VEIN OF GALEN ANEURYSMAL MALFORMATION

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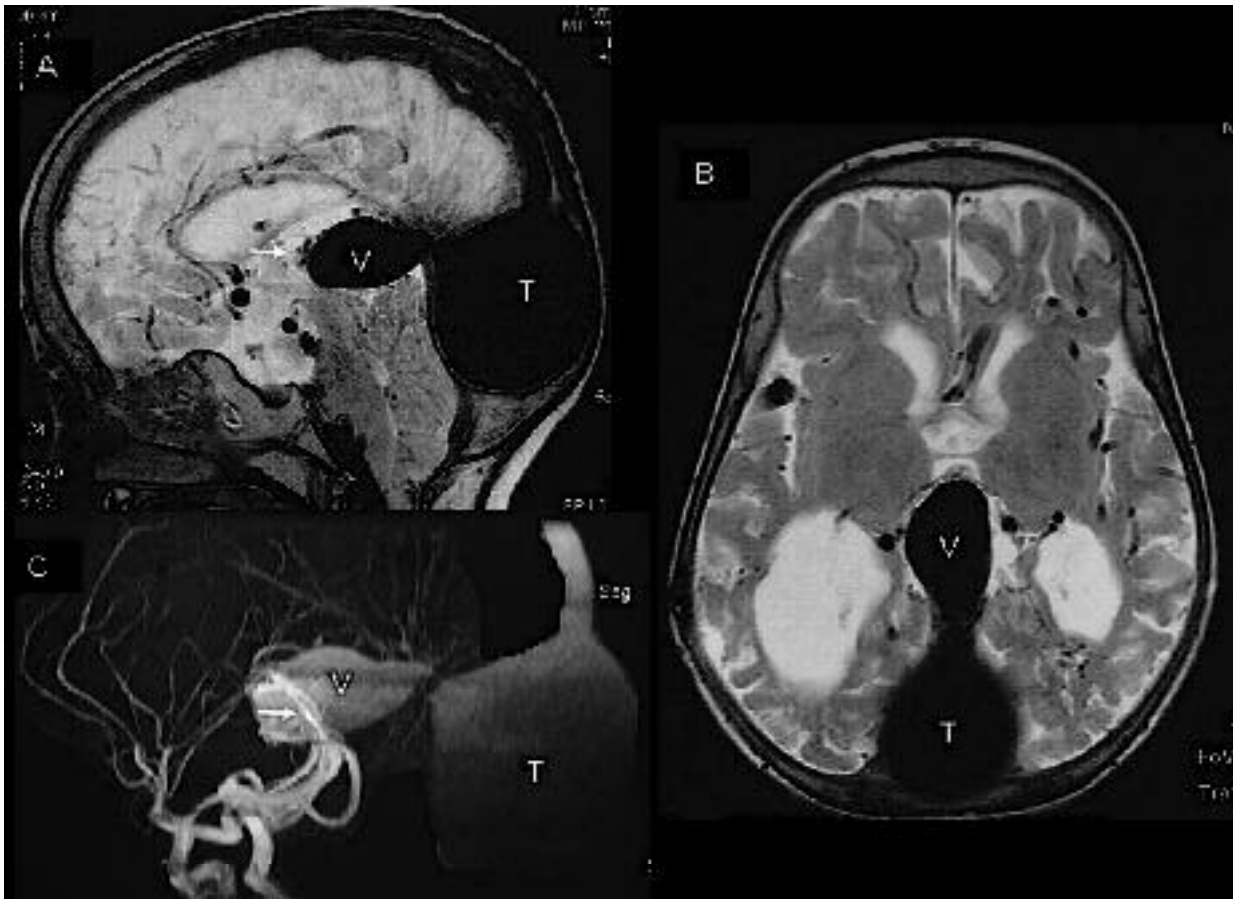

This 4 year-old child presented with markedly delayed milestones. A: T2-weighted saggital MR scan. B: T2-weighted axial MR scan. C: Time Of Flight (TOF) MR angiogram of the intracranial circulation. MR shows an enlarged and dilated promesencephalic vein (V) opening directly into a hugely dilated trocula herophili (T) without an intervening straight sinus. Abnormal feeders arising primarily from the branches of the basilar artery open directly into the wall of the enlarged vein. White arrows: There is contraction of the white matter in the peri-trigonal areas representing ischemic injury due to steal phenomena.

DIAGNOSIS

Vein of Galen Aneurysmal malformation. A formal angiogram was recommended but declined by the family.

COMMENT

The term 'vein of Galen aneurysm' is frequently inappropriately used for a diverse set of vascular anomalies that have in common the dilatation of the vein of Galen.[1] Because of their superficial similarity, arteriovenous malformations resulting in variceal dilatation

of the vein of Galen are sometimes referred to as vein of Galen aneurysmal dilatation (VGAD). The vein of Galen in these cases is inherently normal and also drains brain parenchyma as well as the malformation. True vein of Galen aneurysmal malformation - VGAM - results from development of arteriovenous connections between primitive choroidal vessels and the median promesencephalic vein.[2] The abnormal flow prevents the involution of this embryonic structure which connects either directly to the saggital sinus (as in the illustration) or via a persistent falcine vein. A true vein of Galen is absent in these cases.

VGAMs are rare. Most of these present in early childhood, although the exact presentation is variable. Neonates present with congestive cardiac failure (due to the shunt), infants with hydrocephalus and macro-crania (due to hydrodynamic disturbances presumably due to the venous insufficiency), and developmental delay and seizures in the older child (due to a combination of arterial steal and venous hypertension leading to encephalomalacia).[3] Large shunts may be detectable antenatally. MR imaging is the modality of choice with catheter angiography now reserved for those cases in whom embolization is planned. Without treatment, outcomes are very poor. The advent of trans-catheter embolization techniques have improved the situation; however, the timing and form of intervention remain topics for debate.[4]

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