Cerebral venous malformation complicated by spontaneous thrombosis

Abstract  A case of spontaneous thrombosis and infarction leading to death as complications of a cerebral venous malformation in a 13-year-old boy is reported. This is the first published report of this type of complication occurring in a case of venous angioma. While the biologic behavior of cerebral venous malformations has suggested that they are benign in nature, and the results of surgical management have encouraged a conservative approach, the present case illustrates a potential complication and argues against the assumption that these malformations are completely benign in nature.

Key words  Spontaneous thrombosis · Cerebral venous malformation · Infarction

Introduction

Venous malformations are one of the four cerebral vascular malformations and are described as anomalous medullary veins that converge on a centrally located dilated trunk [7]. Histopathologically they consist of ectatic venous channels situated in brain parenchyma unaffected by gliosis or hemosiderin deposition. The lack of an arterial component along with the unaffected intervening neural parenchyma distinguish these lesions from arteriovenous malformations. The manner in which the veins insinuate themselves within the brain parenchyma gives rise to the characteristic “umbrella” appearance on angiogram, first noted in the early venous phase and termed caput medusae [6]. While thrombosis of cerebral veins is a known clinicopathologic entity, no cases have been cited in the literature that concern thrombosis in a cerebral venous malformation. The present report is the first such case to be described.

Case report

Clinical history

A 13-year-old right-handed boy was well until the day of admission, when he became disoriented and started vomiting. His condition pro-gressed with several more bouts of emesis and development of an ataxic gait. When his disorientation proceeded to lethargy, he was taken to a local urgent care center where he was noted to have right ptosis and a fixed and dilated right pupil. He was immediately transported to a local emergency room where he was intubated. Following a CT scan of his head, the patient was transported to a tertiary pediatric trauma center.

On physical examination at the trauma center, the patient was afebrile. No cephalohematoma, abrasions, lacerations, facial deformity, swelling, or rashes were present. The right pupil remained fixed and dilated; the left pupil was reactive. The patient moved the right upper and lower extremities spontaneously and was able to localize pain stimuli. There was no spontaneous movement on the left. There was slight hyperreflexia on the left side. Babinski reflexes were plantar-responsive bilaterally.

Initial laboratory values upon transfer were Na 137 mmol/l, K 3.6 mmol/l, Cl 103 mmol/l, HCO₃ 20 mmol/l, BUN 12 mg/dl, Cr 1.0 mg/dl, glucose 253 mg/dl. Osmolarity was 296 mmol/l. Blood parameters showed WBC 16.6 10⁹/l, platelets 418 10⁹/l, Hct of 39.9, prothrombin time 13.2 s (11.6–13.9 s), and partial thromboplastin time 24 s (23–33 s). Fibrinogen was 247 mg/dl (140–400 mg/dl). The patient’s CT scan from the referring hospital showed a right-side extra-axial mass with midline shift. Head MRI at the tertiary care center revealed abnormal signal within the right temporal and parietal regions, extending into the right basal ganglia and thalamus. The cerebral angiogram was characterized by a paucity of venous drainage from the aforementioned regions with an abnormal appearance on MRI (Figs. 1, 2). No evidence of vascular malformation, aneurysm, or branch occlusion was seen on the angiogram.

The patient was taken to the pediatric intensive care unit where an intracranial bolt monitor was placed. The initial reading measured between 50 and 60 mmHg. Mannitol therapy with intermittent hyperventilation was instituted to reduce the intracranial pressure,
Multiple abnormally ectatic blood vessels both in the neocortex and in the subjacent white matter which extended to the cortical surface (Fig. 4). Serial axial sections of the brainstem showed diffuse swelling, elongation of the midbrain, and necrosis involving the right cerebellar tonsil.

Microscopic examination showed that many of the abnormally ectatic blood vessels, including the vein of Labbé, contained organizing thrombi (Fig. 5), while some were congested without thrombi. The vessels were endothelial-lined and supported by fibrous connective tissue (Fig. 6). The neural parenchyma intervening between the vessels showed no evidence of gliosis or hemosiderin deposition. Accompanying these changes was eosinophilic neuronal degeneration, which was more marked on the side of the lesion.

Postmortem neuropathology report

On gross examination, the external surfaces of the brain showed diffuse cerebral edema in addition to dilatation and thrombosis of the right vein of Labbé (Fig. 3). Right to left subfalcal herniation, right uncal herniation, and cerebellar tonsillar herniation were present. Serial coronal sections of the cerebral hemispheres revealed an intracerebral hematoma in the right temporo-occipital region measuring 5 cm in greatest dimension, immediately surrounded by necrotic cerebral cortex and dusky discoloration of the cerebral white matter. In the right temporal lobe adjacent to the hematoma there were multiple abnormally ectatic blood vessels both in the neocortex and in the subjacent white matter which extended to the cortical surface (Fig. 4). Serial axial sections of the brainstem showed diffuse swelling, elongation of the midbrain, and necrosis involving the right cerebellar tonsil.

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Discussion

Pfannenstiel first described an intracranial vascular malformation composed entirely of veins in 1887 [1]. However, Cushing and Bailey did not recognize venous malformations as a distinct entity until 1928 [4]. Autopsy reports by McCormick and Sawar suggest that venous malformations are the most common vascular malformation, possibly five times more common than their sister lesion the arteriovenous malformation [9].

Many reports and reviews on venous angiomas point out this relatively high incidence in postmortem studies in relation to its incidence of clinical manifestation. The reported clinical manifestations, namely hemorrhage and seizures, appear to have been quite rare and reported mainly in case reports. This combination of being a relatively common entity but relatively rarely reported clinically led to the assumption that venous malformations are benign in nature. However, the clinical relevance of venous angiomas is still being debated [3–6, 9]. Malik et al. [4] reviewed their experience with venous angiomas and noted that of the 21 patients diagnosed with the entity, 9 presented with intracranial hemorrhage. To date this per-