QUIZ

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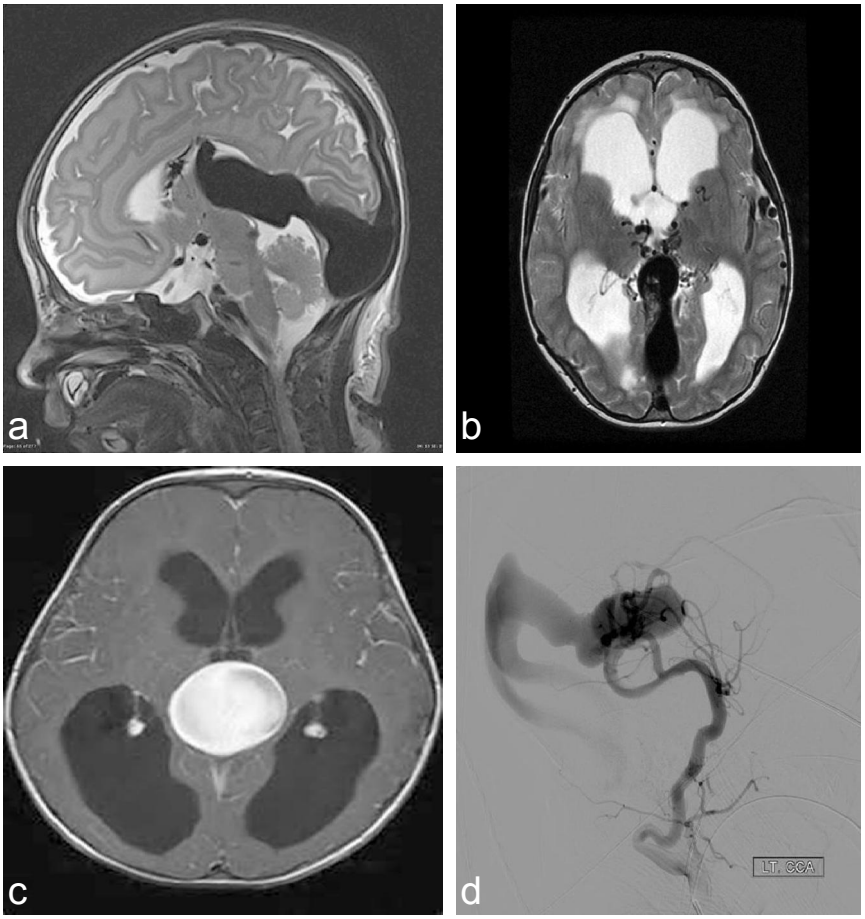
Clinical History

Infant presenting with increase head size



Question

Q1. What are the findings?
Q2. What is the diagnosis?
Q3. What is the gold standard test for diagnosis?
Q4. What are the treatment options?

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QUIZ

Answer

1. (a) Sagittal (b) Axial T2WI showing a hypointense (flow void) area within posterior fossa at the midline draining into Vein of Galen representing aneurysmal dilatation of the feeding and draining vessels. This is associated with dilatation of ventricular system representing hydrocephalus. (c) Axial T1 Post contrast T1WI showing enhancement of the vessels. (d) Venogram showing arteriovenous malformation with feeding vessel and draining vein.

2. Vein of Galen Aneurysmal Malformation.

3. Angiography.

4. Open surgery, Endovascular treatment and Stereotactic radiosurgery. VP Shunt if associated with hydrocephalus.

Discussion

Vein of Galen malformations (VGMs) are rare congenital defects of cerebral vessels. They constitute up to 30% of intracranial vascular malformations presenting among pediatric patients. They are formed between the 6th and 11th week of gestation. The malformation is due to the presence of one or more artero-venous fistulas directing blood flow toward the dilated, persistent proximal median prosencephalic vein. Clinical symptoms depend on the size of arterovenous flow. The most common ones include: antenatal development of high output heart failure, brain hypoperfusion and hydrocephalus.

Ultrasound is a basic examination method allowing for diagnosis of VGMs. MRI technique has recently become a clinically valuable complementary method and is highly valued allowing for precise determination of the size of ventricular system, presence of elevated intraventricular pressure, topographic relationships between pathological vessels and particular brain structures as well as the presence of infarcted and ischemic areas.[1]

Angiography remains the gold standard in full characterisation of the lesion. It enables to individually catheterise feeding vessels. Venous drainage is via the median prosencephalic vein (MPV), the straight sinus (if present) and then out via the transverse/sigmoid sinuses. By definition there should be no drainage to other components of the deep venous system.[2]

Recent advances in the field of intervention neuroradiology has changed the treatment and prognosis of children with VOGAM. Therapeutic options available include no treatment, open surgery, endovascular treatment, and stereotactic radiosurgery. The therapeutic options should be individualized with consideration of age, clinical manifestation of the lesion and the angioarchitecture. Endovascular treatment has improved the results of treatment in recent years. The transarterial approach is preferred and more effective in controlling the symptoms.[3]

Prognosis in VGMs depends on two main factors. The first one is the severity of heart failure, which is directly related to the size of arterovenous shunt, and the second is the extent of cerebral ischemia caused by increased venous pressure and so-called cerebral steal.[4]

References

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