Computed Tomographic Diagnosis of Aneurysmal Dilatation of the Great Vein of Galen in a Male Infant

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ABSTRACT

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A case of great vein of Galen aneurysmal dilatation diagnosed on cranial CT scan in a male infant has been presented. The clinical, radiological features as well as the prognostic factors were also discussed.

Keywords: Aneurysmal dilatation, Computed tomography, great vein of Galen, infant,

INTRODUCTION

The vein of Galen aneurysm, are distinct rare congenital, aneurysmal venous anomalies which occur mainly in the paediatric age group. The incidence is about 1:25000 deliveries, accounting for 1% of all intracranial malformations, however 30% of this anomaly occur in the paediatric age group (Golombek et al., 2004; Recinos et al., 2012). This anomaly is of two clinical findings, the vein of Galen aneurysmal malformation (VGAM) and the vein of Galen aneurysmal dilatation (VGAD). For the vein of Galen aneurysmal dilatation (VGAD), there is venous drainage into a dilated embryologically normal great vein of Galen. Whereas the vein of Galen aneurysmal malformation (VGAM) is an embryological remnant of the median vein of the prosencephalon, there are two variants. The mural type usually associated with single arteriovenous fistula and the choroidal type, which is associated with multiple bilateral arteriovenous fistulas.

Aneurysmal malformations of the vein of Galen typically result in high-output congestive heart failure or may present with developmental delay, hydrocephalus and seizures.

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This is a case of a male infant with computed tomographic scan diagnosis of aneurysmal dilatation of the great vein of Galen. It is presented to highlight the role of intravenous contrast computed topographic evaluation in the diagnosis of this condition.

CASE REPORT

E.D., an 11-month-old male infant presented at the children's emergency department of the University of Benin Teaching Hospital on the 5th of February 2014 with a history of enlarged head of about 7

months duration, failure to thrive and difficulty in breathing of 2 days duration. Pregnancy was uneventful though his mother did not have an obstetric scan. He was delivered by spontaneous vaginal delivery without any complications. About 4 months after delivery, the mother noticed that baby's head was progressively getting enlarged.

There was no history of fever, however the child developed recent poor tolerance to feeding and two days prior to presentation he started having difficulty in breathing. Developmental milestones were delayed as he achieved neck control at 5 months, sat at 8 months, and started crawling at 9 months.

On examination, he was dyspnoeic, respiratory rate was 53 cycles/minute, but breath sounds were vesicular. Pulse rate was 102 beats/minute. The child had a large head, was afebrile, not pale and anicteric. Other systems were essentially normal. Packed cell volume was 33%. Random blood sugar, electrolyte, urea and creatinine were within normal levels.

Transfontanelle sonography showed dilated lateral and third ventricles with reduction of the cerebral mantle (Figure 1). Chest radiograph showed cardiomegaly with bilateral hilar fullness (Figure 2). Cranial computed tomogram scan also showed dilated lateral and third ventricles with reduced cerebral cortical mantle. The fourth ventricle was however normal, indicative of a non-communicating hydrocephalus (Figure 3a, 3b and 3c). On axial view, there was a well-defined, fairly large, rounded hyperdense mass in the midline measuring (5.00cm x8.00cm), which avidly enhanced on contrast administration (Figure 3a and 3b). The mass was continuous with the sagittal sinus on sagittal reconstructed image (Figure 3c). A diagnosis of aneurysmal dilatation of the great vein of Galen complicated by cardiac failure was made. He subsequently was managed for cardiac failure. He had a ventriculoperitoneal shunt to relieve the hydrocephalus and was placed on antibiotics. He is presently being managed in the clinic.

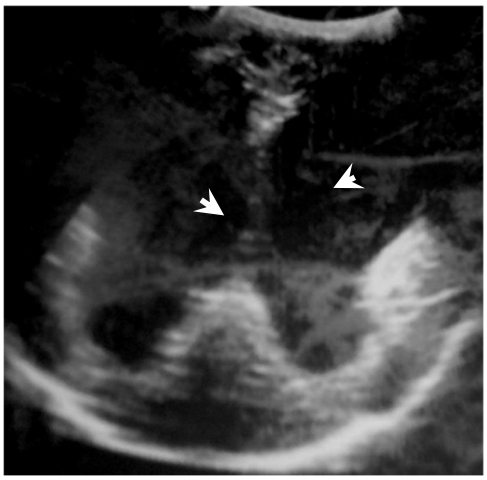

Figure 1: Transfontanelle sonogram: There is dilatation of the lateral and third ventricles (arrows) with reduction in the cerebral cortical mantle.

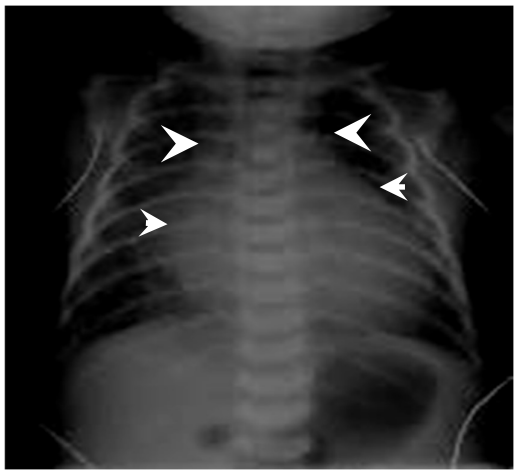

Figure 2: Chest radiograph: There is cardiomegaly (arrows) with bilateral hilar congestion (arrow heads).

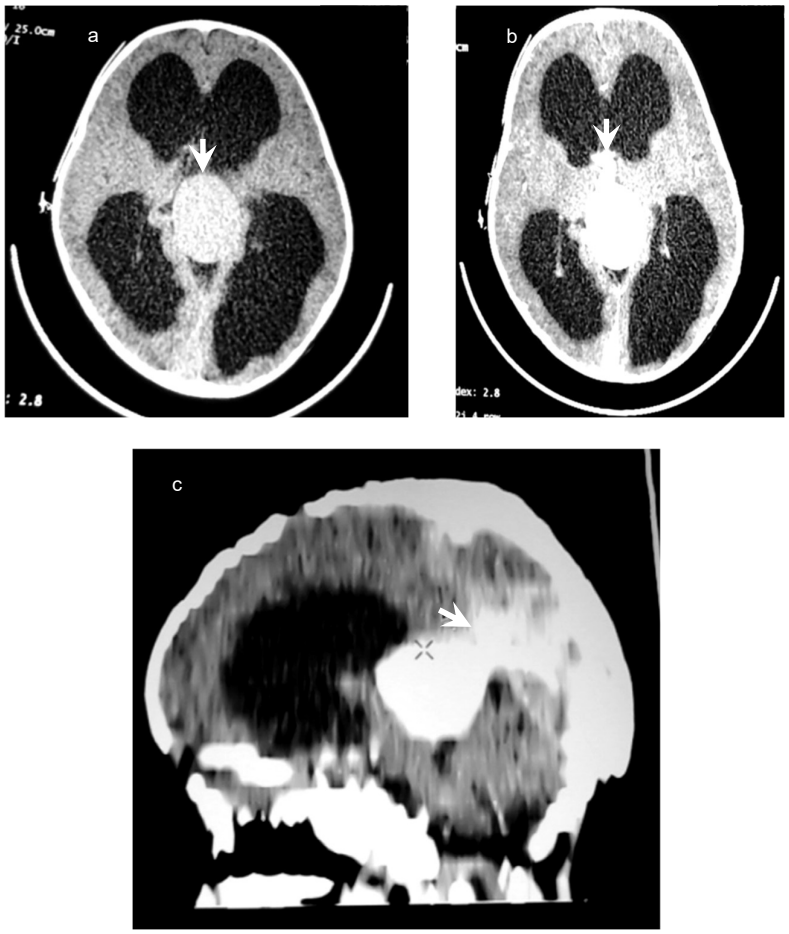

Figure 3: (a) Non contrast enhanced axial cranial computed tomography scan: There is a well-defined, fairly round hyperdense mass in the midline (arrow). measuring (5.00cm x 8.00cm). There is also dilatation of the lateral and third ventricles with reduction in the cerebral mantle. The fourth ventricle was normal(b) Contrast enhanced axial cranial computed tomography scan: The mass is seen to enhance avidly (arrow). (c) Sagittal reconstructed cranial computed tomography scan (contrast enhanced): The mass is seen to be continuous with the sagittal sinus (arrow).

DISCUSSION

The vein of Galen aneurysmal malformation is a congenital vascular malformation that comprises about 30% of paediatric vascular and 1% of all congenital anomalies (Recinos et al., 2012). Vein of Galen malformation results from an aneurysmal malformation with arteriovenous shunting of blood. The congenital malformation develops during the 6^{th}-11^{th} week of fetal development as a persistent embryonic prosencephalic vein of Markowski, thus vein of Galen malformation is actually a misnomer. The vein of Markowski actually drains into the vein of Galen (Incorpora et al., 1999).

During the perinatal period, a timely diagnosis is of paramount importance because the large systemic shunting within the fetal brain often results in a substantial steal of blood, potentially leading to cardiac failure, hydrops and perinatal death (Comstock & Kirk, 1991). Unfortunately, the diagnosis in this patient was made postnatally after the patient had developed cardiac failure.

Vein of Galen malformation usually causes high-output heart failure in the newborn resulting from the decreased resistance and high blood flow in the lesion as was seen in the case presented. Patients with this condition can also present with stroke or steal phenomena that result in progressive hemiparesis. The patient did not present with any central nervous system symptoms Spontaneous intracranial haemorrhage is a rare, complication in this condition.

The malformation may also result in mass effect causing progressive neurological impairment. Alternatively, the malformation may cause obstruction of cerebrospinal fluid outflow resulting in hydrocephalus as was demonstrated on both Transfontanelle scan and cranial CT scan in this

patient (Incorpora et al., 1999). Vein of Galen malformation has been associated with capillary malformation-arteriovenous malformation (CM-AVM), which is a newly recognized autosomal dominant disorder caused by mutations in the RASA1 gene in 6 families (Revencu et al., 2008).

With the widespread use of sonography in antenatal care, most cases of vein of Galen aneurysm are currently detected in utero. This case however was not detected in utero as patient's mother did not have an obstetric scan during pregnancy. The most striking prenatal feature is the detection of a cerebral midline tubular anechoic structure superior to the thalamus, which is contiguous with the dilated saggittal sinus, the "comet tail or keyhole" sign (Sepulveda et al., 1995). In the last 15 years, prenatal diagnosis of this vascular anomaly has been facilitated greatly by the use of colour Doppler sonography. This is crucial in differentiating this lesion from other cystic lesions of the fetal brain because of the marked display of blood flow within the vein of Galen aneurysm (Pilu et al., 1997).

An aneurysm of the vein of Galen can have a varied computed tomogram appearance in infancy. The classical picture is a spherical posterior third ventricular mass with the density of circulating blood contiguous with a dilated straight sinus and with uniform contrast enhancement. This was demonstrated in the case presented. With varying degrees of thrombosis of the aneurysm which may occur in late infancy, the mass can change in density and the dilated straight sinus disappears. With total thrombosis, a precontrast hyperdense rim develops with a low-density centre. This feature was not demonstrated in this patient as the mass enhanced uniformly. The rim enhances but the low-density centre does not change with contrast infusion (Shirkhoda et al., 1981).

On magnetic resonance imaging, the dilated feeding and draining vessels appear as flow-voids on T2-weighted images. Magnetic resonance angiography may also be performed which would better delineate vascular anatomy. Conventional angiography remains the gold standard in full characterisation of the lesion and allows for individual catheterisation of the feeding vessels (Lasjaunias, 1997). Cardiac management of high output heart failure is essential usually involving a paediatric cardiologist as was the case in this patient who eventually improved markedly following management. Seizures which however were not observed in this patient should be managed with antiepileptic medications. Assessment of the child's development is an important part of medical care. Reversible diencephalic syndrome can be seen in some patients, mainly in adults (Gladstone et al., 2001).

Surgical care involves use of a ventriculoperitoneal shunt to relieve the hydrocephalus. This was done for the case presented and his condition improved greatly. Vaso-occlusive therapy including selective catheterization and therapeutic embolization of feeding vessels can be performed (Lylyk et al., 1993). This was not done for this patient due to lack of equipment and expertise. Jones *et al. (2002)* determined prognostic factors using 13 subjects, eight of the thirteen subjects were diagnosed in the neonatal period while the remaining five were diagnosed at age range of four months to thirteen years.

CONCLUSION

The study concluded that children diagnosed with vein of Galen malformation in the neonatal period have a generally much worse prognosis than those diagnosed later in childhood as five of the eight children diagnosed in the neonatal period died. The index patient was not diagnosed in the neonatal period but at 11 months of age which was probably why he had a good prognosis.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

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