Management of midline dural sinus malformations

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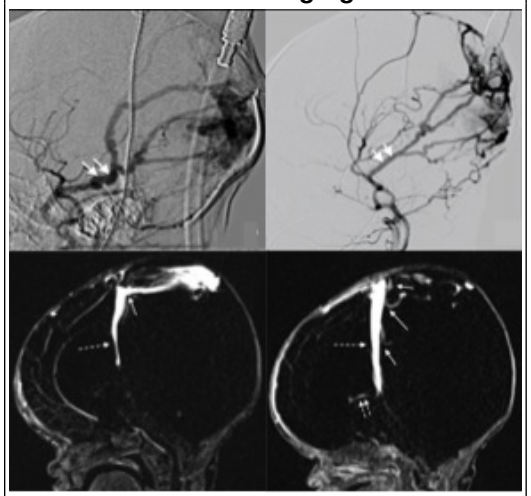
Introduction

Dural sinus malformations (DSMs) are rare pediatric vascular malformations. Two types of DSMs have been recognized: a midline type involving the posterior sinus with giant dural lakes and slow flow mural AV shunting, and a lateral type involving the jugular bulb with otherwise normal sinuses and associated high flow sigmoid sinus arteriovenous fistula. Midline DSMs have been reported to have a poor prognosis and have been traditionally more difficult to cure. We describe three cases of DSMs and discuss the treatment strategy employed for each lesion.

Initial imaging



Top: Patient 1-cerebral angiogram showing multiple AV shunts from the middle meningeal, vertebral, and occipital arteries. Bottom: Patient 2-venous channel at the anterior margin of the DSM, multiple sites of contrast extravasation from the posterior wall into the DSM.

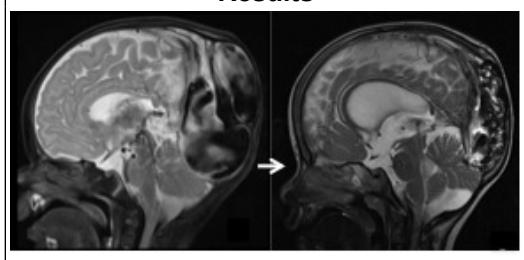
Cases

Patient 1 presented with cardiac failure, intracranial hemorrhage, and seizures. She had a large midline DSM with multiple high flow dural and pial AV shunts. She underwent several staged transarterial embolization of feeders of the DSM, which allowed gradual reduction in flow through the lesion. This was followed by percutaneous approach to eliminate the associated venous pouch. The combination of these procedures achieved complete obliteration of the DSM.

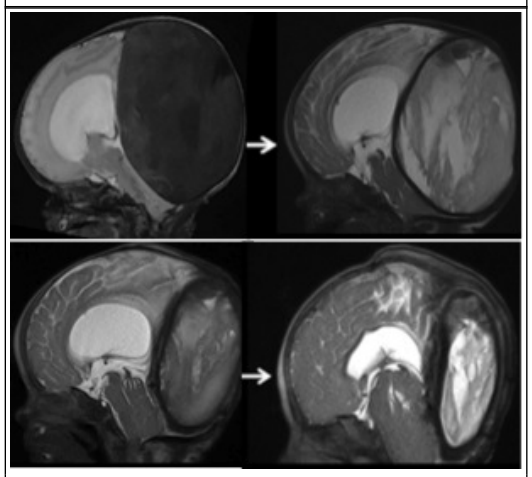
In the second patient, a large midline DSM with no AV connections was identified prenatally and significantly enlargered prior to birth causing ventriculomegaly and brainstem compression. This patient underwent several procedures including multiple percutaneous embolizations, placement of a ventricular access device followed by a shunt, posterior fossa decompressive craniectomy, and cranial expansion. Final imaging showed decrease in the venous pouch with no acute extravasation.

The third patient was found prenatally to have a large DSM involving the torcula with no evidence of ventriculomegaly or brainstem compression. On serial imaging, the DSM continued to decrease in size despite no intervention. She was born without complication and has no symptoms attributable to the DSM.

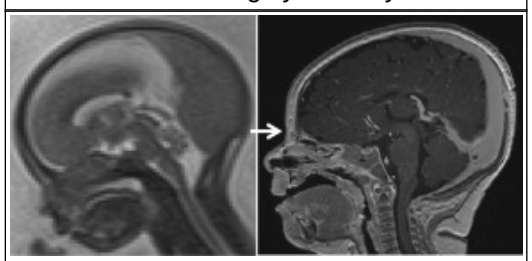
Results



Patient 1: Decrease in thrombosed midline venous pouch after endovascular embolization



Patient 2: Significant reduction of dural sinus malformation and decreased ventriculomegaly over 1 year



Patient 3: Fetal MRI showing a large midline dural sinus malformation. Follow-up MRI shows a decrease in the size of the lesion.

Results

Patient 1 was meeting developmental milestones at 24 month follow up with imaging showing marked decrease in the thrombosed midline venous pouch and no new AV shunts.

At 12 month follow-up, Patient 2 was meeting most developmental milestones, with some motor delay. Serial MRI imaging revealed significant reduction in the size of the DSM and decreased ventriculomegaly.

Patient 3 continues to do well with no intervention and on follow-up imaging at 5 months of age, the DSM remained stable in size with near complete resolution of the prior thrombus within the torcular Herophili and the left transverse sinus.

Conclusions

Early diagnosis and treatment, if necessary, of DSMs is critical to prevent cardiac failure or parenchymal injury from chronic venous hypertension. Management should be decided on individual case basis depending on the angioarchitecture and progression of the lesion. Options including observation, percutaneous/endovascular embolization and adjuvant neurosurgical decompression with CSF diversion is shown in our cases to treat these lesions with good outcomes.