Endovascular embolization with Onyx in the management of sinus pericranii: a case report

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Sinus pericranii (SP) is an uncommon and usually asymptomatic communication between intra- and extracranial venous drainage pathways in which blood flow can circulate bidirectionally through abnormal dilated veins through a skull defect. Diagnosis and evaluation of the venous drainage pattern is important if treatment is contemplated. Cerebral angiography with the use of Dyna CT can be helpful in the diagnosis of SP and its relationship with the skull defect. The authors report what is, to the best of their knowledge, the first case of SP treated by means of endovascular embolization with Onyx. (DOI: 10.3171/2009.8.FOCUS09170)

Key Words • sinus pericranii • endovascular therapy • Dyna CT • embolization • Onyx

Sinus pericranii was initially described by Stromeyer in 1850 as a “blood bag on the skull...in connection with the veins of the diploë and through these with the sinuses of the brain.” Sinus pericranii is an abnormal vascular communication of extracranial venous blood vessels with the intracranial dural venous sinuses via a transosseous diploic vein or emissary vein. The communication can be a complex network of thin-walled veins that forms a varix on the external table of the skull. This abnormal varix is continuous with pericranial and subgaleal veins.

In the majority of cases, the SP becomes apparent and symptomatic as a nontender, nonpulsatile soft-tissue mass, most commonly located on the frontal region close to the midline connected to the SSS though a skull defect. Off-midline lesions are extremely rare. We present a case of this rare condition that was successfully treated with endovascular embolization with the use of Onyx. To our knowledge, embolization with the use of Onyx as a definitive treatment of SP has not been previously reported.

Case Report

History and Examination. This 27-year-old woman presented with a discoloration of the skin at the level of the posterosalateral aspect of the left orbit associated with a nontender soft mass. The mass and discoloration had been present since early childhood, and they had extended and increased in size over the last years. The mass increased in size with the Valsalva maneuver. Findings on physical examination, including a full neurological examination, were otherwise unremarkable.

Contrast-enhanced MR imaging revealed multiple enlarged subgaleal vessels that enhanced homogeneously and showed flow void. The lesion extended from the left fronto-orbital region posteriorly to the temporal fossa, extending along the temporalis muscle on both medial and lateral surfaces (Fig. 1).

Cerebral angiography showed normal arterial and capillary phases. The delayed venous phase demonstrated abnormal communication of the most anterior aspect of the SSS via a midline frontal bone foramen (Fig. 2C) involving only the inner table of the frontal bone. This foramen contained a diploic vein (Fig. 2A and B) that connected the SSS with a large tangled venous structure that extended over the malar eminence and into the soft tissues of the infratemporal fossa.

Three-dimensional imaging with the use of DynaCT better delineates the intraosseous channel along the frontal bone and the connection from the anterior aspect of the SSS (Fig. 2D and E). The bone 3D reconstruction imaging documents the intraosseous channel with the external foramen just posterior to the left orbit (Fig. 2C). The venous channels then extend along the orbital process of the zygomatic bone and into the inferior temporal fossa (Fig. 2E and F).

Embolization and Postoperative Course. Under general anesthesia, a percutaneous injection of the frontal diploic vein (Fig. 3A) demonstrated multiple irregular channels with reflux into the most anterior aspect of the SSS. There was reflux into the contralateral small frontal diploic vein and anterograde flow into the multiple ve-
nous vascular channels located in the soft tissues of the left periorbital region. After the diagnostic arteriogram a microcatheter (Fig. 3B) was navigated into the junction of the SSS and the diploic vein, and multiple hydrocoils (Fig. 3D and E) and Onyx (Fig. 3C) were used to completely obliterate the connection between those structures. The postembolization right and left internal carotid artery injections demonstrated normal anterograde flow into the intracranial circulation. There was normal filling of the anterior aspect of the SSS without evidence of venous hypertension. There was complete disconnection from the anterior aspect of the SSS and the left frontal diploic vein. Minimal filling of the diploic vein in the right frontal region was detected. General anesthesia is recommended, although it is not mandatory, because Onyx injection can be painful.

The postoperative course was uneventful and the patient did not experience headaches or neurological deficits. The serpiginous extracranial veins decreased in size significantly after embolization. The patient was discharged home 2 days after the procedure.

Discussion

Sinus pericranii has been defined as “an uncommon, usually asymptomatic condition involving abnormal communication between the intra- and extracranial venous drainage pathways.” The communication usually occurs via dilated emissary or diploic veins that leave the skull through an aberrant skull defect that is usually lined with connective tissue or endothelial cells. The etiology of SP is unknown, although the condition was described as early as 1760 by Percival Pott. Some authors suggest an acquired pathophysiology because of the development of SP after head trauma, skull fracture, emissary vein tear, or birth trauma. However, the constant association with other vascular anomalies (for example, angiomas, aneurysmal malformation of an internal cerebral vein, subcortaneous venous cavernoma) supports a congenital cause such as failure of regression of the venous plexus
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between the periosteum and dura during early brain development or failure of the closure of intracranial sutures.\textsuperscript{10} The presence of endothelial lining defines a congenital SP and distinguishes it from the acquired type.\textsuperscript{7}

Sinus pericranii affects mainly young people, 80\% of cases occurring in patients younger than 40 years of age. It appears to occur with approximately equal frequency in males and female, although males may be more affected due to a higher incidence of head injury.\textsuperscript{2,15} The majority of SP cases occur along the midline, with 40\% of the lesions being frontal, 34\% parietal, 23\% occipital, and 4\% temporal.\textsuperscript{4} The case described in the current report is a rare instance of an SP located in the temporal fossa, although the skull defect where the emissary vein exited the skull was on the frontal region.

Patients with SP usually present with mild headache, localized tenderness, and a sensation of fullness due to the mass extension. The natural history is that of a slowly enlarging or persistent lesion. Differential diagnosis is varied and includes leptomeningeal cyst, encephalocele, epidermoid tumor, arteriovenous malformation, and cavernous hemangioma; further evaluation is always recommended before surgery, as the treatment for these conditions is different. Clinical findings of SP, demonstrated by manual and dynamic maneuvers (for example, Valsalva maneuver) on physical examination, combined with characteristic neurovascular imaging findings can help guide the correct diagnosis. A simple CT scan will show a nonenhancing soft-tissue mass along with an underlying skull defect. An MR imaging study with administration of contrast medium will demonstrate a soft-tissue mass of mixed signal intensity, with areas of flow void (Fig. 1). Invasive angiography will show the lesion in the venous phase (Fig. 2A and B), and as described above, Dyna CT angiography is very useful in showing the morphological characteristics of the vessels in a very detailed fashion and their relation to the bone structures as well as the drainage of the SP into the dural venous sinuses, usually the SSS (Fig. 3A–C). Cerebral angiography and venography should be performed before any treatment plan is formulated. Digital subtraction angiography is the best diagnostic modality for excluding other vascular malformations, including arterial and venous anomalies that can mimic SP, and it can demonstrate direct or indirect drainage of a venous anomaly into the SP, both of which could change the plan of treatment. Moreover, digital subtraction angiography can be used to assess the venous dynamics and their relation to the SP. In a recent angiographic classification based on flow dynamics, there were 2 different patterns of SP: 1) dominant, in which the venous flow uses the SP to drain the brain parenchyma and bypasses the normal venous outlets; and 2) accessory, in which only a portion of the brain's venous outflow occurs through the SP.\textsuperscript{6}

Treatment for SP is very controversial and has mainly been recommended for cosmetic reasons, for prevention of hemorrhage, and because of the possible risk of air embolism.\textsuperscript{1,13,17} Surgery has been the usual method of treatment. Typically, craniectomy of the involved bone has been proposed—with division of the diploic veins and cranioplasty.\textsuperscript{4} Although surgery is curative, some studies have reported significant hemorrhage as a result of dural
sinus lacerations. Other conservative approaches have been proposed, since the bone channels are often small enough to be occluded with bone wax.

In case of an SP in which the cerebral venous pattern is accessory, surgical or endovascular treatment can be an option; as previously described, treatment of a dominant SP is contraindicated because it may lead to venous congestion, venous infarction, or bleeding. In the present case, indications for endovascular treatment were relief of facial pain and the desirability of a minimally invasive intervention for cosmetic reasons. Endovascular treatment of SP with N-butyl cyanoacrylate glue has been proposed. Since its approval by the US Food and Drug Administration, the liquid embolic agent Onyx (ethylene-vinyl alcohol copolymer) has been used in the endovascular treatment of intracranial arteriovenous malformations, arteriovenous fistulas, and other venous malformations with excellent results. We decided to combine the use of coils and Onyx to obtain a fast obliteration of the dural vein, minimizing the number of coils used.

The current case also illustrates the efficacy of Dyna CT (Fig. 3A–C) in demonstrating the morphological characteristics of the SP and its relationship with osseous structures. In addition, digital subtraction angiography was used to demonstrate the flow pattern—in this case it was an accessory flow pattern; the emissary vein originates from the anterior portion of the SSS and connects to the SP located in the temporal fossa. We never observed dominant venous flow, so we proceeded with treatment. Onyx was used as the embolic agent (Fig. 3D–E). Based on previous good results obtained with Onyx for obliteration of dural and cerebral AVFs, the use of this agent in high–blood flow vessels is reliable and safe. Its use in low-flow vessels such as veins can be considered even safer.

Conclusions
Sinus pericranii is a rare venous anomaly that connects intra- and extracranial venous drainage pathways. We agree with the authors of previous reports that, based on angiographic findings, the assessment of venous drainage patterns is crucial to determine the possibility of treatment. As previously described, an endovascular approach can be a safe method for definitive treatment. To the best of our knowledge, this is the first report of endovascular treatment of an SP anomaly with the use of Onyx. As with surgery, the goal was to permanently occlude the intrasosseous connection of the SP to the dural sinus, and this was successfully accomplished.

Disclaimer
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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