Interesting Case of Facial Palsy

Authors
Sowmiya .M¹, Suja², Madhavan K³, Dr Priya Dharshini⁴, Sivaprakash⁵
¹Final Year Postgraduate, Department of General Medicine, ²Assistant Professor, General Medicine
³Professor in General Medicine, ⁴Assistant Professor, General Medicine
⁵Associate Professor, General Medicine

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 pros encephalic 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. Valacyclovir 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

1. Vein of Galen malformations: Review A. K. Gupta, D. R. Varma Department of Radiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvendram


3. Diagnosis and Management of Vein of Galen Aneurysmal Malformations, Philippe Gailloud MD1, Declan P O'Riordan MD2, Ingrid Burger BS1, Olivier Levrier MD1, George Jallo MD3, Rafael J Tamargo MD4, Kieran J Murphy MD1 and Christoph.

4. U Lehmann MD2 Vein of Galen aneurysmal malformations: critical analysis of the
literature with proposal of a new classification system A review Martin

5. M. Mortazavi, M.D.,1 Christoph J. Griessenauer, M.D.,1 Paul Foreman, M.D.,1 Reza Bavarsad Shahripour, M.D.,2 Mohammadali M. Shoja, M.D.,2 Curtis J. Rozzelle, M.D.,2 R. Shane Tubbs, Ph.D.,2 Winfield Stitt Fisher III, M.D.,1 and Takanori Fukushima, M.D.3 1

6. Division of Neurosurgery, Department of Surgery, University of Alabama at Birmingham; 2 Pediatric Neurosurgery, Children’s Hospital, Birmingham, Alabama; and 3 Division of Neurological Surgery, Dula University, Durham, North Carolina