Teaching NeuroImages: Macrocephaly with subcortical calcifications in vein of Galen aneurysmal malformation

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A previously asymptomatic 3-year-old boy presented with a large head, generalized seizures, and vision impairment for 7 months. Examination revealed macrocephaly (52 cm) and prominent scalp and facial veins. There was bilateral disc pallor and sixth nerve palsies. Developmental assessment and the remainder of the neurologic and systemic examinations were unremarkable. Neuroimaging led to a diagnosis of vein of Galen aneurysmal malformation (VGAM) (figures 1 and 2).

The clinical presentation in VGAM is either related to the high output cardiac failure or cerebral venous congestion and abnormal CSF flow. In untreated VGAMs, ischemia due to venous congestion in watershed region results in development of subcortical white matter calcifications and subependymal atrophy with ventricular dilation.¹

REFERENCE

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