CASE REPORT

Antenatal diagnosis of aneurysmal malformation of the vein of Galen

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Accepted 20 November 2015

▸ http://dx.doi.org/10.1136/bcr-2012-006529

SUMMARY

Vein of Galen malformation (VGAM) results from an aneurysmal aberration with an arteriovenous shunting of blood. Vein of Galen aneurysmal malformations are the most frequent arteriovenous malformations in infants and fetuses. The congenital malformation develops during weeks 6–11 of fetal development. Infants often die from high-output congestive heart failure.

BACKGROUND

This case is important because vein of Galen malformation (VGAM) is a rare congenital anomaly that often increases in size with advancing age and is associated with hydrocephalus, seizures, intracranial haemorrhage, heart failure and even resultant death. So early diagnosis and treatment is important in order to avoid such consequences.

CASE PRESENTATION

A 25-year-old woman with an 8-month history of amenorrhoea presented to hospital for routine antenatal check-up and was advised sonography for fetal wellbeing. She was a second gravida with one live child and no known previous congenital malformations.

INVESTIGATIONS

Ultrasonography examination revealed that fetal maturity was 31 weeks 5 days. All fetal biometric parameters were correlating with the mother’s gestational age according to her last menstrual period. We identified a 24×15 mm solitary, oval cystic lesion in the midline in the cranium extending into the right paramedian region of the posterior fossa. The lesion appeared to be separate from the occipital horns of the lateral ventricles. Both cerebella were normal. Ventricles with choroid plexuses were normal. No other abnormalities were found in the cranium. On colour Doppler, the lesion showed characteristic bidirectional blood flow (figure 1). No other gross congenital anomalies were detected. The fetus was found to have mild cardiomegaly with situs solitus. Sonographic evaluation of the fetal heart did not reveal any other abnormality to justify the cause of the cardiomegaly.

Diagnosis of aneurysmal malformation of vein of Galen was made based on sonographic features and on the mild cardiomegaly.

OUTCOME AND FOLLOW-UP

The patient was delivered at full term through vaginal route. Physical examination of the baby was unremarkable. The baby was called for post-natal evaluation at the radiology department on the second day. On cranial sonography, a 32×33 mm cystic lesion was found in the midline and right paramedian region in the posterior fossa showing bidirectional blood flow correlating with antenatal findings (figure 2). No other abnormalities were found in the cranium. Echocardiography showed mild cardiomegaly with otherwise normal morphology, signifying high output as the cause of the cardiomegaly.

Antenatal diagnosis of VGAM was confirmed. On discharge from hospital on the fifth post-partum day, we explained the need for CT angiography and other interventions to the relatives of

![Figure 1](url) Antenatal ultrasonography image showing cystic lesion in posterior fossa that, on Doppler, showed bidirectional colour flow—vein of Galen malformation.
the patient. For these, the patient was referred to a specialist
centre. Unfortunately, the patient was lost to follow-up.

**DISCUSSION**

The vein of Galen is located under the cerebral hemispheres
and drains the anterior and central regions of the brain. VGAM
results from an aneurysmal aberration with arteriovenous shunt-
ing of blood due to either thalamic arteriovenous malformation
or a choroidal arteriovenous fistula.

Vein of Galen aneurysmal malformations are the most fre-
quent arteriovenous malformations in infants and fetuses.1 The
malformation increases greatly in size with age, although the
mechanism of the increase is unknown.2 The right side cardiac
chambers and pulmonary arteries also develop mild to severe
dilation.3 The congenital malformation develops during weeks
6–11 of fetal development.

**Frequency:** unknown

**Race:** VGAM occurs in all races

**Sex:** Boys and girls are affected equally

**Age:** VGAM is a congenital malformation; therefore, it may
present at birth or in early childhood

It commonly presents in the neonatal period, although it may
present in early childhood. Malformations often lead to cardiac
failure, cranial bruits and hydrocephaly, and subarachnoid haem-
orrhage and seizures in neonates.4 Heart failure is due to the
size of the arteriovenous shunt, which can steal 80% or more of
cardiac output, with large volumes of blood under high pressure
returning to the right heart and pulmonary circulation, and
sinus venosus atrial septal defects.3 4 It is also the most common
cause of death in such patients.5

**Imaging studies**

**Cranial ultrasound**

▸ This will help to localise or identify the lesion.
▸ Doppler studies can help further to understand the haemo-
dynamics of the lesion.

**Cranial MRI and/or CT scan**

▸ These studies will help confirm diagnosis and define the
degree of involvement. Imaging studies will also help deter-
mine any accompanying hydrocephalus. Cerebral MRI shows
large flow void in the central region with enlarged straight
sinus and drainage to transverse sinuses.
▸ MR angiography can help to delineate vascular supply. MRI
venogram shows VGAM with draining veins.

**Cranial angiography:** Cranial angiography is required to
define the extent of aneurysmal dilation and details for arterial
feeders. Venogram shows draining vasculature for VGAM.

**Cardiac ultrasound:** This study may be indicated to assess left
ventricular function.

**Medication summary**

▸ No specific medical therapy is available
▸ Seizures usually are managed with antiepileptics.6

**Patient care**

▸ Infants who undergo surgical ligation or selective embolisa-
tion should be reimaged to assess results of the therapeutic
intervention
▸ Head circumference measurements should be obtained in
patients who undergo ventriculoperitoneal shunt to treat
hydrocephalus. The patient should be monitored for the
development of hydrocephalus.

**Complications**

Complications usually associated with VGAMs are usually intra-
cranial haemorrhages.7 Over half the patients with VGAM have
a malformation that cannot be corrected. Patients frequently die
in the neonatal period or in early infancy. Studies have shown
that 77% of untreated cases result in mortality.8 Even after sur-
gical treatment, the mortality rate remains as high as 39.4%.8

**Learning points**

▸ Vein of Galen malformation is an uncommon congenital
malformation that should be kept in consideration for
diagnosis if imaging features are suggestive.
▸ Vein of Galen malformation may be mistaken for a cystic
lesion of the brain if no colour Doppler is performed. Colour
Doppler helps to differentiate cystic lesions from vascular
aneurysms.
▸ Vein of Galen malformations are associated with
hydrocephalus, seizures, intracranial haemorrhage, heart
failure and death. So early diagnosis and treatment is
important in order to avoid such consequences.

**Competing interests** None declared.
REFERENCES