP is presented early in life either with mild symptoms e.g pain and tenderness or severe symptoms related to hemodynamic disturbance e.g bradycardi[8], however the most common presentation is non-pulsatile soft scalp swelling (Fig 1) which becomes tense on crying and straining, scalp swelling causing cosmetic disfigurement was the main complaint for all the cases in this study, such cases should be clinically and radiologically differentiated from other scalp lumps before proceeding to treatment [8]. Parents seek medical advice with or without history of head trauma which could be missed as well, majority (67%) of patients in our study gave positive history of trauma.

Though some cases are strongly linked to head trauma, the frequent association with other cranial vascular anomalies especially developmental venous anomalies supports the congenital theory of the origin of SP such as failure of regression of interperiosteodural venous plexus or non-closure of skull sutures [5,6].

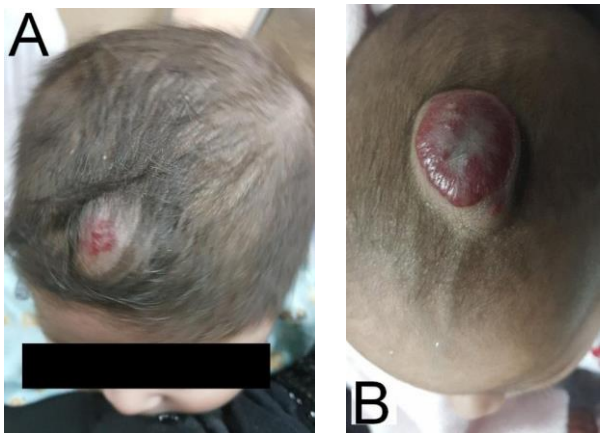


**Figure 1.** Main presentation of SP as a scalp swelling, usually

devoting hair on top when increases in size. Related to coronal suture (A), behind the coronal suture (B). Both infants with open fontanile.

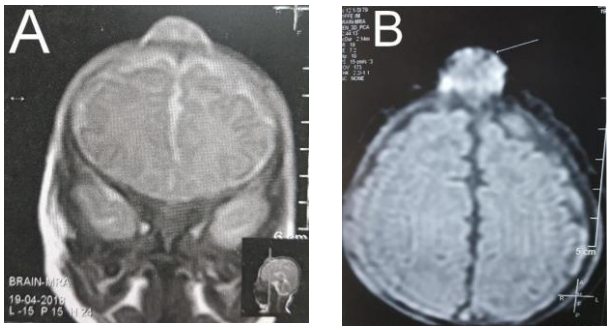


**Figure 2.** MRI T2 coronal (A), and axial T1 of two different cases showing midline SP related to SSS.

CT scan is required to identify skull erosion and enlarged diploic vein. MRI (Fig 2) is needed to exclude parenchymal lesions and AVM's [9], no cases of co-existing intracranial pathology were encountered in this study, while MRV we found beneficial to classify SP via a non-invasive tool.

The necessity of digital subtraction angiography (DSA), though concluded by Gandolfo is not indicated in all cases according to our experience. In our study, MRV was sufficient to classify SP into primary and accessory, no dominant cases were encountered in this study. MRV was not among the radiology options for Gandolf series [10]. We suggest that DSA could be reserved to cases with associated intracranial vascular anomaly or with suspected significant venous drainage into the varix.

Pathologically SP is categorized into two main groups, dominant where it represents a drainage to the cortex and accessory where part of the cortex or the dural sinus drains directly and solely into the varix 10. Another classification suggested by Brook et al 11 to categorize SP according to the feasibility of endovascular treatment, A, B, C. Where A stands for the dominant variant, B for the accessory variant and C variant in which no parenchymal venous drainage into the varix. Type C is the safest for endovascular embolization and type A is an absolute contraindication for.

Histological classification by Nakasu[12] listed three different types; a) fibrous architecture with cavernoma like structure, b) venous varix with endothelial lining and c) herniated dural venous sinus.

Though spontaneous regression of SP through thrombosis had been reported, cases underwent surgery in this series were large and parents decided to undergo surgery [4,13]. Surgical management includes disconnection of the emissary vein (Fig 3) with excision of the mass when applicable, which was performed for all our cases. For three patients in this study (50 %), disconnection of the vein was enough to collapse the mass completely with no actual mass to be excised, for these cases only biopsy of the potential cavity was performed, one of which showed irrelevant sample. Some authors reported closure of bone defect with bone wax or cranioplasty for larger defects which was not necessary in this series. Serious complication such dural sinus laceration and hemorrhage were reported [14], though not encountered in this study, we believe this complication is liable to happen with Nakasu type C which could be excluded preoperatively and intra-operatively with microsurgical microscope.

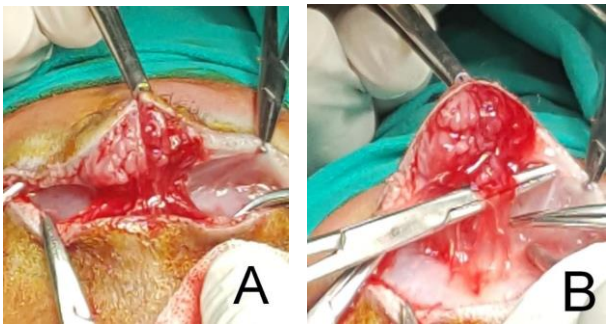


**Figure 3.** Surgical management, subcutaneous dissection to identify the connection vein thoroughly (A), identification of the connection vein, skeletonization and cauterization (B).

Endovascular therapy is a valid means for treatment of SP, it can be performed either via direct puncture, transvenous or combined. Endovascular therapy carries the risk of skin sloughing and thromboembolic events, also it is questionable to consider it less invasive than surgery in infantile age group included in this study. In a case report by Brook et al [11], nBCA was used to embolize the varix however cosmetic outcome is questionable as it initiates an inflammatory reaction which might lead to varix hardening. Another case report by Kessler et al[15] reported good cosmetic result using direct absolute alcohol injection after transvenous endovascular closure of the in/outflow point. The concerns about absolute alcohol leakage into circulation are serious, connection site to systemic

circulation should be confirmed to be sealed before proceeding to injection. Also, theoretical assumption that embolization might lead to development of new vascular anomalies in the region surrounding the occluded one is a reasonable concern.

Although most of cases of Pavanello et al series[8], the largest to our knowledge, were treated via endovascular techniques, it included only 7 infantile cases which were managed conservatively except for one which was treated surgically during the corrective surgery of the associated craniosynostosis. The same study reported spontaneous resolution of a number of accessory cases, an option was not accepted for all the parents in this study due to the disfigurement at time of presentation.

Another large study by Gandoflo et al [10] included 15 infants, six of which required surgical intervention, four of them were dominant variant, with good outcome for all cases including dominant cases which were managed surgically. These results make the necessity of DSA, which is an invasive maneuver, questionable since categorizing SP via DSA didn't change the role of surgery, also significant brain parenchymal drainage is diagnosed with MRV (Fig 4).

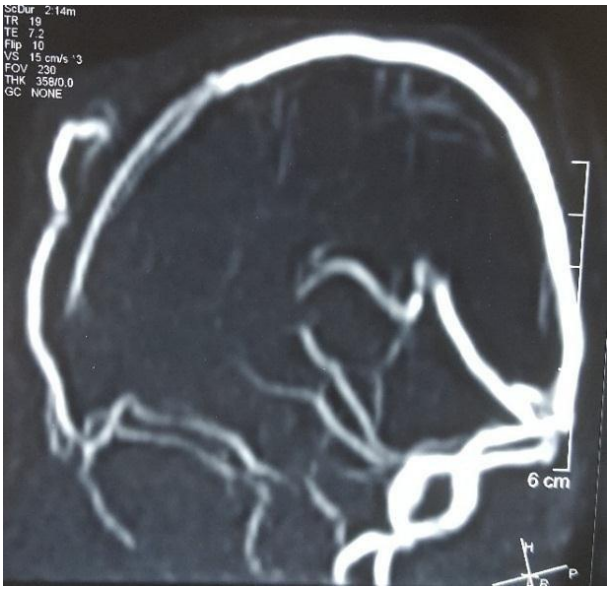


**Figure 4.** MRV showing accessory SP, related to SSS. With no significant brain parenchymal drainage into the varix.

In all means, the extent of treatment depends on the degree of normal brain parenchyma draining into the DVA, as a rule no normal parenchymal brain drainage should be compromised.

## Conclusion

SP is a rare condition in infants characterized by a subgaleal venous varix connected to intracranial venous system via abnormally enlarged emissary vein(s), surgical management of sinus pericranii in infants is safe method for accessory cases with no significant parenchymal drainage into the varix.

## Abbreviations

AVM: Arteriovenous Malformation;
CT: Computed Tomography;
DSA: Digital Subtraction Angiography;
DVA: Developmental Venous Anomaly;
MRI: Magnetic Resonance Imaging;
MRV: Magnetic Resonance Venogram;
nBCA: N-butyl cyanoacrylate;
SP: Sinus Pericranii.

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