

Vein Of Galen Malformation

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Citation

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Abstract

Vein of Galen malformation is an uncommon intra-cranial anomaly accounting for 1-2% of all intra-cranial vascular malformations and up to 30% of pediatric vascular malformations. It results from arteriovenous connections between deep choroidal arteries and the median prosencephalic vein of Markowski (MPW). It is the most common extra cardiac cause of high output failure in newborn. Presenting here a case where a term female baby presented with high output failure. On Neurosonogram imaging findings suggestive of Vein of Galen Malformation was noted.

INTRODUCTION

Vein of Galen malformations, better termed as a median Prosencephalic vein malformation, is an uncommon intracranial anomaly accounting for 1-2% of intracranial vascular abnormalities and up to 30% of all pediatric cerebral vascular malformations¹. Arteriovenous fistula between deep choroidal arteries and the median prosencephalic vein of Markowski (MPV) develops during 6-11 weeks of gestation, which prevents normal regression of MPV². It is the most common extra cardiac cause of high output cardiac failure in newborn.

CLINICAL PRESENTATION

A term female baby, with a normal APGAR score, birth weight 2500gm and head circumference 36cms, presented with history of improper feeding since day one. There was no history of breathlessness or cyanosis. On examination, pansystolic murmur was detected. ECHO showed presence of an atrial septal defect of moderate size, patent foramen Ovale with left to right shunt and features of high output cardiac failure. Baby was referred to our department for abdominal ultrasound and neurosonogram.

Ultrasound examination of abdomen was normal.

Neurosonogram revealed a well-defined anechoic rounded lesion in the brain almost in the midline located posterior to third ventricle and superior to cerebellum under splenium of corpus callosum.

On Colour Doppler imaging, there was complete colour fill of the lesion with turbulent flow, with multiple tortuous feeding arteries and draining veins. Spectral tracing showed

high velocity venous flow within the lesion. There was no hydrocephalus or intracranial hemorrhage.

Image (LtoR) 1&2

(Coronal and Midsagittal): The midline anechoic lesion posterior to third ventricle is seen on neurosonogram.

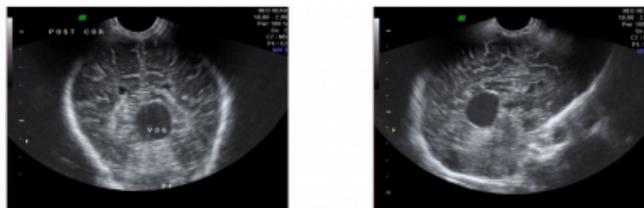


Image (LtoR) 3&4

Filling up of the anechoic lesion with multiple tortuous feeding arteries and draining veins on colour doppler.

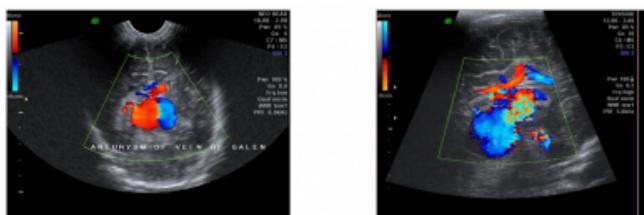
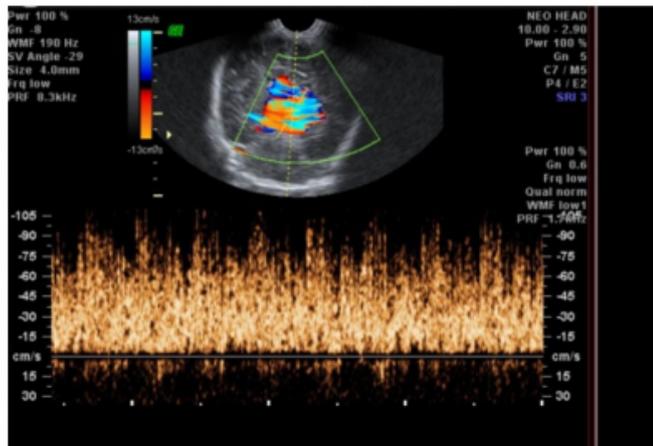


Image 5

Spectral tracing shows high velocity venous flow within the lesion.



DISCUSSION

The vein of Galen is a short vein with a large diameter. Arteriovenous fistula between deep choroidal arteries and the median prosencephalic vein of Markowski (MPV) develops during 6-11 weeks of gestation. Flow from the fistula prevents the normal regression of MPV. In neonates it presents as high output cardiac failure and a cranial bruit whereas in infants it presents with macrocephaly and developmental delays.

This condition can be detected prenatally by ultrasound or MRI. In neonates presenting with cardiac failure, a cardiac

echo should be done. MRI should be done to establish the vascular anatomy. Angiography remains the gold standard and will be done if the patient requires embolization.

The congestive heart failure should be treated first in neonates. The treatment of choice for VOGM is Embolization. Surgery should be considered only in cases where embolization is not successful as it has a higher mortality risk³.

CONCLUSION

This report emphasizes the role of Sonography for the diagnosis of vascular abnormalities, like Vein of Galen malformation in patients presenting with congestive cardiac failure and its role in antenatal detection, which aids in management and follow up.

References

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