CASE REPORT

Sinus pericranii: long-term outcome in a 10-year-old boy with a review of literature

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SUMMARY

Sinus pericranii is a rare cranial venous anomaly in which there is an abnormal communication between intracranial dural sinuses and extracranial venous structures, usually via an emissary transosseous vein. We report the case of a 10-year-old boy who was successfully managed conservatively. The clinical findings, investigations and management are discussed. We focus on the long-term management of these patients, with a review of the relevant literature.

BACKGROUND

Sinus pericranii (SP) was initially described by Hecker in 1845 and the term sinus pericranii was later coined by Stromeyer.¹ Due to its rarity, pathogenesis and natural history remains unclear. This condition is usually identified clinically and can be further diagnosed with imaging such as MRI scanning, which usually shows an abnormal communication between dural sinuses and the cranial vault. The long-term outcomes of SP are poorly understood, and this report aims to add to the current understanding of conservative management.

CASE PRESENTATION

We describe a case of SP in a 10-year-old boy who was noted to have a large frontal scalp swelling at birth. This was managed conservatively and at 5 months of age, he subsequently had an MRI brain scan which showed a vascular malformation involving the soft tissues of the scalp and communicating with the superior sagittal sinus. An ultrasound scan at 10 months old showed that his left frontal parietal vascular malformation persisted, with reduced areas of vascularity and increased areas of haematoma formation. It also showed that in the midline the communication between the superior sagittal sinus and the subgaleal vessels persisted.

Neurological examination was unremarkable and he had normal eye movements with no squint and had a normal funduscopy. At 20 months, he was making good progress and there were no concerns from a neurodevelopmental point of view. He still had a visible area of pulsation overlying the vascular malformation. His head circumference remained between the 50th and 75th centile for age. He was then followed up with further ultrasound scans. The possibility of an intervention had been discussed early on with the child's parents. They were well

informed of the risks of trauma sequelae, thrombosis or haemorrhage but given the fact that he was progressing well without neurological compromise, they were reassured and happy with him attending regular follow-up appointments.

Two years ago, he was reviewed in clinic as he was experiencing headaches that happened two to three times a week. These were not associated with nausea and vomiting or visual disturbances. His mother was asked to keep a detailed diary of these episodes to clarify if this was following a specific pattern and whether they were linked to a rise in intracranial pressure (ICP). He subsequently attended an ophthalmology appointment which was normal. The team believed that the headaches were not related to the SP, therefore it was not felt appropriate to carry out further invasive testing to rule out raised ICP. There were also concerns about a poor diet and low body weight. However, neurological development appeared unaffected. He was therefore referred to the neurosurgical service for further management.

During this review recently, examination revealed an irregular bifrontal calvarial thinning in the midline on either side with an obvious pulsation noted, associated with blue discolouration throughout this swelling. The surrounding skull appeared abnormal, with an area of irregularities under the dysplastic skin.

INVESTIGATIONS

He had an annual review in the clinic. An incidental finding of a 31×14 mm arachnoid cyst in the right temporal fossa was identified with further MRI imaging in 2011. The scan also showed a decrease in the size of the subcutaneous component of the vascular lesion on his scalp. It was also noted that he lacked hair over the bony defect in the vortex area, with an appearance almost of cutis aplasia.

Further neuroimaging (CT and MRI brain with MR venogram (MRV)) was organised to assess for any change in the anatomy.

The CT head showed bilateral deficiencies in ossification of the skull vault involving the posterior frontal and anterior parietal bones, with very little ossification in the region of the normal location of the anterior fontanelle (figures 1 and 2). Posterior to this, more marked on the left than the right, there appeared to be only an inner table of the skull, with some scalloping. MRI head showed a discontinuity of the normal fat signal from the scalp to the left

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Figure 1 Three-dimensional CT defect involving the posterior, frontal and anterior parietal bones, with poor ossification in the region of the anterior fontanelle.

of the midline, in the region of the vertex (figure 3). This is less clearly defined than previously but is up to 4 cm in diameter. The MRV (figures 4-6) showed multiple vascular channels in communication with the superior sagittal sinus, more prominent on the right than the left, with no significant changes since 2011.

While angiography can provide unique information about the dynamic pattern of this condition, this was not performed in this case. This was discussed in the vascular multidisciplinary team meeting and as this child was well clinically, the risks of

Figure 2 Three-dimensional CT demonstrating the bilateral deficiencies in ossification of the skull vault.

Figure 3 MRI, sagittal view showing discontinuity of the normal fat signal along the scalp.

angiography were considered to outweigh the benefits. This was discussed with the family who were happy to have MR angiography if needed.

DIFFERENTIAL DIAGNOSIS

- ▶ Atretic cephalocele;
- ▶ Dural fistula or arteriovenous malformation;

Figure 4 MR venogram, transverse view showing prominent vascular channels in communication with the superior sagittal sinus.

Figure 5 MR venogram, coronal view highlighting vascular channels in communication with the superior sagittal sinus.

Figure 6 MR venogram, sagittal view identifies collateral vessels running between the superior and inferior sagittal sinuses.

- ▶ Arteriovenous fistulae;
- ▶ Subepicranial varix (dilated scalp venous sac without communication with intracranial dural sinuses);
- ▶ Dermoid or epidermoid sinus;
- ▶ Sebaceous, epidermoid, dermoid cyst;
- ▶ Cephalohaematoma;
- ▶ Infantile haemangioma;
- ▶ Aplasia cutis congenital;
- ▶ Cutaneous meningioma;
- ▶ Meningoencephalocele and cranial meningocele.

TREATMENT

This patient was successfully managed with conservative measures and regular follow-up appointments. There was no developmental abnormality noted. Radiological follow-up added to the understanding of the nature of SP. There were no apparent changes in the vascular channels on MRV noted since imaging from 2011.

OUTCOME AND FOLLOW-UP

This case confirms that this condition can safely be managed conservatively when followed up regularly. MRI scanning has proven to be an effective mean of safely monitoring SP without the impact of radiation that comes with other imaging modalities such as CT scanning.

DISCUSSION

This condition is mostly identified by a palpable mass which is usually asymptomatic with some reports of headaches or local pain associated with it. The main concern is usually cosmetic due to the bulging nature of the malformation. Although it is typically a congenital finding, it has also been found to be secondary to trauma.² The distribution of the lesions is more commonly found in the frontal region (43%), followed by parietal (36%) and occipital regions (7.4%).³

Confirmation of this anomaly can be identified by various imaging techniques such as CT scan, MRI and Doppler ultrasonography. CT scanning allows direct visualisation of the bony defect. MRI with or without MR angiography is believed to be of great benefit at providing a definitive diagnosis of SP and its drainage into the dural sinuses.⁴ Angiography can separate SP into two patterns; dominant and accessory.

There is little available evidence with regards to available guidelines and recommendations concerning the management of this condition. A recent retrospective study by Pavanello *et al*, looking at the diagnosis and management in 21 paediatric patients, suggests that only accessory SP is amenable to treatment, whereas dominant sinus pericranii must be preserved due to its potential life-threatening complications associated with it, such as haemorrhage and venous congestion.^{5,6}

Endovascular approach to treatment has proven to be safe and effective when treated for cosmetic reasons, in cases of accessory SP. Digital subtraction angiography has proven to be effective in excluding other vascular malformations as well as assessing the intracranial venous dynamics and their relationship to the SP.⁷ While this has been rarely reported in the available literature, endovascular embolisation appears to have good results when patients are identified for potential complications.^{7,8} This method of managing SP is less invasive than surgery but still carries the risk of causing skin necrosis and embolic events.⁹

Surgical management has been demonstrated in a retrospective study conducted by Park *et al* where 10 patients underwent operations for SP.¹⁰ All patients with SP associated with

cranosynostosis were managed conservatively. This study concluded that surgery can be performed in cases without intracranial venous compromise. Surgery can be safely carried out with thorough preoperative planning and imaging of the cranial vault.

Conservative management has rarely been reported in the literature with regards to follow-up. One case reports the case of a 6-year-old who was safely managed through this method.¹¹ Surveillance proved an effected method of monitoring the condition, with no reported complications. Another retrospective study identified four patients with untreated accessory SP, two of which resolved spontaneously, one was lost in follow-up and the remaining one was unchanged.

This case highlights the success in conservative management of this rare condition. Regular follow-up appointments and repeat scans can ensure disease progression is monitored closely and, as shown in this case, long-term follow-up of such patients is a safe method of managing SP. While the available literature mentions management options of this rare condition, no evidence has been found with regards to long-term outcomes. As mentioned in the literature, this condition is more commonly treated for cosmetic reasons. Surgery should be avoided in cases of dominant SP due

to the high risks associated with it. The evidence for the management of this condition remains limited and more research would be beneficial in the options available to successfully manage SP.

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Learning points

- This case highlights and reiterates the benefit of conservative management in sinus pericranii (SP) treatment paradigm.
- Cosmetic reasons remain the main indication for surgery in SP. However, the potential indications for surgery, which include the prevention of sequelae particularly haemorrhage or traumatic changes in patients with skull galloping, should be understood in the management of such a rare condition.
- Surgery should be avoided in cases of dominant SP.

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