



Interesting Case of Facial Palsy

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Abstract

Vein of Galen malformations are rare anomalies that constitute only 1% of all intracranial malformations.

35 Year Old Female Presented with Deviation of angle of mouth to the right side associated with giddiness. MRI Brain with MRV done, Showed vein of galen malformation (median prosencephalic AV fistula) causing mass effect and obstructive hydrocephalus. Because of its rarity in adults, there is still insufficient information about this disease during adult age.

Introduction

Vein of Galen malformations are rare anomalies that constitute only 1% of all intracranial malformations. However they represent 30 % in pediatric age group, to understand its valid clinical presentations, knowledge has to date back to its embryology. During the third phase of intrinsic vascularization, the median prosencephalic vein has to regress, failure of which will result in aneurismal malformation. With new and increasing techniques, it has been increasingly diagnosed in prenatal period. In adults the presentations can vary depending on its mass effects. It almost never bleeds, continuing developments in diagnostic and interventional aspects have radically changed the management of these cases.

Case History

35 years old female who was a known Hypothyroid (on T. Thyroxine 100mcg) came with chief

complaints of Deviation of mouth to right side, a/w Facial Pain and giddiness for 4 days, Watering of Left eye for 4 days.

There was no history of Tinnitus/ Hearing difficulty/ Weakness of limbs/ Diminution of Vision/ diplopia.

On Examination, Conscious, Oriented, Afebrile

PR : 80 bpm

BP : 140/90 mmhg

RR : 18/minute

No pallor, icterus, cyanosis, clubbing, lymphadenopathy, oedema.

CVS : S1S2 +

Respiratory System: B/L A/E + ; NVBS

PA : Soft, Bowel Sounds +

CNS: Power on B/L UL and LL – Normal

Bulk: B/L upper and lower limbs : Normal

TONE – B/L UL and LL- NORMAL,

Deep Tendon Reflexes: B/L Normal

Sensory System – Normal

Cranial Nerve - 7

Loss of forehead wrinkling on the left side
 Deviation of angle of mouth to right side
 Improper closure of left eyelid with Bell's Phenomenon

Other Cranial Nerves Normal

Cerebellum

Tandem Walking Swaying to Left Side
 finger finger , finger-nose incoordination present
 No Autonomic Disturbances

- A working diagnosis of left Bell's Palsy With Left Cerebellar involvement was made, probably due to CP angle tumor.

- She was started on T. Valcyclovir 1gm TDS, T. Prednisolone 40mg OD, T. Thyronorm 100mcg OD

- ENT and Ophthalmology opinion was obtained and they labelled her as Bell's Palsy with Lagophthalmos, advised to continue antiviral and steroids

- MRI Brain with MRV done , showed Vein of Galen Malformation (Median Prosencephalic Av Fistula) Causing Mass Effect And Obstructive Hydrocephalus.)

- Neuroradiology opinion was obtained and was advised for diagnostic cerebral angiogram which revealed single whole high flow fistula with aneurysmal fistula with aneurysmal dilatation of vein of galen.

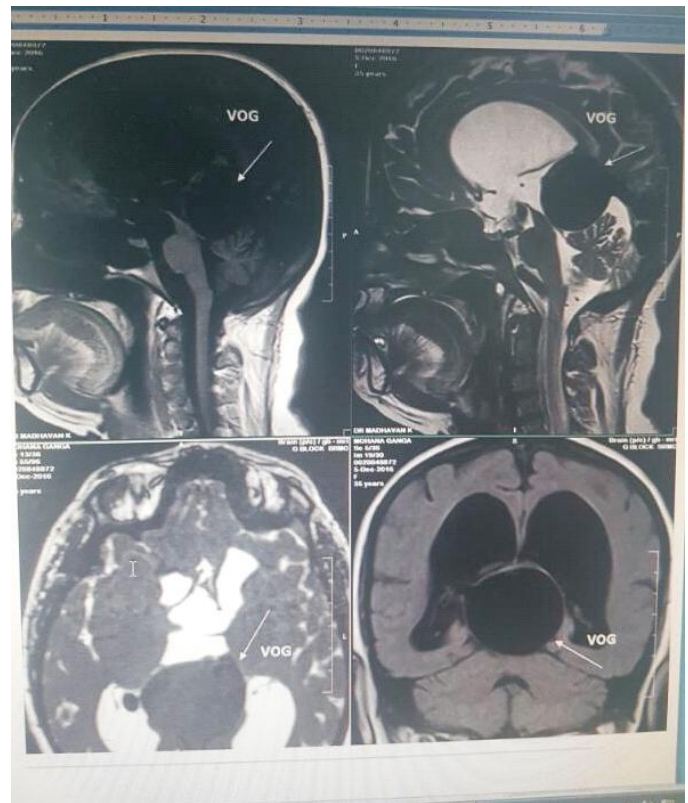
- Interventional Radiologist advised for Glue Embolization.

- Patient counseled glue embolisation, but patient wanted a second opinion . Patient was discharged at request with following drug advise

T. Levipil 500mg BD

T. Thyroxine 100 mcg OD

T. prednisolone was tapered and stopped.



Discussion

- Vein of Galen aneurysmal malformation (VGAM) was first described by Steinhel in 1895. incidence is 1 in 25000. Most often the VGAM is detected in the postnatal period.

VOG is a Congenital malformation develops during weeks 6-11 of fetal development as a persistent embryonic prosencephalic vein of Markowski Vein of Gallen – Internal Cerebral Vein and Basal Vein of Rosenthal.

Vein of Galen malformation has been associated with capillary malformation- autosomal dominant disorder, caused by mutations in the *RASA1* gene known as Parkes Weber syndrome.

Angiographic identification of the two major types of the abnormality aneurysmal malformation aneurysmal dilatation.

- Aneurysmal malformation does not drain the normal brain tissue nor the normal venous system. Alternate drainage pathways develop by the 12th week of gestation and persist into fetal life.



Clinical Features

• Infants

Up to 25% high-output congestive cardiac failure,
Older children – macrocephaly or hydrocephalus.

• Adults

headache

seizures.

neurological deficits

epistaxis,

proptosis.

Diagnosis

• MRI can demonstrate the location of fistula, presence of any nidus, the arterial components, the venous sac as well as the status of venous drainage. Thrombosis of the venous sac is also depicted well.

• Angiography remains the gold standard for the evaluation of VOGMs.

Medical Management

- Anti-seizure medications to manage convulsions
- Cardiac management of congestive heart failure

Management

• Lasjaunias and co-workers, have described a 21-point scale based on cardiac function, cerebral function, hepatic function, respiratory function and renal function. A score of less than 8 usually indicates a poor prognosis– no emergency treatment
A score of 8–12 - indication for emergency endovascular management.

A score of > 12 - delay the endovascular procedure, by medical management

Embolization

- specially designed coils,
- glues
- spheres, which plug its vessels.

A shunt to manage hydrocephalus

Conclusion

Because of its rarity in adults, there is still insufficient information about this disease during adult age.

Use of oral contraceptives postpartum status, sickle cell anemia, and aseptic meningitis were risk factors related to thrombosis of the vein of Galen.

References

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