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Letter to Editor

Hemangioma calcificans: A rare entity with epileptogenic potential

Dear Editor,

Hemangioma calcificans (HC) is a very rare intracranial vascular tumor considered to be a variant of cavernous hemangioma or cavernoma.[1] In 1890, Brenner and Carson[2] reported the first known case of this lesion. In 1948, Penfield and Ward[3] introduced the term "haemangioma calcificans" and established the lesion as a definite clinico-radiological entity having epileptic features. Later, Shafey et al.[4] described the lesion as a cause of cerebral calculi (brain stones). Less than 30 cases of HC have been reported in the literature till date.[5]

We describe an additional case of HC in a young male who presented with focal motor seizures.

An 18-year-old male presented with a 1-year history of relatively frequent left-sided focal motor seizures without any incidence of secondary generalization. The patient had a history of primary treatment with antiepileptic drugs in a rural hospital for the last 8 months, following which there was partial improvement of his symptoms. On examination, no significant neurological or neuropsychological abnormality was noticed. General examination and routine tests were also unremarkable. Computerized tomography (CT) scan of the brain revealed a well-circumscribed calcified lesion in the right frontoparietal region without any features of mass effect, midline shift or perilesional edema. Right frontoparietal craniotomy was performed to excise the lesion. Grossly, the resected specimen [Figure 1a] was stony hard, yellowish brown in color measuring 3 cm × 2.4 cm × 1.3 cm. The cut-surface was granular with areas of hemorrhage. Part of the specimen was decalcified and routinely processed for light microscopic examination. On histopathological examination, the lesion was composed of numerous thin-walled vessels of varying caliber, most of which were dilated like cavernous channels [Figure 1b]. Most of the areas displayed intravascular calcified laminated hyaline structures resembling psammoma bodies [Figure 2a], often superimposed on thrombi. Calcification as well as ossification was observed along some of the vessel walls [Figures 2b-c]. Foci of old and fresh hemorrhages and hemosiderin-laden macrophages were scattered in some areas of fibro-collagenous stroma intervening with the calcified vascular channels. The stroma was mostly loose but areas of hyalinization were also noted. No neural tissue was appreciated in between the vascular channels, although the surrounding brain parenchyma featured reactive gliosis [Figure 2d]. The postoperative period was uncomplicated and the patient was completely seizure free on follow-up after 6 months. No residual mass was detected on postoperative follow-up CT scan.

HC is a benign variant of cavernoma that mostly presents with seizures in patients of varying age groups starting from the first decade to the eighth decade.

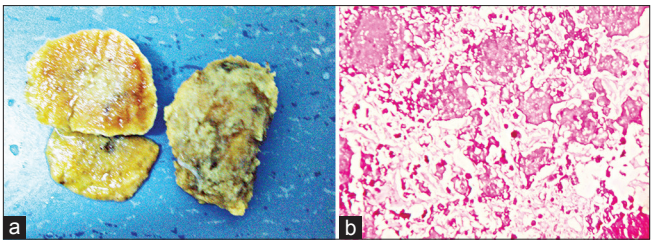

Figure 1: (a) Resected specimen of hemangioma calcificans. (b) Numerous dilated vascular channels with calcifications (H and E, ×40)

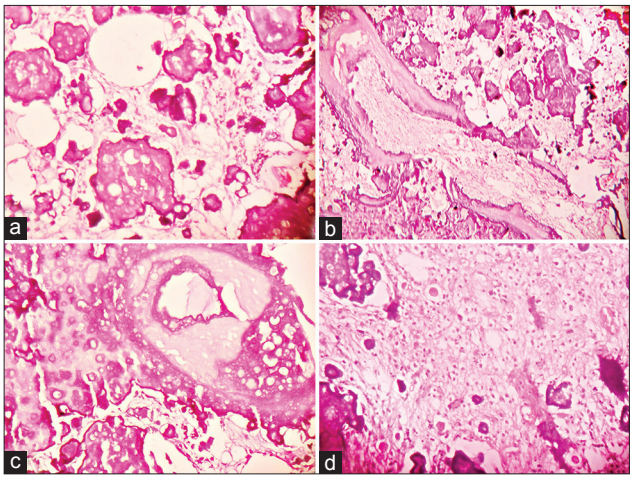

Figure 2: (a) Photomicrograph of hemangioma calcificans showing laminated psammoma body-like hyaline structures with loose intervening fibro-collagenous stroma (H and E, ×100). (b) Photomicrograph showing calcification and ossification along the vessel wall (H and E, ×100). (c) Calcification and ossification along the vessel wall under a high-power objective (H and E, ×400). (d) Photomicrograph showing reactive gliosis in the surrounding brain parenchyma (H and E, ×400)

Although the hemorrhage in HC is rare, cavernoma typically presents with devastating spontaneous repeated intracranial hemorrhage in addition to seizures.[5] There is no evidence of intervening normal neural tissue in HC, indicating its hamartomatous origin, whereas in cavernomas, tongues of dense fibrillary neuroglia may penetrate the compact mass of closely apposed dilated, thin-walled, vascular sinusoids (caverns) for variable depths between the peripheral blood spaces sparing the central part of the lesion. Another vascular lesion, capillary telangiectasia, is characterized by dilated capillaries that are separated by normal brain tissue throughout the lesion with rare chances of hemorrhage.[5,6]

Location of HC is subcortical and often in the temporal lobe, and needs to be differentiated from other calcified intracranial lesions with seizures like low-grade glioma with calcification, meningioma, parasitic cyst, cavernoma, Sturge Weber syndrome, endarteritis calcificans cerebri, calcified hamartoma, granuloma and abscess.[5] Progressive closure of the end arteries in the brain parenchyma, which produces an area of local necrosis, is probably the primary

pathological process related to HC. The degenerated area then becomes secondarily calcified and calcium is laid down along the walls of the involved vessels.[7]

Harbaugh *et al*.[8] reported the very rare case of HC presenting as spontaneous intraventricular hemorrhage in a 44-year-old woman. Hanakita *et al*.[9] described a case with peritumoral brain atrophy and an enlarged subarachnoid space.

The readily accessible lesions of HC should be resected because of the refractory epileptogenic properties, potential for growth and the risk of intracranial hemorrhage, and possible complications like raised intracranial pressure, hydrocephalus and paraparesis. Finally, in most cases, surgical resection leads to complete cure.[5,10]

In the present case, the definitive diagnosis was almost impossible even after the CT scan of the brain. However, histopathology of the resected specimen revealing the intravascular laminated calcospherites, conspicuous absence of intervening neural tissue, surrounding reactive gliosis with supportive clinical history and radiological findings helped us to frame the final diagnosis of HC.

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