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Case Report

Dural-based infantile hemangioma of the posterior fossa: Case report

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Abstract

Background: The authors present the unique case of a dural-based, infantile hemangioma located in the posterior fossa of a 15-day-old infant.

Case Description: The patient presented with hydrocephalus. The lesion was identified by magnetic resonance imaging and was subsequently resected. Diagnosis of the lesion was confirmed with immunohistochemistry staining. The patient's hospital course was complicated by transverse sinus thrombosis and a cerebrospinal fluid leak that were treated with anticoagulation therapy and ventriculoperitoneal shunt placement, respectively.

Conclusion: Although hemangiomas are benign entities, our patient's lesion was in the posterior fossa causing compression and hydrocephalus that necessitated resection. We encourage others to consider the possibility of hemangioma in the differential diagnosis of dural-based posterior fossa lesions in infants.

Key Words: Glucose transporter 1, infant, intracranial hemangioma, neonatal ultrasonography, posterior fossa

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INTRODUCTION

The most prevalent benign neoplasm of infancy is the infantile hemangioma.[2] These lesions primarily occur in extracranial locations such as the skin, whereas intracranial hemangiomas are a less frequently observed entity. We present a unique case of a dural-based posterior fossa infantile hemangioma in a 15-day-old infant.

CASE REPORT

The patient is a 15-day-old dizygotic twin girl born via uncomplicated vaginal delivery. She was referred to our institution as a result of findings of ventriculomegaly noted on prenatal screening ultrasonography. The cause of the hydrocephalus was unknown at that time. On examination, the patient had a soft but full anterior fontanelle, along with dilated scalp veins. The head circumference was greater than the 97th percentile for gestational age. No focal neurologic deficits were appreciated on examination. Postnatal cranial ultrasonography revealed progressive hydrocephalus as well as a midline echogenic mass in the posterior fossa with mass effect on the cerebellum. Magnetic resonance imaging (MRI) was obtained, which revealed

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a very large dural-based extraaxial mass along the dorsal aspect of the posterior fossa [Figure 1]. The mass was well-circumscribed and heterogeneous, but mostly T1 isointense and T2 hypointense with flow voids and evidence of previous hemorrhage within it. Avid contrast enhancement was seen. There was marked mass effect on the cerebellum, fourth ventricle, and brainstem with resultant obstructive hydrocephalus and evidence of transependymal flow. The differential diagnoses of an intracranial vascular lesion in this 15-day-old infant included hemangioma, hemangioblastoma, vascular malformation, and other neonatal tumor types, such as soft-tissue sarcoma.[10] In addition, meningioma could not initially be definitively excluded because of the dural attachment.

Owing to the compressive nature of the lesion and the progressive hydrocephalus, the infant was taken to the operating room for resection of the posterior fossa lesion. Prior to the resection, an external ventricular drain (EVD) was placed in the occipital horn of the right lateral ventricle with care to avoid the egress of cerebral spinal fluid until the tumor was resected. A suboccipital craniectomy and C1 laminectomy were performed. The lesion appeared extremely vascular and clearly emanated from the dura without intraaxial involvement. Because of the patient's size and the low circulating blood volume, minimizing blood loss was imperative. Blood products were on standby. Care was taken not to breach the venous system as the mass was circumferentially dissected away from the surrounding tentorium and brain parenchyma. Cavitron ultrasonic aspiration and bipolar cautery were used to reduce the blood supply to the tumor externally while it was debulked internally. Following resection of the lesion, a large dural defect remained. A dural graft was used to close the defect with the additional support of a large piece of non-suturable DuraGen (Integra Life Sciences, Plainsboro, New Jersey) and Tisseel (Baxter Healthcare, Deerfield, Illinois) because the paper thin nature of the patient's dura made watertight closure challenging.

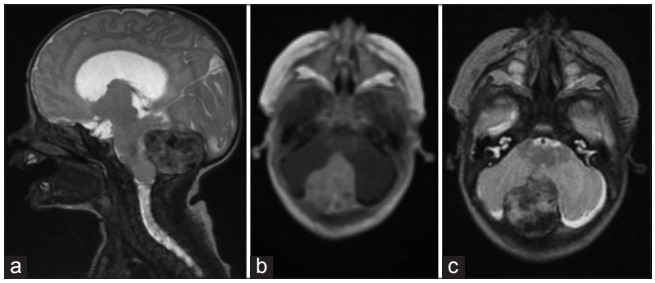

Figure 1: Preoperative images. (a) Sagittal T2 image demonstrating a heterogeneous posterior fossa lesion with compression of the cerebellum and enlargement of the supratentorial ventricles. (b) Contrast-enhanced axial T1 image of the posterior fossa lesion demonstrating avid contrast enhancement. (c) Axial T2 MRI showing significant heterogeneity within the lesion with effacement of the fourth ventricle

Because of our patient's small body habitus (2.7 kg), minimizing hemorrhage and maintaining intravascular volume were of utmost importance. In sum, throughout the duration of the case, our patient received 160 ml of lactated Ringer's solution, 90 ml of packed red blood cells, 50 cc of fresh frozen plasma, 40 ml of platelets, and 10 ml of cryoprecipitate. Despite efforts taken to reduce volume loss, the estimated blood loss was 150 ml, whereas the patient's estimated total circulating blood volume was only 230 ml and urine output was 50 ml.

Postoperative imaging revealed gross total resection of the mass and significant decompression of the brainstem and cerebellum [Figure 2]. The immediate postoperative course was complicated by seizures and thrombosis of the right transverse sinus, for which therapy with levetiracetam and Lovenox (Sanofi-Aventis, Bridgewater, New Jersey) was started. Initially, intracranial pressures were normal and a several-day trial of EVD clamping was successful, allowing the removal of the EVD. Unfortunately, during the follow-up period, the patient developed a persistent cerebrospinal fluid (CSF) leak, enlarging head circumference, and enlarging ventricular size. Infection was excluded by CSF sampling. Ultimately, spinal fluid diversion with ventriculoperitoneal shunt placement was required.

The pathology report of the lesion showed positive staining of 20% for MIB-1, a marker for actively proliferating cells. Stains for glucose transporter 1 (GLUT1) and CD31 were also positive, indicating that the lesion was an infantile hemangioma.[5,6]

At the time of the most recent office evaluation 4 months since her operation and 6 weeks from shunt placement, the patient had completed her course of anticoagulation therapy with follow-up imaging demonstrating patency of all venous sinuses. There has been no recurrence of the mass. She remains on antiepileptic medications for

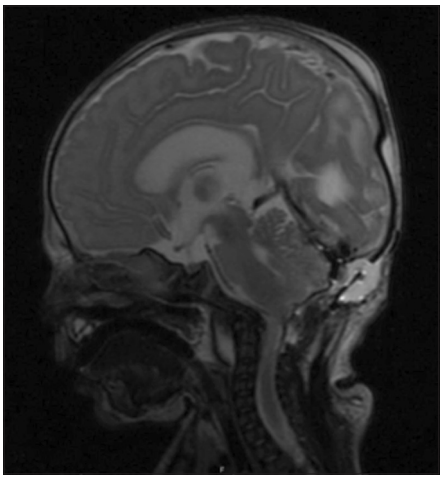

Figure 2: Postoperative sagittal T2-weighted image showing gross total resection with decompression of the cerebellum

seizure prophylaxis but has not had seizure activity. The patient continues to meet her developmental milestones in subsequent outpatient follow up visits.

DISCUSSION

One percent of infantile hemangiomas involve the central nervous system, and approximately half of these are intracranial.[10] Most intracranial infantile hemangiomas classically involve the extraaxial basal cisterns, subarachnoid and ventricular spaces, and cavernous sinus.[9] One study describes a case of an infantile hemangioma in the fourth ventricle; however, there was no evidence of dural attachment.[4] There have been two reported cases of dural-based infantile hemangiomas in the literature, neither of which was located in the posterior fossa [Table 1].[9,11] As described in a previous report of cases of infantile hemangioma involving the central nervous system,[10] our case demonstrates similar findings on MRI, including heterogeneity and avid contrast enhancement. Given these classic findings, we suspected that this was a vascular lesion. The intraaxial versus extraaxial nature of the lesion was not clearly delineated until resection. Confirmation of the final diagnosis was revealed on immunohistochemical staining, which included GLUT1 and CD31, both of which are reliable markers for infantile hemangioma.[5,6] GLUT1, a glucose transporter present in normal capillary endothelial cells constituting the blood–brain barrier, is a particularly useful marker of infantile hemangiomas because it differentiates those from other vascular tumors and malformations that have been shown to lack GLUT1.[7,8] In addition, the presence of GLUT1 may indicate that the tumor is in a proliferating stage rather than an involution stage as GLUT1 positive cells have been shown to display properties of facultative stem cells.[3,12] Histologically, infantile hemangiomas classically show lobular, solid, and infiltrative patterns with small capillaries, plump endothelium, and a thin basement membrane.[10] Several other associated abnormalities have been observed in infants with infantile hemangioma, including arterial anomalies, ipsilateral cerebellar hypoplasia, Dandy–Walker malformation, moyamoya-like proliferation of the internal carotid artery terminus, and other manifestations of the posterior fossa–hemangioma–arterial lesions–cardiac abnormalities/aortic coarctation–eye abnormalities (PHACE) syndrome.[10] However, our patient's infantile hemangioma appeared to be an isolated finding.

Despite the benign nature of intracranial infantile hemangioma, mass effect from these lesions can cause considerable and permanent damage to the developing brain. Therefore, intervention is required, especially in the face of hydrocephalus, seizures, or focal neurologic deficits. Because our patient had progressing hydrocephalus with enlarged head circumference and effacement of the fourth ventricle with mass effect on the cerebellum, we did not believe a trial of steroids or propranolol would be a reasonable management option, especially because the diagnosis had yet to be confirmed. However, if the lesion had been found incidentally in a less dangerous location and the diagnosis was biopsy proven, conservative management with observation and corticosteroids may be considered because cases of regression after medical therapy have been reported.[1,10] Our patient exhibited marked hydrocephalus as do 10% of patients with intracranial infantile hemangiomas.[9] Two proposed mechanisms of hydrocephalus due to infantile hemangioma include thrombosis of surrounding small venous structures and mass effect from the lesion causing an obstructive hydrocephalus. In our patient, the latter is likely the etiology given the compressive nature of the lesion on the fourth ventricle. Although a vascular lesion of this caliber in adults would require potential preoperative embolization and more extensive vascular studies, concern for iatrogenic injury was great given our patient's small body habitus and small caliber of vessels. Therefore, it was concluded that the overall risk-benefit ratio of preoperative embolization did not warrant such an intervention.

The postoperative transverse sinus thrombosis seen in our patient was likely due to direct surgical manipulation and retraction of the brain parenchyma and venous structures. Volume loss during the procedure may also have been a contributing factor. Our patient did not experience any other signs or symptoms of hypercoagulability, nor is there any reported association of infantile hemangiomas with a hypercoagulable state. In addition, the postoperative timing of the thrombosis and its close proximity to the surgical site lead us to believe it was a sequela of the surgical procedure rather than of the disease process itself.

Table 1: Case reports of dural-based infantile hemangiomas

Authors, year	Age, sex	Presentation	Location	Diagnosis confirmation	Management	Complications
Willing *et al*., 1993[11]	17 months, male	Generalized and focal seizures	Right temporal	Histopathology (no staining used)	Resection	Postoperative seizures
Philpott *et al*., 2012[9]	12 months, girl (monozygotic twin)	Enlarged head circumference	Right parietal	Immunohistochemistry (GLUT1)	Resection	None
Shakir *et al*.	15 days, girl (dizygotic twin)	Ventriculomegaly on screening ultrasound	Midline posterior fossa	Immunohistochemistry (GLUT1)	Resection	Postoperative seizures, sinus thrombosis

CONCLUSION

Although hemangiomas overall are common in infants, the unique nature of our case report stems from its location in the posterior fossa and dural attachment. To our knowledge, this has not been described in the literature. The diagnosis was suspected by classic MRI features and was confirmed by histopathology staining including GLUT1 and CD31. Although hemangiomas are benign entities, our patient's lesion was in the posterior fossa causing compression and hydrocephalus that necessitated resection. We encourage others to consider the possibility of hemangioma in the differential diagnosis of dural-based posterior fossa lesions in infants.

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Conflicts of interest

There are no conflicts of interest.

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