

Case Report

Torcular Dural Sinus Malformation in the Early Childhood: A Case Report

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Abstract

Arteriovenous malformations (AVM) are rare diseases in pediatric age groups, and dural sinus malformation (DSM) has a lower incidence rate. DSMs are associated with mild male dominance, and onset symptoms appear around 5 months of age. The most common clinical presentation is macrocrania, seizures, psychomotor delay, intracranial hemorrhage, congestive heart failure, and brain ischemia. Early recognition of these lesions is essential to prevent brain injury for ischemia and intracranial hypertension. Case description: We discuss the case of a 8 month-old boy presenting with macrocrania secondary to torcular dural sinus malformation. This case was successfully treated by the endovascular procedure, reaching the goal of the treatment to obliterate the arterial portion of the fistula while preserving cerebral venous drainage to reduce the pial reflux to prevent venous hypertension and ischemic complications.

Keywords: Torcula, Dural sinus malformation, Dural arteriovenous shunt.

Abbreviation:MMA- Middle meningeal artery, OA- Occipital artery.

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Introduction:

Arteriovenous malformations (AVM) have a low incidence with an annual case detection rate estimated as 0.9-1.5 per 100,000 population worldwide ¹. Dural sinus malformation (DSM) has even

a lower incidence, representing 4.8% to 10% of the AVM in the pediatric age group ^{2,3}. We discuss the case of a 8-month-old boy presenting a torcular dural sinus malformation successfully treated by the endovascular procedure.

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Case Report:

A 8 month-old boy was admitted to the neurosurgery department with macro-cranium and irritability. General examination reveals: height is 66 cm (15th to 50th percentile) and weight is 6.2 kg (15th percentile). Head circumference is 49 cm (more than 97th percentile), bulged tensed fontanelle, occipital cranial bruit with sutural diastasis on local examination of the head. There is good eye contact with regular eye movements; pupils are equal bilaterally with normal light reactions. He has no motor deficit, and he can sit. CT scan of the brain (Figure 1A) shows a substantial isodense occipital lesion with enlarged ventricles and periventricular lucency. Contrast CT scan (Figure 1B) shows excellent homogenous contrast enhancement with signs of venous HTN.

A magnetic resonance imaging (MRI) (Figure2) shows the presence of obstructive hydrocephalus with a

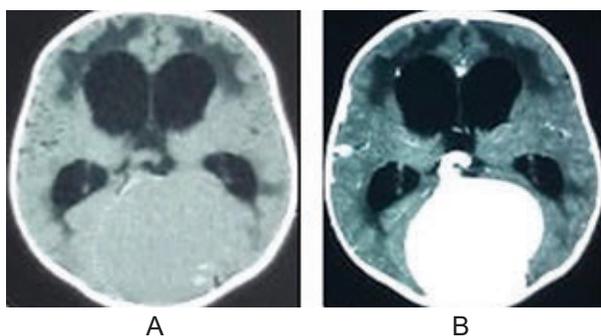


Fig.-1: CT scan A) plain; B) contrast; occipital contrast-enhancing lesion, venous HTN, has hydrocephalus.

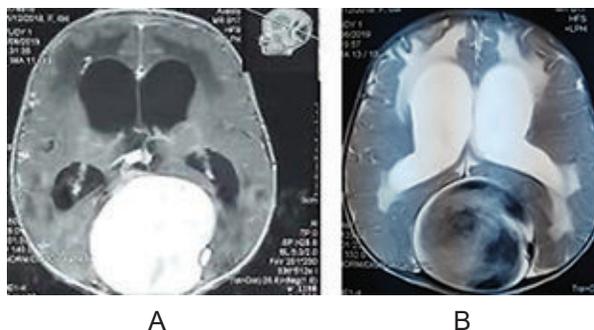


Fig.-2: MRI A) Homogenous contrast uptake; B) T2 flow void lesion.

contrast-enhancing extradural large, round heterogeneous but predominantly high dense occipital lesion. On T2WI, there is a central hypointense signal suggestive of flow void, which was diagnosed as vascular abnormality (Figure3).

The patient was referred to a digital angiography which showed dural direct arteriovenous fistula fed by both middle meningeal and left occipital artery and drained to the torcula leading to enlargement of the transverse sinus, sigmoid sinus, including the torcula, with no signs of jugular bulb stenosis, pial reflux or cavernous sinus abnormality.cavernous sinus abnormality.

A two-staged endovascular closure (21 days apart) was chosen for management. The goal was intra-arterial catheterization with complete occlusion at the union of the artery and torcula. The navigation was performed using Apollo (Medtronic) catheter with a 0.0008 Chikai microwire. The material used to treat the fistula was ONYX 18.

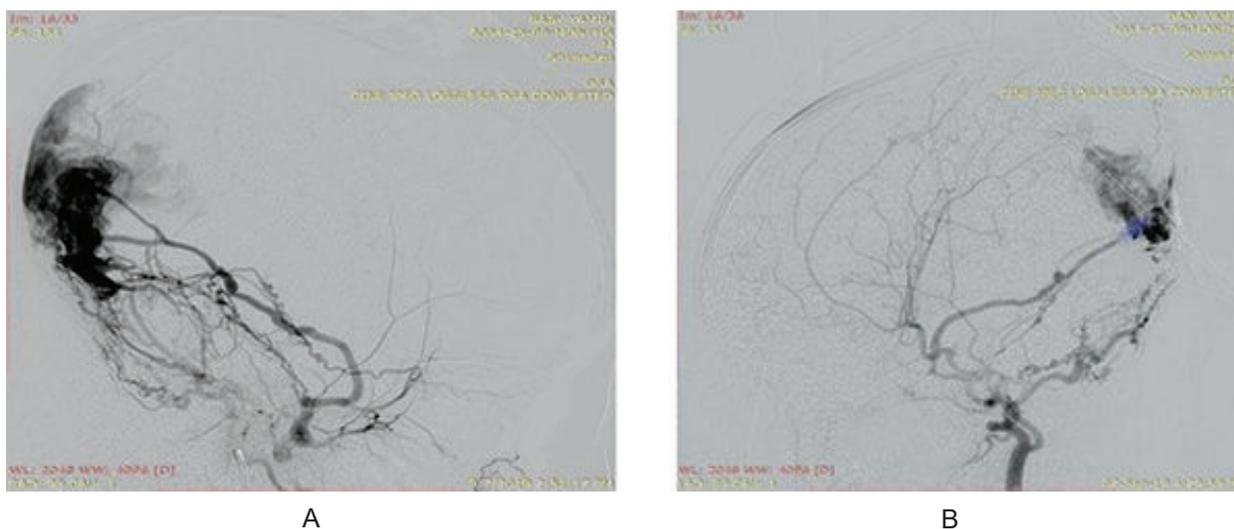


Fig.-3: Venous Lake with feeder A) Right MMA; B) Left MMA & OA.

The first session was carried through the right middle meningeal artery and partial embolization (Figure 4 A, B, C). After twenty-one days, the rest of the embolization was completed through the left middle meningeal and left occipital artery (Figure 5-A).The

post-embolization angiography showed complete resolution of the fistula (Figure 5-B).

Postoperative contrast CT scan shows a hypodense aspect with an essential decrease in size with periventricular lucency and no signs of venous HTN with visible sulci & gyri (Figure 6).

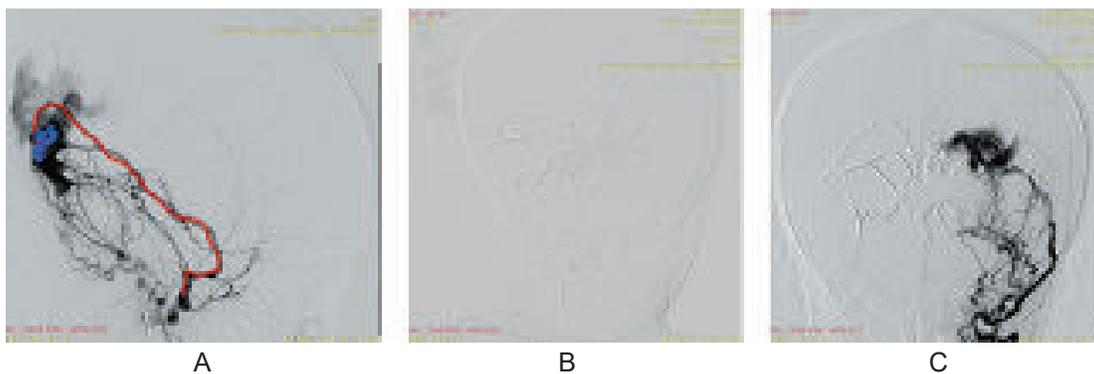


Fig.-4: Right MMA access A) Route; B) After embolization; C)Residual.

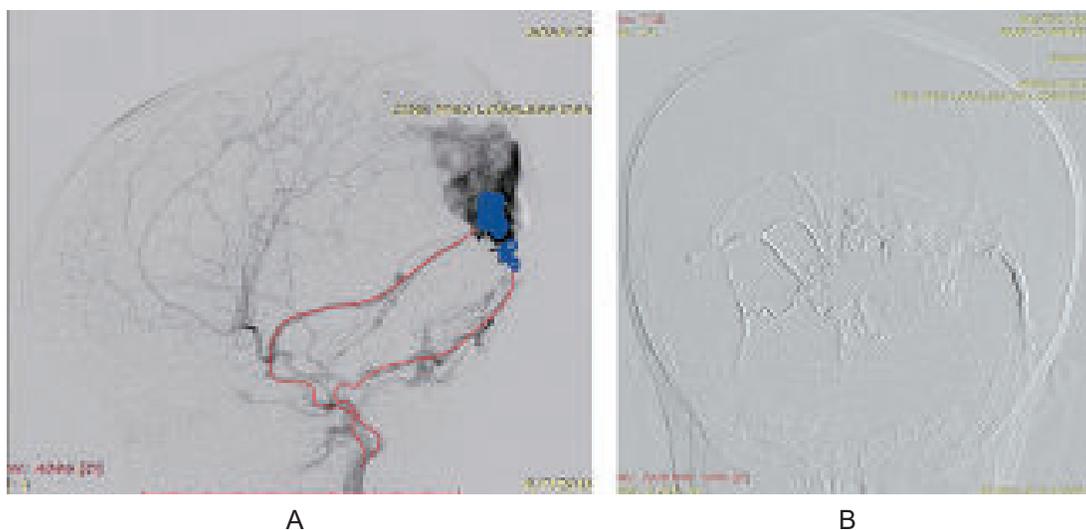


Fig.-5: A) Embolization through left MMA & OA. B)Complete embolization.



Fig.-6: CT scan shows thrombosed lesion with no signs of intracranial HTN.

Discussion:

DSM is the consequence of non maturation of a sinus segment, and it leads to secondary arteriovenous shunt in the affected part of the sinus. So DSM with their mural arteriovenous shunt (AVS) is different from neonatal and infant AVS, since shunts in DSMs seem to be a secondary phenomena at the sinus level, usually with low flow characteristics². In a recent series of DSM with giant lakes published by Barbosa et al.², studying AVM in the pediatric age group, a vein of Galen malformation (VGAM) is the primary pathology, representing 51.1%. Dural arteriovenous shunts (DAVS) represented 8.3% of all AVM in children². Within the pediatric DAVS, three types of lesions can be individualized: 1) DSM; 2) infantile type of DAVS, and 3) adult type of DAVS.

DSM is a type of lesion in which the arteriovenous shunts are usually secondary and accessory to the sinus malformation. Infantile type of DAVS are often multifocal, with no sinus malformation, however present with large sinus and sometimes, secondary jugular occlusion. In the adult form of DAVS, the venous sinus are usually small and some times partially thrombosed and can be secondary to another local event⁷. DSM with giant lakes involving the torcular and/ or adjacent sinuses represent 57.7% of all DAVS in children². Therefore, DSM with giant lakes represents 4.8% of all AVM².

DSM is associated with a male dominance (1.9:1), and the DSM symptoms usually appear around 5 months of age, although some lesions are identified in the antenatal period. Some of the most common clinical presentations are macrocrania, seizures, psychomotor development delay, intracranial hemorrhage, congestive heart failure, and brain damage^{2,5-7}.

Although the high mortality rate, about 38% to 46%, endovascular management is the first therapeutic choice^{2,4,6}. Other essential factors are medical therapy with support to the treatment of cardiac failure if present and anticoagulation⁴. The goal of the treatment is to obliterate the arterial portion of the fistula while preserving cerebral venous drainage to reduce the pial reflux to prevent venous hypertension and ischemic complications^{2,4,7}. According to Barbosa et al.², 75% of the patients who were able

to be submitted to endovascular therapy presented good outcomes, with only minor neurologic deficits.

The prognosis of such lesions is usually poor, with an overall mortality of 38%⁶. Barbosa et al.² highlighted some harmful prognostic factors: brain damage, DSM involving torcula and absence of cavernous capture; and good prognostic factors: DSM away from the torcula, bilateral presence, and bilateral presence flow at cavernous sinuses, and absence of jugular bulb dysmaturation.

Conclusion:

Early recognition of these lesions is essential to prevent brain injury for ischemia and intracranial hypertension. The endovascular treatment represents the gold standard option for such lesions. Although the development of embolization techniques in the last years, new technologies and further understanding of the disease are needed to achieve better treatment results.

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