Sinus pericranii in the setting of a posterior fossa pilocytic astrocytoma: illustrative case

Youngkyung Jung, MD,1 Jennifer L. Quon, MD, MHS,2 and James M. Drake, MSc, MBBCh1-3

Departments of 1Neurosurgery, 3Surgery, University of Toronto, Toronto, Ontario, Canada; and 2Department of Neurosurgery, Hospital for Sick Children, Toronto, Ontario, Canada

BACKGROUND Sinus pericranii (SP) is a rare vascular anomaly, with an uncertain etiology. Often discovered as superficial lesions, they can be primary or secondary in nature. Herein, we report a rare case of SP in the setting of a large posterior fossa pilocytic astrocytoma associated with a significant venous network.

OBSERVATIONS A 12-year-old male presented with acute clinical deterioration in extremis with a 2-month history of lethargy and headaches. Outside plain computed tomography imaging revealed a large posterior fossa cystic lesion, probably a tumor, with severe hydrocephalus. There was also a midline small skull defect at the opisthocranion, without visible vascular anomalies. An external ventricular drain was placed with rapid recovery. Contrast imaging revealed a large midline SP emanating from occipital bone with a large intraosseous, and subcutaneous venous plexus in the midline draining inferiorty into venous plexus around the craniocervical junction. A posterior fossa craniotomy without contrast imaging could have resulted in catastrophic hemorrhage. A small modified off-center craniotomy provided access to the tumor with a gross total excision.

LESSONS SP is a rare but significant phenomenon. Its presence does not necessarily preclude resection of underlying tumors, provided that a careful preoperative assessment of the venous anomaly is undertaken.

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KEYWORDS sinus pericranii; pilocytic astrocytoma; dural sinus hypoplasia

Sinus pericranii (SP) is an exceedingly rare low-flow venous anomaly characterized by an aberrant connection between the intracranial and extracranial venous system, most commonly involving the superior sagittal sinus.1 It most commonly presents as a nonpulsatile and palpable soft tissue lesion.

Primary SP exists as an isolated entity, whereas secondary variants are associated with anomalies such as dural sinus hypoplasia or craniosynostosis.2 SP can also be divided into dominant and accessory subtypes. In dominant SPs, most venous outflow occurs through the malformation, whereas with accessory SP, a minority of the venous outflow occurs through the SP.1

Here, we describe a rare case of secondary and dominant SP in the context of a pilocytic astrocytoma with significant intracranial hypertension and bilateral transverse sinus compression. Illustrative Case

A previously healthy 12-year-old male presented acutely with 2 months of ataxia, headaches, and lethargy. Upon presentation to hospital, he became apneic, bradycardic, with decreased level of consciousness necessitating emergent intubation. Noncontrast computed tomography (CT) demonstrated a large (6.4 × 6.2 × 5.9 cm) posterior fossa lesion, with mixed solid and cystic components (Fig. 1). The lesion produced significant mass effect, resulting in obstructive hydrocephalus, and crowding of the foramen magnum. Of note, there was also an associated bony defect superior to the torcula with bony erosions along the sagittal sinus.

Upon arrival to our center, the patient had a Glasgow Coma Scale (GCS) score of 9 (E2VTM6) and underwent emergent right frontal external ventricular drainage (EVD). Brain magnetic resonance imaging ABBREVIATIONS

CT = computed tomography; CTA = computed tomography angiography; CTV = computed tomography venography; EVD = external ventricular drain; GCS = Glasgow Coma Scale; MRI = magnetic resonance imaging; SP = sinus pericranii.

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(MRI) was subsequently performed and was notable for SP along the posterior fossa connecting the torcula to a series of prominent emissary veins along the occipital and suboccipital regions (Fig. 2). A computed tomography angiography (CTA)/computed tomography venography (CTV) was also performed to better visualize the vasculature for operative planning, demonstrating bilateral transverse sinus hypoplasia.

The patient underwent an image-guided posterior fossa craniotomy for resection of the cystic lesion. A lateral curvilinear skin incision was made to avoid the bony defect overlying the torcula and medial and inferior vasculature in the skin and underlying soft tissue. We could see several large veins draining in the underlying soft tissue of the skin flap. We performed a paramedian craniotomy that allowed lateral access to the mural nodule while avoiding the bony erosion of the torcula. Once the dura was open, the cystic lesion was readily identified using ultrasound guidance drained through a small corticectomy using an EVD catheter. After the cyst was decompressed the tumor was more readily visible under the medial aspect of the dural opening. There were several large veins extending up toward the tentorium and transverse sinus that we were careful to preserve while resecting the tumor.

The procedure was well-tolerated, and the patient was extubated the next day. He remained neurologically intact with no new postoperative deficits. Postoperative MRI demonstrated gross-total resection of the tumor with subsequent improvement in mass effect on the brainstem and cerebellum as well as a reduction in ventricle size (Fig. 3). The transverse sinuses were more visible on the postoperative magnetic resonance venogram (MRV), likely secondary to reduced compression (Fig. 3). However, the patient continued to have high EVD output, and we decided to proceed with an endoscopic third ventriculostomy, as there was some reluctance in passing a shunt tunnel near the venous network. Pathological evaluation by frozen section was consistent with a pilocytic astrocytoma.

Discussion

SP is an uncommon vascular lesion, the pathophysiology of which is unclear. There are theories of increased intracranial pressure and venous injury playing a role in the development of secondary SP. Although hydrocephalus and craniosynostosis have been associated with SP, intracranial tumors appear to be an uncommon etiology, with no prior reported cases in the literature.

Observations

There have been several documented cases of intracranial neoplasms with dural sinus invasion or compression with associated venous hypertension, none of which were reported to develop SP (Table 1). In one patient with metastatic prostate cancer, there was associated bilateral transverse sinus stenosis, which improved with radiation therapy and reduction of mass effect. In another patient with

FIG. 1. Noncontrast CTs. A: Axial view demonstrating a large cystic lesion in the posterior fossa. Note the supratentorial hydrocephalus with transependymal flow. B: Sagittal view showing a large cystic and solid lesion in the posterior fossa. There is a visible bony defect in the occiput and frontal bone. C: Sagittal view, bone window, showing a bony defect in the occiput with a preserved inner table, and bony erosions along the frontal bone, posterior to the coronal suture.

FIG. 2. Preoperative images. Axial T1-weighted MRI with contrast (A), demonstrating sinus pericrania coursing along the posterior fossa. Axial T1-weighted MRI with contrast (B) showing a network of emissary veins along the occipital region. Sagittal T1-weighted MRI with contrast (C), demonstrating a large cystic lesion causing obstructive hydrocephalus with effacement of the 4th ventricle. Magnetic resonance venogram, sagittal (D) and coronal (E) views, demonstrating bilaterally hypoplastic or compressed transverse sinuses, with occipital sinus pericranii connecting the torcula to a series of emissary veins.
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CN = cranial nerve; FU = follow-up; HA = headache.
an epidermoid cyst, there was diploic venous drainage, although no signs of SP.\textsuperscript{6} Of note, both cases were associated with compression of the torcular, as seen in our case report. In addition to occlusion of the torcular, a notable element of this case was the associated bony erosions, reflective of a chronic process, which is consistent with the slow-growing nature of pilocytic astrocytoma. Although the pathophysiology of SP remains unclear, it is possible this unusual case was a result of the chronic compression of the slow-growing pilocytic astrocytoma at the level of the torcular giving rise to the extensive network of venous drainage and bilateral sinus occlusions/hypoplasia. Furthermore, since this patient had not had prior imaging, we cannot exclude the possibility that there may have been an underlying vascular anomaly or SP. A formal diagnostic subtraction angiogram was not performed, and we cannot rule out other developmental venous anomalies, which are associated with primary SP.\textsuperscript{1,2} A primary SP in conjunction with compression on the torcular may account for the extensive venous hypertension, bony erosion, and expansive venous dilatation/vasculature seen in this case. Another unique consideration for this case was the inverse relationship between age and angiogenesis, which may account for the extensive vascular networks seen in this pediatric patient.\textsuperscript{5}

In respect to treatment options, generally, experts suggest only operating on secondary SP with adequate collaterals; dominant SPs are operated on, for risk of causing thrombosis of the limited venous outflow channels.\textsuperscript{1,2} There is a preference for endovascular options, namely, transvenous or percutaneous embolization or occlusion, with surgery reserved for cases with accompanying vascular lesions.\textsuperscript{2} Overall, treatment is suggested for cases causing significant persistent psychological distress for patients and their families, and associated venous malformations (e.g., arteriovenous malformations, developmental venous anomalies, etc.).\textsuperscript{1,2}

**Lessons**

SP is a rare vascular phenomenon that may occur with posterior fossa lesions. Recognizing this vascular variant prior to tumor resection is essential for operative planning due to the significant risk of significant intraoperative hemorrhage with a standard midline craniotomy. Further, sacrificing critical venous vasculature with a standard opening may additionally compromise the compensatory venous drainage system. To our knowledge, this is the first known report of SP in a pilocytic astrocytoma. We demonstrate here that a modified posterior fossa craniotomy is a safe approach for resection of such lesions.

Although we present a unique case of secondary SP, there are several limitations to our report. As previously mentioned, we did not perform a formal digital subtraction angiography to further characterize the vascular anatomy. This is also a single case report, limiting our ability to draw definitive conclusions regarding the epidemiology, pathophysiology, and natural history of this phenomenon.

**References**


**Disclosures**

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

**Author Contributions**

Conception and design: Jung, Quon. Acquisition of data: Quon. Analysis and interpretation of data: Quon. Drafting the article: Jung, Quon. Critically revising the article: all authors. Reviewed submitted version of manuscript: Jung, Quon. Approved the final version of the manuscript on behalf of all authors: Drake. Study supervision: Drake, Quon.

**Correspondence**

James M. Drake: Hospital for Sick Children, Toronto, ON, Canada. james.drake@sickkids.ca.