

NEONATAL GIANT DURAL SINUS ECTASIA: A CASE REPORT AND REVIEW OF LITERATURE

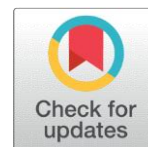
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ABSTRACT

Background: Neonatal giant dural sinus ectasia (DSE) is a rare congenital vascular malformation of the posterior major dural venous sinuses, often presenting as giant venous pouch. Subsequently, thrombosed lesions can progress spontaneously, and the lesions are accompanied by high flow dural arteriovenous fistulas (dAVFs) resulting in high rates of morbidity and mortality.

Case Presentation: Our case about full term female neonate who developed respiratory distress, severe cyanosis, and high flow cardiac output failure immediately after birth, was initially mimicking vein of Galen malformation and Ebstein anomaly featured on cranial ultrasound and bedside echocardiogram.

Neuroimaging MRI, MRA and MRV findings emphasize the complex giant DSE noted at the torcular herophili with high flow dAVFs, supplied by the middle meningeal and right occipital arteries, which was complicated with venous thrombosis.

Then, on day 6 of life, transcranial endovascular embolization was done and intraoperative course and unfortunately was complicated by severe access site hemorrhages and 20 minutes' cardiac arrest receiving urgent resuscitation. Then procedure resumed requiring coils and embolic liquid agents, demonstrating reduction in size of shunt by more than 50%. However, post intervention, the patient developed severe metabolic acidosis, and consumptive coagulopathy, bilateral high grade GMHs and stage III acute kidney injury, despite maximal quadruple inotropic support as well as

peritoneal dialysis catheter placement. Eventually, on day 9 of life, she succumbed to refractory shock and multiorgan failure.

Conclusion: Neonatal DSE with coexisting high flow dural AVFs represents a life-threatening entity competent of precipitating rapid, fatal multiorgan failure despite urgent intervention. Overall, prognosis determined by arteriovenous degree shunting, impairment of venous outflow, and secondary brain parenchyma injury.

Keywords: Dural Sinus Malformation, Neonate, Vascular Malformation, Neuroimaging, Intracranial Sinus Thrombosis, Dural Sinus Ectasia

1. INTRODUCTION

Neonatal dural sinus ectasia is a rare dural venous sinuses congenital malformation, it usually involving the torcular herophili or adjacent major posterior sinuses and often described as a giant venous lake or pouch, occasionally associated with thrombosis or arteriovenous shunting. [1-4] Although uncommon, it is clinically important because it can produce mass effect, venous hypertension, cardiac insufficiency, coagulopathy, hydrocephalus, cerebral ischemia, and death in severe cases. [2,4, 5-8]

2. BACKGROUND

The most cases were detected incidentally on prenatal routine ultrasound, as posterior fossa mass or midline extraaxial mass. However, the fetal MRI is helpful to confirm the venous origin, asses for compression or associated brain injury as well as define thrombosis. [6,8-12]

The presentation of diseased neonates ranges from thrombosed asymptomatic lesions, to bulging fontanel, macrocrania, hydrocephalus, seizures, cranial bruit, respiratory compromise, disseminated intravascular coagulation and congestive heart failure. [1, 4-6, 8]

Reported cases of congenital ectasia at the torcular herophili with thrombosis and bilateral polymicrogyria, suggest early fetal venous ischemia, may contribute to

neonatal cortical malformations, with Neuroimaging MR playing diagnostic key role. [6]

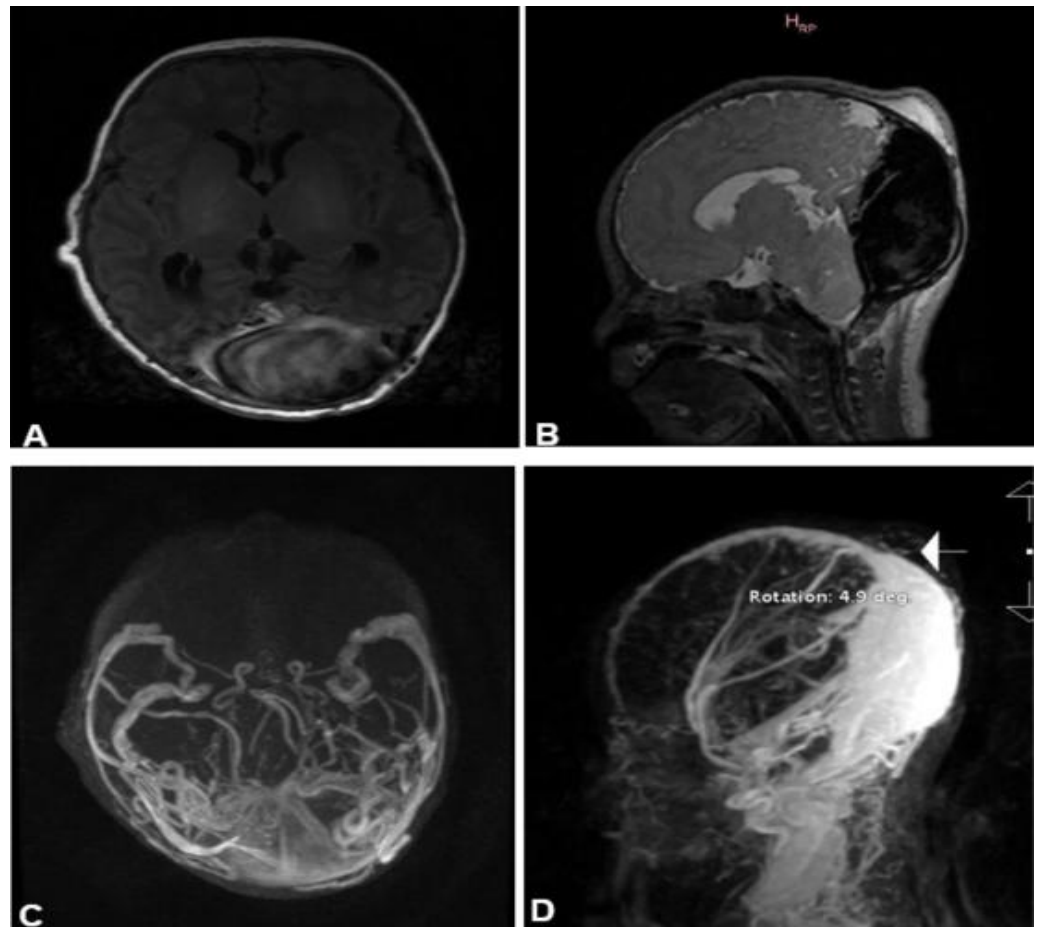


Figure 1. 7 Day girl case of DSE MRI: Feature of dural sinus malformation, in particular giant dural venous sinus ectasia at the confluence of sinuses with mass effect. Large extra-axial heterogeneous T1 hypointense lesion occupying and expanding the region of confluence of sinuses consistent with giant dural venous sinus ectasia (A) it associated with mass effect in the form of splaying of the posterior aspect of interhemispheric fissure with superolateral displacement of the bilateral posterior hemispheres, compression of the occipital and bilateral temporal lobes and dilatation of the temporal horns (A, B). anterior displacement and compression of the cerebellum with partial effacement of the fourth ventricle. The inferior cerebellar leaflets are inferiorly displaced (B). The giant ectatic dural venous sinus associated with dilatation of the left superior petrosal sinus, posterior emissary veins, and bilateral internal jugular veins. Suspected multiple coexisting arteriovenous fistulas. Preserved flow in anterior and posterior circulations arteries with diffusely reduced caliber which could be attributed to AV shunting with steal phenomenon (C,D).

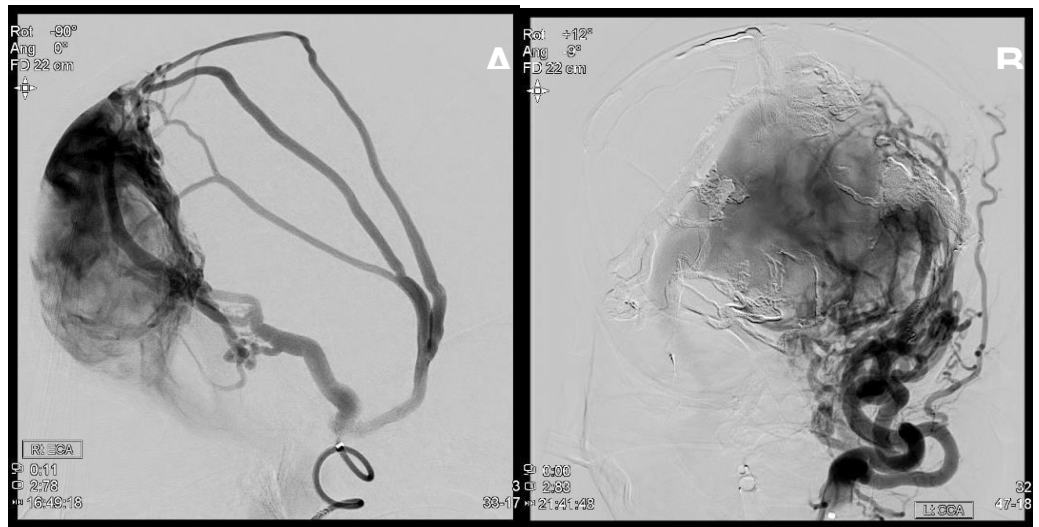


Figure 2. 7 Days girl case of DSE, status post trans-arterial embolization: A complex of high flow arteriovenous shunting representing of dural sinus malformation with arteriovenous fistula (A) Successfully targeted embolization of bilateral middle meningeal arteries and right occipital artery distal branches using liquid embolic material achieving more than 50% (B).

3. CASE PRESENTATION

A full term female neonate, born at 37 weeks of gestation via emergency cesarian section due to decreased fetal movement and failure to progress, presented immediately after birth with severe respiratory distress, cyanosis, and marked cardiomegaly. Initial echocardiography raised suspicion for Ebstein's anomaly, promoting transfer to a tertiary centre where a bedside cranial ultrasound instead revealed features highly suggestive of a vein of galen malformation alongside 0.3 cm right caudothalamic groove cyst. Due to progressive respiratory failure and worsening high cardiac output failure state, the patient was selectively intubated and mechanically ventilated on her second day of life to facilitate advanced neuroimaging.

Brain MRI, MRA, and MRV confirmed a complex dural sinus malformation characterized by a giant extra-axial dural venous sinus ectasia at the confluence of sinuses, multiple coexisting dural arteriovenous fistulas supplied predominantly by the tortuous bilateral middle meningeal and right occipital

arteries, and signs of perinatal venous ischemia with punctate ependymal hemorrhages.

Refractory high output heart failure necessitated an urgent, first stage transarterial endovascular embolization on day six of life using liquid embolic materials and coils, which successfully achieved greater than 50 percent shunt reduction but was intraoperatively complicated by severe access site bleeding and a 20 minutes cardiorespiratory arrest requiring resuscitation before returning to the NICU. Postoperatively, the patient exhibited profound hemodynamic instability, severe metabolic and respiratory acidosis, and progressive bilateral periventricular intraparenchymal hemorrhages on follow up cranial head ultrasound.

A post procedural echocardiogram demonstrated severe systemic pulmonary hypertension, an engorged right heart with moderate to severe tricuspid regurgitation, a patent foramen ovale and patent ductus arteriosus with persistent right to left shunting and severe left ventricular underfilling.

The clinical course rapidly deteriorated into multi organ failure, marked by a progression from severe oliguria to complete anuria secondary to stage three of acute kidney injury and profound fluid overload/ anasarca.

Despite emergency bedside insertion of a peritoneal dialysis catheter, which could not be utilized due to shock, maximum quadruple inotropic support, and aggressive blood product transfusions for secondary consumptive coagulopathy, the neonate developed severe bradycardia and unrecordable oxygen desaturation, ultimately dying from refractory shock on her ninth day of life.

4. DISCUSSION

The giant dural sinus ectasia in neonates is an extremely rare congenital venous malformation with variable anatomy, clinical course, and outcome, so individual case reports remain useful for defining the diagnosis and prognosis. [1,3-4,16,17]

The appearance of this entity is typically giant venous pouch or lake centered on the torcular or adjacent dural sinuses and usually associated with thrombosis and sometimes accompanied with arteriovenous shunting. [1-3, 17,18]

4.1 Interpretation

Our present case fits the reported pattern of prenatal or early neonatal detection, and often after identification of the posterior fossa mass usually seen on midline extra axial on ultrasound scan. The MRI findings clarifying venous origin, mass effect or even associated thrombosis and further complications. [2,7,11,16]

Distinguishing the imaging findings from other similar entities matters because giant dural sinus ectasia can mimic extra-axial lesions, for example; extraaxial hemorrhage, brain tumors, subdural collection or cysts, and vein of galen malformation and misclassification lead to delay appropriate monitoring and proper time intervention. [1,4,7-8]

The disease nature history was variable, earlier studies mentioned high mortality and severe neurological morbidity, particularly in lesions involving the torcular or accompanied by the brain damage and persistent shunting, whereas more recent studies demonstrate that isolated thrombosed lesions is not merely a complication but often part of the lesion's natural evolution, with slow flow predisposing clot formation, can regress spontaneously and have favorable neurodevelopmental outcomes, in selected neonates managed conservatively. [1-6, 13-14]

4.2 Prognostic Context

The cases were accompanied by cardiac failure, venous hypertension, parenchymal injury, ventriculomegaly, venous hypertension, and persistent arteriovenous shunting, has been reported with poorer outcomes. [1,8,15,17]

Favorable outcomes is reported in most evolved isolated thrombosed lesions with preserved brain parenchyma, and the absence of ventriculomegaly, particularly

when serial imaging demonstrates decreasing size and increasing internal heterogeneity, without persistent arterial feeders. [3,4,9]

Appearance of thrombosis as frequent part of lesion evolution that may accompany spontaneous involution, it can also worsen to serious complications as progressive thrombosis to consumptive coagulopathy and venous outflow obstruction, as well as rapid postnatal decompensation even when limited initial symptoms. [1, 6-7, 11]

4.3 Prognostic Features

Table 1: Reported prognostic features in neonatal or fetal dural sinus malformation/ectasia.

Feature	Association
Decreasing lesion size	Better outcome [9]
Thrombosed isolated pouch	Often favorable course [6]
Brain injury or patent feeders	Worse outcome [3]
Ventriculomegaly or parenchymal lesions	Poor prognosis [17]
Cardiac overload or venous hypertension	Higher risk, often prompts intervention [15]

4.4 Management Implications

The management of these cases depends on pathophysiology rather than the size of the lesions. [3,11]

Follow up conservatively reported for isolated, regressed and nonarterial lesions, whereas endovascular treatment was useful in worsening hydrocephalus, cardiac overload venous shunting or congestion, as published in literature. [9,11-13]

Embolization of the shunts are main reported strategy with improvement of the venous hypertension, and cardiac failure, more recently venous remodeling in some cases. [3,12,17-18]

Conversely, surgical management has been demonstrating limited effect in some cases of neonatal ectasia. The option of medical treatment as anticoagulation remains selective and controversial, as well as newer small series support medical treatment which lead to reduction of the venous pouch and residual shunt after embolization. [2-3,18]

Eventually, our case report illustrating the alarming imaging appearance of these entities with highly possible different natural history. The giant neonatal dural ectasia, outcome depends mostly on associated shunting, venous outflow compromise and brain consequence injury rather than the presence of venous pouch alone. [3,4,9,12]. However, long term neurodevelopmental outcomes remain poorly defined. Extended follow up and further research are recommended to improve clinical management and family counseling.

5. CONCLUSIONS

Neonatal giant dural sinus ectasia with dural arteriovenous fistulas can lead to rapid multiorgan failure and severe brain injury despite urgent endovascular treatment. Complex arterialized malformations remain high risk, life-threatening entities. Prognostic stratification is dictated by shunt severity and parenchymal damage regardless of the lesion size. Early multimodal neuroimaging guiding timely intervention and family counselling.

6. DECLARATIONS

Ethics approval and consent to participate: Institutional review board approval was obtained/waived according to institutional policy, King Fahad City, Riyadh, Saudi Arabia.

7. CONSENT FOR PUBLICATION

Not applicable.

8. AVAILABILITY OF SUPPORTING DATA

The data used during the current study are not publicly available due to patient privacy but are available from the corresponding author upon reasonable request.

9. COMPETING INTEREST

The authors declare that they have no conflict of interest.

10. FUNDING

This research received no specific grant from any funding agency.

11. AUTHORS' CONTRIBUTIONS

Dr.Salha acted as the person who conducted this study. Dr.Salha and Dr.Abdullah was involved in the literature review, methodology and start writing of this manuscript, following the patient data and writing the introduction and discussion sections. Dr.Ahmed and Dr.Ibrahim were involved in the literature search, conception, study design, and acquisition, as well as in developing research idea, as well as reviewing the manuscript.

12. ACKNOWLEDGEMENTS

Not Applicable.

13. DECLARATION OF GENERATIVE AI

During the preparation of this work the authors used GPT-4.1 (OpenAI, San Francisco, USA) to improve the readability of some of the sections, assure homogeneity of the passages and reduce redundancies in writing. After using this tool/service, the authors reviewed and edited the content as needed and take full responsibility for the content of the publication.

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