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Case Report

Giant vein of Galen malformation in an adult

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ABSTRACT

Vein of Galen malformations (VoGMs) are rare vascular malformations resulting from persistent shunting of primitive choroidal vessels into the median prosencephalic vein of Markowski. VoGMs are associated with poor clinical outcome with a reported 76.7% mortality if left untreated. We present an exceedingly rare case of a giant, untreated VoGM measuring 7.8 × 5.5 × 7 cm in a 42-year-old man. The embryologic origin, classification, clinical manifestations, and treatment options of VoGMs are discussed with a review of pertinent literature.

Introduction

Vein of Galen malformations (VoGMs) are rare vascular malformations arising from persistent shunting of primitive choroidal vessels into the median prosencephalic vein of Markowski [1–6]. Patients with VoGMs usually present with high-output cardiac failure and hydrocephalus during infancy, with other delayed sequelae such as developmental delay and seizures. It has been associated with poor clinical outcome with a reported mortality of up to 76.7% if left untreated [7].

We present an exceedingly rare case of a giant, untreated VoGM measuring 7.8 × 5.5 × 7 cm in a 42-year-old man. The embryologic origin, classification, clinical manifestations, and treatment options of VoGMs are discussed with a review of pertinent literature.

Case report

A 42-year-old man with a history of seizures, developmental delay, and ventricular peritoneal shunt (VPS) was admitted to our institution for evaluation and treatment of sepsis. Given the history of VPS, a computed tomography (CT) of the brain was obtained which revealed marked calvarial hyperostosis likely due to his antiepileptic use and a large aneurysmal

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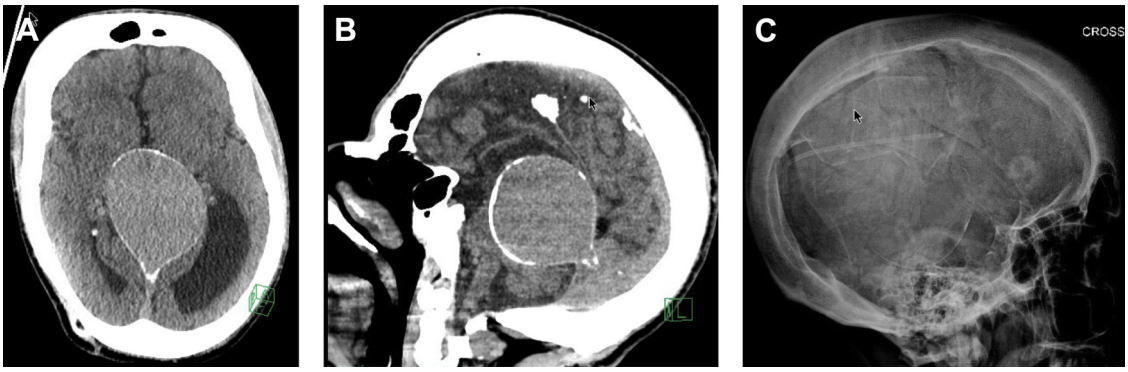

Fig. 1 – (A) A 42-year-old male with history of developmental seizures and ventricular peritoneal shunt with a giant VoGM. Noncontrast axial CT scan shows a giant midline venous dilation in the region of the quadrigeminal plate. Note the calcified rim in the wall of the venous malformation. (B) Noncontrast sagittal CT scan demonstrating a giant midline venous dilation and faint outline of its vascular communication with the straight sinus. There is also marked calvarial hyperostosis. (C) Lateral skull x-ray showing a calcified rim characteristic of a VoGM. CT, computed tomography; VoGM, vein of Galen malformation.

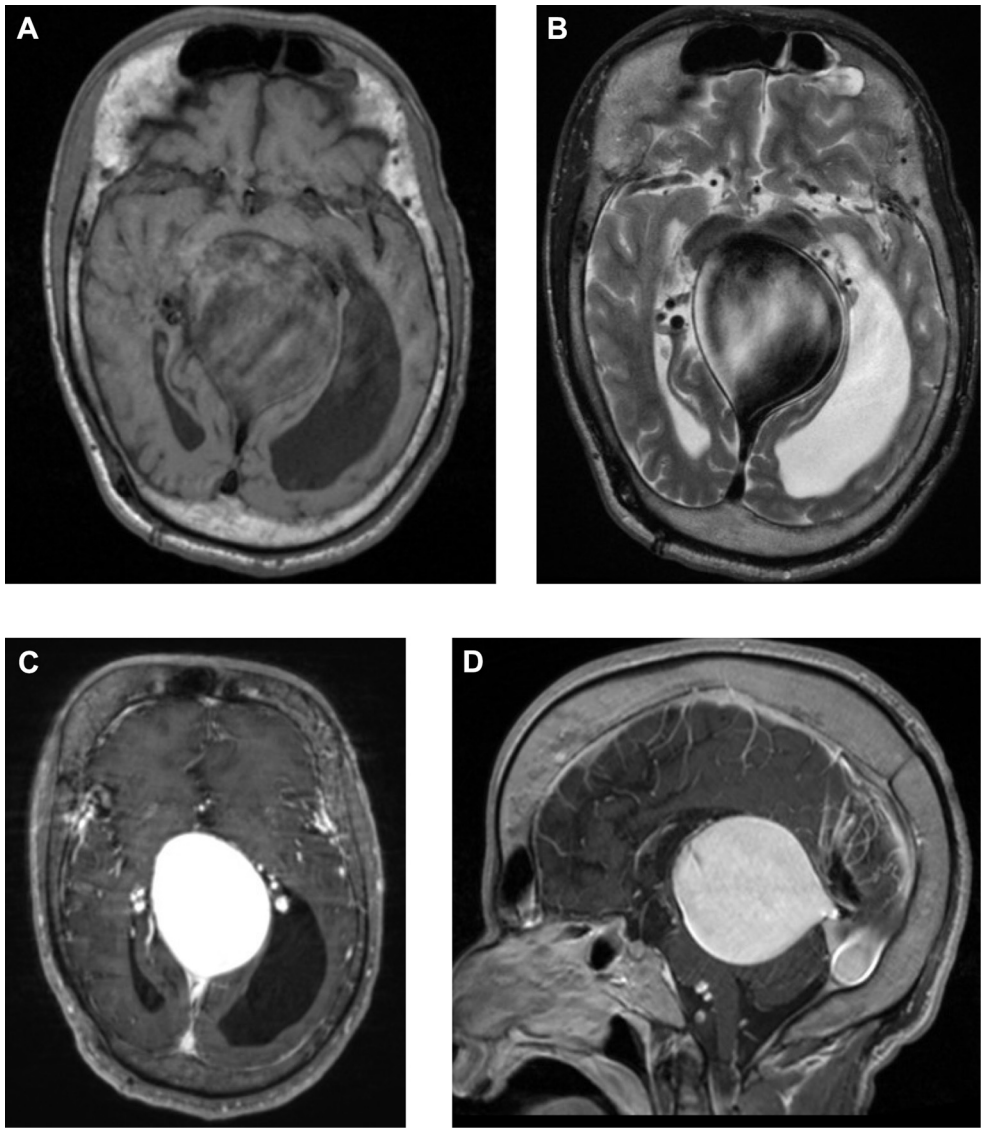

Fig. 2 – (A) Axial T1W, (B) axial T2W, (C) T1W postcontrast, and (D) sagittal T1W postcontrast MRI demonstrating a giant VoGM. MRI, magnetic resonance imaging; VoGM, vein of Galen malformation.