Case Report

VEIN OF GALEN ANEURISMAL MALFORMATION: DIAGNOSIS CONFIRMED ON MRI AND CT ANGIOGRAPHY- A CASE REPORT OF 2.5 YEARS OLD CHILD AND REVIEW OF THE LITERATURE.

Saeed Mazher*, Junaid Ashraf*, Qazi Muhammad Zeeshan*, Farrukh Zulfiqar*, Shiraz Ahmed*

*Department of Neurosurgery, DUHS University of Health Sciences, Karachi.

ABSTRACT:

In this uncommon case report, we present a case of 2.5years old child with Vein of Galen Malformation who presented with history of headache used to relieve by taking analgesics. The purpose of this report is to increase the awareness about this challenging diagnostic dilemma demanding early recognition and decrease morbidity and mortality.

KEY WORDS: *Vein of Galen Aneurysm Malformation (VGAM). Magnetic Resonance Imaging (MRI). Headache*

INTRODUCTION:

Vein of Galen aneurysmal Malformation was first described by Steinhel in 1895[1] . It is one of the rare congenital anomalies of intracerebral circulation which constitutes about 1% of all intracerebral vascular malformations. However it represents a reported incidence of 30% in pediatric vascular malformations. [2,3]. In the literature it was found that Jaeger et al., first published a case report of VOGAM.

During early development the large median prosencephalic vein of Markowski, gradually involutes and eventually persist as the great vein of Galen[5]. Thus VGM is assumed to be the persistent structure of the embryonic median prosencephalic vien of Markowski. The VGAM develops during 6th to 11th weeks of gestation and can already be diagnosed by ultrasound. But often the VGAM detected in post natal period[6].

The two most widely used classification systems have been described by Yasargil and Lasjaunias. Yasargil classified in to four categories Table-1[7]. While Lasjaunias and colleagues classified in to choroidal and mural types depending on the location of fistula.

These patients usually present with varied symptoms depending on the age of the patient and on the anatomical angioarchitecture of malformation. In neonates there is choroidal type of VGAM, so multiple choroidal arteries emptying in to venous pouch leading to volume overloading is often dominating raised cardiac output, tachycardia, cardiomegaly and cardiac insufficiency, pulmonary hypertension even multiple organ failure. In infancy mural type of VGAM may present with bigger shunt as direct arteriovenous fistulas with in wall of the median prosencephalic vein of Markowski leading to hydrocephalus, macrocrania and developmental retardation. Rarely VGAM patient's presentation found to be epileptic seizures. In different part of the world the clinical picture can differ from the general description in the literature.[8]

Calcification is seen in about half the patients with thrombosed VGAM; compared to only 14% of patients without thrombosis Where as in contrast CT ,the presence of a central thrombus and peripheral circulating blood along the wall of the sac can produce Target sign[9] . Presence of calcification is one of the poor predictors for the tendency to thrombose subsequently. Calcification is rarely seen before 15 years and complete calcification of the sac is extremely rare[10].

We report a case of VGAM who presented with headache and CT demonstration of the presence of thrombus within the aneurysmal sac.

CASE REPORT:

A 2.5 years old child with normal birth and developmental history, presented to us due to complain of frequent headache for one year used to subside by analgesic. Prenatal and

Corresponding Author:
Saeed Mazher Dept. of Neurosurgery, DUHS Karachi Email: saeed.mazhar@hotmail.com

post natal history was uneventful. Other four siblings had no significant complain reported in the family history and having healthy lives. On clinical examination there was no abnormal finding which could be elicited during thorough neurological examination and systemic review.

His routine blood examination revealed heamoglobin level as 9.3g/dl with peripheral morphological picture of Hypochromic Anisocytosis. Moreover his inflammatory markers were slightly raised. Rest of the laboratory evaluation was found to be unremarkable, except the serum calcium was towards the lower side.

On CT scan brain there was a huge lesion with well circumscribed margins showing rim of calcifications. It was reported as more favouable towards hydatid cyst with consideration of a thrombosed VGAM in differentials Fig 1. As to confirm MRI brain was recommended which revealed the diagnosis of VGAM with hydrocephalus. On the basis of MRI brain, differential diagnosis of Hydatid cyst was excluded and a well defined non enhancing lesion measuring about 5 x 5 cm was appreciated in the left posteroinferior temporal region causing extrinsic compression over the occipital horn of the left lateral ventricle, midbrain, thalamus, cerebral aqueduct and quadrigeminal cistern Fig 2. His Echinococcus antibody titer was also performed which was reported as non significant.

To proceed further as per protocol we got his cerebral angiogram done which revealed a large well defined rounded multi-lamellated hypodense lesion showing multiple peripheral rim calcifications in the quadrigeminal cistern projecting to the left of the midline. In 3-D image reconstruction, its size was documented as 5.6x5.3cm and extends 5.7cm in craniocaudal direction. However MD CTcerebral angiography showed in Fig 3 reported as thrombosed aneurysm of Vein of Galen with obstructive hydrocephalus.

Due to the headache initially it was considerable to perform intervention but surgery can worsen the situation therefore it was postponed and endovascular embolization was planned.

As the patient is not having neurological deficit or cardiac problem hence he is in our close follow up and the patient has been allotted an appointment with neuroendovascular interventionist. It was recommended to wait for few months for endovascular treatment to be instituted.

Table 1:

YASARGIL CLASSIFICATION OF VEIN OF GALEN MALFORMATIONS
Pure cisternal fistula between pericallosal arteries, posterior cerebral artery and vein of Galen.
Fistulous connections between the thalamoperforators and vein of Galen.
Mixed form with both Type1 and Type2 lesions.
Plexiform AVM with one or more niduses within the mesencephalon or thalamus with draining veins emptying in to the vein of Galen.

Fig 1:
CT scan brain plain and contrast showing rim of calcification in the lesion.

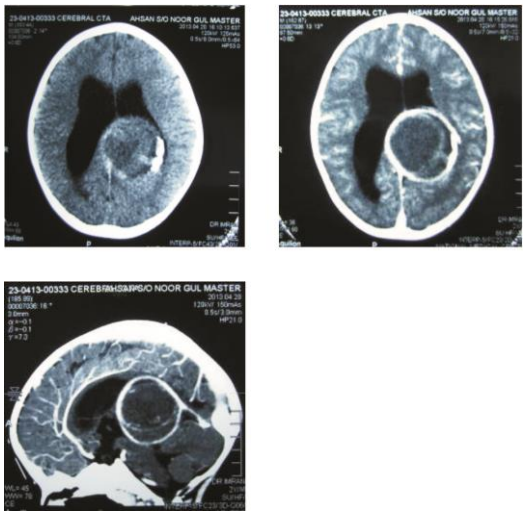


Fig 2:
MRI brain contrast & T2W images showing lesion location with extension.

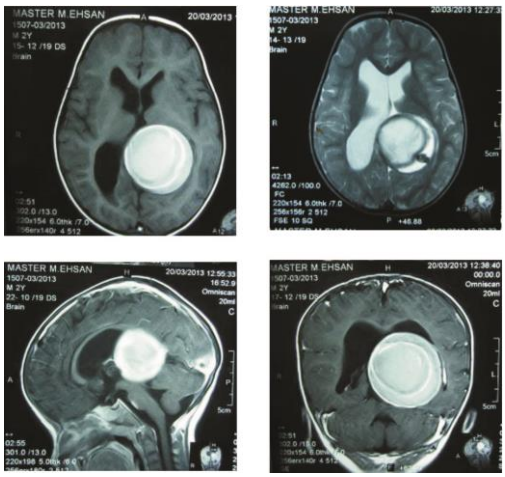
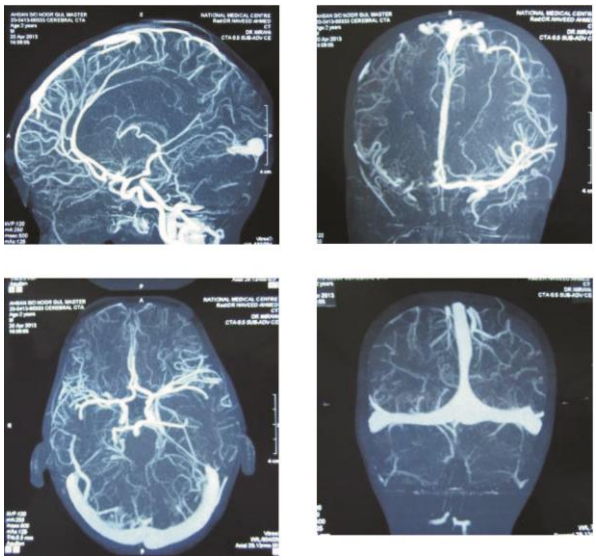


Fig 3:
MD CT cerebral angiogram showing VGAM



DISCUSSION:

The reported mortality rate in neonates is higher than 90%, most of the patients die within first week of diagnosis or treatment[11]. On literature review it was documented that in untreated patients the mortality among newborns with cardiac insufficiency accounts to 100%, while in infancy it's 72%[12].

According to literature review hydrocephalus reported in 46.8% patients of VGAM, and increased venous sinus pressure has been considered of primary importance in the development of hydrocephalus in these patients[13]. There is huge pressure difference in the dural venous sinus and the CSF intraventricular pressure before embolization, while post embolization there is remarkable fall in the pressure of dural venous sinuses[14]. Hence it has also been reported that ventricular shunting may worsen the cerebral venous hypertension, before resolving the arteriovenous shunt as it is not tolerable to infants and therefore should be preceded by emergency embolization[13, 16]. Due to this reason we avoided the shunting in our case.

The safest treatment modality for a VGAM and also therapy of choice is endovascular embolization. Using transarterial route with special glue is better than tranvenous embolization or surgical treatment[15, 16]. Timing and method of endovascular embolization depends on the clinical sign and symptoms of the patient[17]. Tranvenous approach mentioned in literature but it was associated with increased risk of bleeding complications[15].

The Bicetre-score is one of the important tools to assess the therapeutic management of VGAM child. The basis of this score is defined by Lasjaunias who has published his experience with more than 300 patients of VGAM. While using this scoring system one can evaluate the cardiac, cerebral, respiratory, hepatic and neurological state of the patient. If cardiovascular and neurological symptoms are stable (Bicetre-score>12), the treatment can be postponed up to the age of 5 to 6 month[18, 19]. With a Bicetre score of 8-12 points, an emergency endovascular embolization should be performed. In case of profound neurological deficit or medically uncontrolled cardiac insufficiency (Bicetre score <8) there is no indication for an invasive procedure[15]. It is well documented that pre-existing brain damage characterized by atrophy and parenchymal calcifications indicated a poor prognosis with bad outcome[20].Most of the research work portrayed that VGAM patients usually respond well to endovascular treatment and simultaneously reported good cognitive and functional outcome[21].

Patients with VGAM presenting symptoms during the neonatal period have a worse prognosis regarding the success of endovascular embolization[22]

CONCLUSION:

However patients usually presents with poor Bicetre score at the time diagnosis which demands early treatment. In our case the diagnosis was established by MRI brain and MD CT cerebral angiogram with Bicetre score >12. Therefore it is recommended to establish close watch full follow up by the help of multidisciplinary approach, so that it can be treated by endovascular method before onset of symptoms without complications.

ACKNOWLEDGMENT:

The author would like to express his sincere gratitude to Professor Junaid Ashraf for criticism, encouragement and support.

REFERENCES:

1. Dandy WE. Experimental hydrocephalus. Annals Surgery. 1919; 70(2):129-142.
2. Long DM, Seljeskog EL, Chou SN et al Gaint arteriovenous malformations of infancy and childhood. J Neurosurg. 1974;40:304-12
3. Gailloud P, ORiordan DP Burger et al. Diagnosis and management of vein of galen aneurysmal malformations. J Perinatol. 2005;25:542-51
4. Jaeger JR, Forbes RP, Dandy WE. Bilateral congenital cerebral arteriovenous communication aneurysm. Trans Am Neurol Assoc. 1937;63:173-6
5. Raybaud CA, Strother CM, Hald JK et al. Aneurysms of the vein of Galen: embryonic considerations and anatomical features relating to the pathogenesis of the malformation. Neuroradiology. 1989;31:109-128
6. Cumming GR. Circulation in neonates with intracranial arteriovenous fistula and cardiac failure. American Journal of Cardiology. 1980; 45(5):1019-1024.
7. Yasargil MG. Microneurosurgery IIIB New York: Thieme Medical Publishers; 1988. Pp.323-57
8. Gupta AK, Rao VRK, Varma DR, et al. Evaluation, management and long term follow up of vein of Galen malformation. Journal of Neurosurgery. 2006; 105(1):26-33.
9. Nikas DC, Proetor MR, Seott RM. Spontaneous thrombosis of vein of Galen aneurysmal malformation. Paediatr Neurosurg 1999;31:33-9.
10. Chapman S, Hockley AD. Calcification of aneurysm of the vein of Galen. Pediatr Radiol 1989;19: 541-2
11. Johnston. IH, Whittle. IR, Besser M et al. Vein of Galen malformation: diagnosis and management. Neurosurgery. 20, 747-58.
12. Khullar D, Andeejani AMI, Bulsara KR. Evolution of treatment options for vein of Galen malformations. Journal of Neurosurgery. 2010;6 (5):444-451.
13. Zerah M, Garcia-Monaco, Rodesch G et al. Hydrodynamics in vein of Galen malformations. Child's Nerv Syst. 1992;8:111-7.
14. Mickle JP, Quisling RG. The transtorcular embolization of vein of Galen aneurysms. J Neurosurg 1986;64:731-5.
15. Lasjaunias PL, Chng SM, Sachet M et al. The management of vein of Galen aneurysmal malformations. Neurosurgery. 2006;59 (5):184-194.
16. Lasjaunias P, Garcia-Monaco R, Rodesch G et al. Vein of Galen malformation. Endovascular management of 43 cases. Chils's nervous System. 1991;7 (7):360-367.
17. Beucher G, Fossey C, Belloy F et al. Antenatal diagnosis and management of vein of Galen aneurysm: review illustrated by a case report. Journal Gynecologie Obstetrisue et Biologie de la Reproduction. 2005;34 (6):613-619.
18. Mitchell PJ, Rosenfeld JV, Dargaville P et al. Endovascular management of vein of Galen aneurysmal malformations presenting in the neonatal period. American Journal of Neuroradiology. 2001; 22(7):1403-1409
19. Alvarez H, Monaco RG, Rodesch G et al. Vein of Galen aneurysmal malformations. Neuroimaging Clinics of Noth America. 2007;17 (2):189-206.
20. Frawley GP, Dargaville PA, Mitchell PJ et al. Clinical course and medical management of neonates with severe cardiac failure related to vein of Galen malformation. Arch Dis Child Fetal Neonatal Ed. 2002;87:144-9
21. Ellis JA, Orr L, Ii PC et al. Cognitive and functional status after vein of Galen malformation endovascular occlusion. World Journal of Radiology. 2012;4(3):83-88
22. Jones BV, Ball WS, Tomsick TA et al. Vein of Galen aneurysmal malformation: diagnosis and treatment of 13 children with extended clinical follow-up. American Journal of Neuroradiology 2002;23 (10):1717-1724.

Submitted for publication: 26-11-2013

Accepted for publication: 20-05-2014