Sinus pericranii was first described by Stromeyer in 1850 as a “blood bag on the skull . . . in connection with the veins of the diploe and through these with the sinuses of the brain.”

Most cases of SP become clinically apparent as nonpulsatile, often blue-tinged, soft-tissue masses located in the frontal region, along or close to the midline, which disappear on compression and reduce in size with upright posture. Conversely, SP enlarges with maneuvers that increase intracranial pressure, such as coughing and Valsalva. We present a case of this rare condition successfully treated by transvenous embolization, as well as a review of the literature and discussion of treatment options. To our knowledge, definitive treatment of SP by transvenous embolization has not been previously described.

Case Report

History and Examination. This 3-year-old boy presented with a small, soft, blue-tinged mass at the vertex of his head, just to the right of the midline, with multiple radiating serpiginous scalp veins (Fig. 1). Though the mass and scalp veins were present from birth, they had progressively increased in size and extent. The mass varied in size, increasing with Valsalva maneuver and crying. A bone defect underlying the mass was palpable, as were additional curvilinear grooves in the skull. Findings on physical examination were otherwise unremarkable. The patient had no significant medical history and no siblings.

An unenhanced head CT examination revealed a 1.2-cm soft-tissue attenuation mass in the scalp overlying a small defect in the right parietal bone. A CT angiography study demonstrated enhancement of the mass, which had a direct connection to the SSS via a large emissary vein (Fig. 2). Large superficial subgaleal draining veins, connecting to the mass, were present over the skull. Reconstructed 3D images showed the mass overlying the right parietal foramen and demonstrated curvilinear grooves within the skull, corresponding to the course of the large draining veins (Fig. 3).

Digital subtraction angiography was performed from both arterial and venous access. The venous phase of a right ICA angiogram confirmed communication between
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Fig. 1. Photographs of patient’s skull with patient in Trendelenburg position (A) and upright (B). In panel A, large serpiginous veins may be seen emanating from the region of the right parietal foramen (arrow). When the patient was upright (B), the veins collapsed.

the SSS and a large extracranial vein through a defect in the parietal calvaria, consistent with SP (Fig. 4A and B). Antegrade flow of contrast material toward superficial facial veins was seen. Drainage of major cortical veins, including the vein of Trolard, was into the SSS. A DVA was seen in the right frontal lobe; it did not communicate with the SP. A transvenous microcatheter was advanced into the SP via the SSS. Superselective venography of the SP demonstrated filling of superficial scalp veins with negligible retrograde flow into the SSS (Fig. 4C).

Treatment and Postoperative Course. After careful review of the diagnostic angiogram by the surgical and neurointerventional teams, an endovascular approach was pursued. A regulated pressure dressing was applied to the scalp overlying the SP. The microcatheter tip was positioned in the distal external portion of the osseous channel. Under direct fluoroscopic visualization, N-butyl cyanoacrylate glue mixed 1:1 with Lipiodol was injected through the microcatheter until occlusion of the intraosseous portion of the venous communication was demonstrated (Fig. 5A). The posttreatment angiogram demonstrated no drainage from the SSS to the superficial scalp veins, proving successful obliteration of the SP (Fig. 5B).

The postoperative course was uneventful and the patient experienced no headache or neurological deficit. The serpiginous extracranial veins significantly decreased in size following the procedure. The patient was discharged 2 days after the procedure and 1 year later had had no recurrence.

Fig. 2. Coronal image from CT angiogram demonstrating contrast medium within the SSS (black arrow) in communication with extracranial veins through a venous channel traversing the right parietal foramen (arrowhead).

Fig. 3. Three-dimensional volume-rendered CT reconstruction demonstrating bilateral parietal foramina (A) and after superimposition of the soft tissues (B), including overlying veins, the SP traversing the right parietal foramen.

Discussion

Sinus pericranii is a rare, usually asymptomatic condition that is characterized by an abnormal communication between the intra- and extracranial venous drainage pathways. The etiology of SP is unknown. Development of SP after head trauma suggests an acquired pathophysiology, but its frequent association with other intracranial vascular anomalies, most commonly DVA, supports a congenital cause such as failure of closure of the cranial sutures or failure of regression of the interperiosteodural venous plexus during early brain development. The presence of an endothelial lining histologically defines a congenital SP and distinguishes between the congenital and acquired types.

Typically, SP presents in younger patients; in one report the mean age was 20 years. The condition occurs with equal incidence in male and female patients, although males are more commonly affected in posttraumatic cases. In most patients SP occurs along the midline, with location reported as 40% frontal, 34% parietal, 23% occipital, and 4% temporal. Off-midline locations are exceptional. There are a number of reported associations with SP, including venous malformations, blue-rubber bleb nevus syndrome, aneurysmal internal cerebral vein, and vascular lesions of the cerebellum or retina in the setting of von Hippel–Lindau syndrome.

Clinical symptoms of SP are often mild, including headache, sensations of pressure or fullness, vertigo, or localized pain. The prognosis is almost always favorable, and the natural history commonly shows no increase in size after puberty. Spontaneous partial thrombosis has also been described.

The differential diagnosis for SP is broad and includes cavernous hemangioma, arteriovenous malformation, subepicranial varix or hyroma, epidermoid tumor, encephalocele, and leptomeningeal cyst. The clinical features of SP, as demonstrated by manual and hemodynamic maneuvers on physical examination, together with the characteristic imaging findings, are strongly suggestive of the diagnosis. Unenhanced CT may demonstrate a soft tissue lesion and associated bone defect. Magnetic resonance imaging reveals a characteristic soft-tissue mass of mixed
signal intensity, with areas of signal void corresponding to blood flow. Computed tomography angiography and contrast-enhanced MR imaging may demonstrate drainage of the SP into the dural venous sinuses, usually the SSS. Computed tomography angiography and MR angiography/venography may also demonstrate any enlarged superficial cortical veins, associated vascular anomalies, and the caliber of normal anatomic venous drainage pathways.  

Treatment of SP has mainly been recommended for reasons of cosmesis, prevention of hemorrhage, and the risks of air embolism.  

Surgery is the established method of treatment, with the first recognized successful surgery for SP having been reported in 1902. Cranietomy of the involved skull with division of the diploic veins followed by cranioplasty has been recommended. Several studies have reported significant hemorrhage during surgery as a result of dural sinus laceration. Recently, disconnection of emissary veins alone has been thought to be adequate to prevent recurrence, with bone wax being used for closure of the cranial abnormalities.

Although noninvasive imaging techniques can provide a definitive diagnosis of SP, cerebral angiography and venography should be performed if treatment is being considered. Digital subtraction angiography best excludes other vascular malformations mimicking SP or direct drainage of a DVA into the SP, both of which would significantly alter the treatment plan. In addition, DSA best assesses the intracranial venous dynamics and their relationship to the SP, information that is crucial to treatment planning. Gandolfo et al. recently classified SP based on flow dynamics identified at angiography. Two basic patterns of SP were identified: 1) “dominant,” in which the main stream of contrast flow uses the SP to drain the brain parenchyma, bypassing the usual venous outlets, and 2) “accessory,” in which only a portion of the brain’s venous outflow occurs through the extradiploic vessels. A dominant flow pattern is considered a contraindication to both surgical and endovascular treatment, in view of the potentially life-threatening complications, including bleeding, venous congestion and/or infarction, and hemorrhage.

The feasibility of treatment of SP with an accessory flow pattern is dependent on the degree of brain parenchymal venous drainage into the dural sinuses, as demonstrated on angiography (Fig. 6). At no time should normal brain parenchymal venous drainage be compromised. Additionally, drainage of a DVA into the SP is a contraindication to treatment. Downstream stenoses in the dural sinuses and jugular bulbs should be considered relative contraindications to treatment.

In our case, cortical veins in the region of the SP drained into the sagittal sinus and there was no evidence of stenosis in the sagittal sinus distally. Although our patient did have a DVA, angiography clearly demonstrated independent drainage of the DVA into the SSS, without connection to the SP. Therefore, we considered treatment by an endovascular approach safe and potentially effective.

Conclusions

Sinus pericranii is a rare, abnormal communication between the intracranial and extracranial venous drainage pathways. The pattern of brain drainage in the region of the SP, as demonstrated by angiography, is crucial to assessing treatment options and planning intervention. Although surgery is the more established method of treatment, a transvenous endovascular approach can be safely used for definitive embolization. The endovascular approach provides for visualization of both intracranial and extracranial venous connections to the SP, allowing for possible greater intraprocedural control. As with surgery, the goal of endovascular intervention should be to permanently occlude the intracranial connection of the SP to the dural sinus.

Fig. 4. Lateral (A) and oblique (B) views from the venous phase of a right ICA injection demonstrating communication between the SSS and a large superficial vein (white arrow) through a channel in the parietal calvaria (black arrow). Note DVA (arrowhead) draining into the SSS, without connection to the SP. C: Lateral view during selective injection into the calvarial channel demonstrating antegrade flow toward the superficial veins.

Fig. 5. Posttreatment images. A: Unsubtracted lateral view demonstrates N-butyl cyanoacrylate glue cast traversing the osseous channel, extending into the proximal aspect of an extracranial draining vein. B: Lateral view from the venous phase of a right ICA injection demonstrates normal flow within the SSS and no opacification of extracranial veins, proving successful obliteration of the SP.
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Fig. 6. Artist's rendering of SP drainage patterns. Venous drainage of the brain is depicted as a DVA for purposes of illustration. A: “Dominant” pattern: Brain drainage is directly into the extracranial SP without accessory drainage into the dural sinus; embolization is contraindicated with this pattern of drainage, as it may lead to venous infarct and/or hemorrhage. B: “Accessory” pattern: Brain drainage is into both the SP and the dural sinus; the safety of SP treatment depends on the degree of brain drainage into the dural sinus relative to the SP. C: Normal brain drainage into dural sinuses with a direct connection demonstrated between the dural sinus and extracranial veins through a transosseous channel; treatment of SP is considered to be safe.

Disclaimer
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