



Case Report

Haemangioma Calcificans: An Unusual Cause of Intracerebral Haemorrhage in the Young

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Abstract

A case of temporal lobe hemangioma calcificans is described which is rare and less than 30 cases have been reported in the literature. This is considered to be benign variant of cerebral cavernous hemangioma. This rare case with diagnosis, treatment and clinic-pathological correlation along with review of literature is presented.

Introduction

Haemangioma calcificans (HC), a rare and unusual variant of cavernous hemangioma was first reported by Brenner and Carson in 1890^[1]. Since then less than 30 cases of HC have been reported in the literature till date^[2]. It was only in 1948 that Penfield and Ward coined the term 'haemangioma calcificans' and established it as a definite clinico-radiological entity^[3]. Shafey *et al* described the lesion as a cause of cerebral calculi (brain stones) having a predilection for the temporal lobe and being highly epileptogenic^[4].

Case Report

A 33-year-old male was taken to a hospital with complaints of sudden onset holocranial severe headache associated with vomiting and loss of consciousness. Patient was a nonsmoker, non

alcoholic, vegetarian with no known co morbidities, no history of trauma or any significant relevant past or family history. On admission, his vitals were normal and GCS was 15/15 without any focal neurological deficit. Urgent NCCT head revealed perimesencephalic bleed (subarachnoid haemorrhage: SAH, possibly aneurysmal) without any ventricular or parenchymal extension or any midline shift.

He was referred to a tertiary care neurosurgical centre, where CT angiogram brain on the next day was reported as normal without any major arteriovenous malformation or aneurysm. During the next two weeks his clinical condition improved, till he suddenly had an episode of status epilepticus. NCCT brain showed left Sylvian fissure bleed, in the form of blood in left inferior cistern, quadrigeminal cistern and along gyri of

left frontal lobe with mild compression of left lateral ventricle. Cerebral angiogram showed saccular dilatation in the left posterior communicating artery without any definite aneurysm but associated with vasospasm. An urgent left pterional craniotomy further failed to reveal any aneurysm. He was offered wrapping of the ICA using temporalis muscle. However subsequently he developed status epilepticus with aspiration pneumonia owing to which his condition deteriorated over the next few days till he suffered a cardiorespiratory arrest and passed away after a week.

Limited brain autopsy was performed during which brain parenchyma on the left hemisphere around the site of surgery in the fronto-parietal lobe appeared congested, oedematous and necrosed. Overall brain parenchyma was grossly gelatinous in consistency with marked alteration of normal morphology, which was more marked on the left hemisphere and appeared to flow out of the skull (fig1a). Subsequently coronal sections showed presence of subarachnoid hemorrhage (SAH), in the region of Sylvian fissure (fig 1b). Presence of intracerebral hemorrhage (ICH) measuring 3.5 x 2.5 cm in size in the left medial temporal lobe, with multiple ectatically dilated and congested vessel in the near vicinity, largest measuring around 3 mm in diameter was also observed. In addition, multiple areas of parenchymal congestion and dilated vessels were

noted in the left frontal, parietal and medial temporal lobes. Sections from the right hemisphere, cerebellum and brain stem displayed widespread congestion without any evidence of hemorrhage or space occupying lesion. On gross examination the vertebrobasilar artery, posterior cerebral (bilateral), posterior communicating arteries (bilateral) and right middle cerebral artery appeared dilated and congested, without any evidence of rupture or aneurysm.

Sections from the region of left Sylvian fissure confirmed SAH, while sections from the ICH area in the medial temporal lobe (Lt) was consistent with cerebral cavernous malformation (CCM) (fig 2a). Sections showed a poorly circumscribed lesion composed of tightly packed conglomerate of capillary type vessels with varying thickness (as seen on Masson Trichrome stain-fig 2b) without any internal elastic lamina (as evaluated on elastic Van Gieson stain-fig 2c). There was no intervening brain parenchyma between these vessels, as highlighted by GFAP-immunohistochemistry. In addition, a large area showed prominent psammomatous calcification of vascular channels (fig 2d), which was consistent with haemangioma calcificans (HC). There was no evidence of fibroatheromatous plaque, emboli, thrombus formation, arteriovenous malformation (AVM), aneurysm, rupture or cerebral amyloid angiopathy (after Congo Red stain and Serum Amyloid A immunohistochemistry).

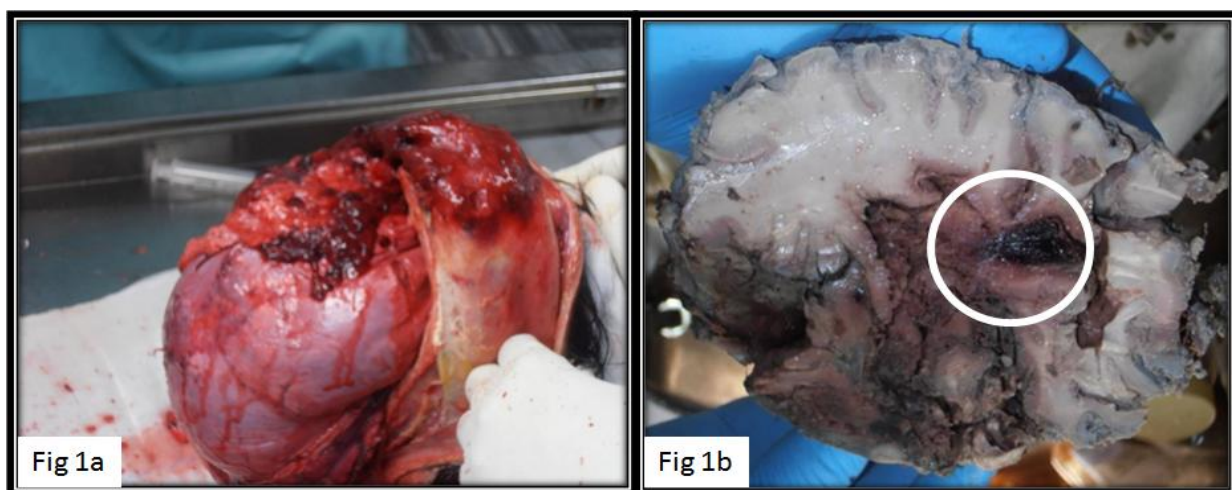


Fig 1 (a) showing enlarged brain with gelatinous consistency.
(b) showing sylvian fissure bleed

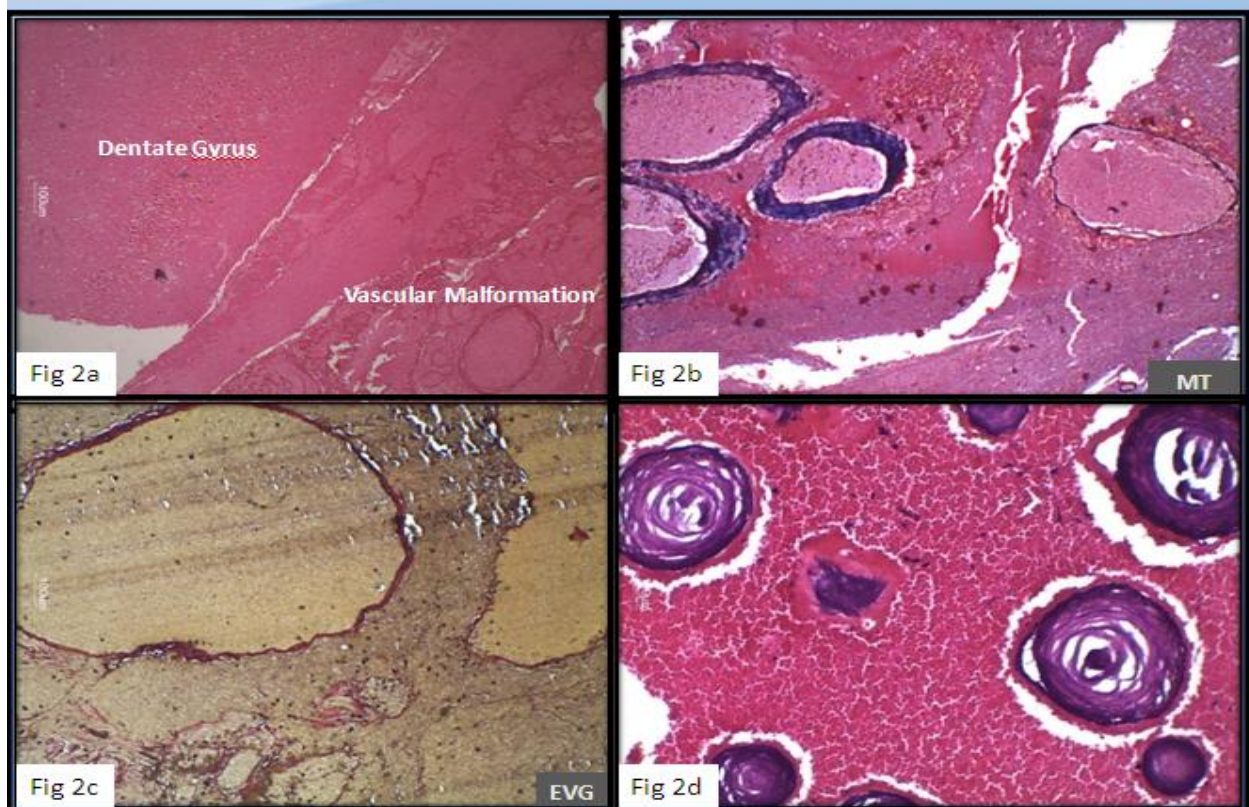


Fig 2 Show temporal cavernous malformation (a) with varying caliber vessels as seen in Masson trichrome stain (b) and without internal elastic lamina as seen in elastic Van Gieson stain (c) and psammoma bodies (d)

Discussion

HC is an unusual and rare variant of cavernous haemangioma of the brain. This relatively rare intracranial tumour is characterized by the presence of a calcified nodule in or near the brain and mostly present with seizures and intracranial haemorrhage.

This rare benign vascular neoplasm has been reported in patients of varying age groups starting from the first decade to eighth decade^[5]. The patient is frequently an adult of either sex, who develops seizures due to the large size of the lesion that causes raised intracranial pressure. Less often the patient presents as intracranial haemorrhage. Harbaugh *et al.*^[6] documented spontaneous intraventricular hemorrhage in a 44-year-old woman with HC, while the case reported by Hanakita *et al.*^[7] had peritumoral brain atrophy with an enlarged subarachnoid space. The tumour is usually solitary and located in the subcortical region and quite often affects the temporal lobe. The index case was also a young male, who

initially presented with intracranial haemorrhage and developed status epilepticus subsequently.

The pathogenesis of HC has been attributed to progressive closure of the end arteries in the brain parenchyma, which initially results in an area of local necrosis, followed by degenerative changes that gets secondarily calcified due to calcium being laid down along the walls of the affected vessels^[8,9]. It needs to be differentiated from other intracranial psammomatous calcifications which can be neoplastic or non-neoplastic in nature. The neoplastic lesions include oligodendrogliomas, meningioma, ganglioglioma and neurocytoma, while non-neoplastic entities consist of parasitic cyst, Sturge Weber syndrome, endarteritis calcificans cerebri, granulomas, calcified hamartoma and abscess.^[5,9]

Microscopically, HC are composed of densely packed capillary channels with widespread calcification, which unlike arteriovenous malformations are poorly circumscribed, lack a nidus, have no internal elastic lamina and show

absence of intervening brain parenchyma. Further, they are distinguished from capillary telangiectasia by the absence of intervening normal brain tissue throughout the lesion with rare chances of hemorrhage.^[10] Ultrastructural examination of HC reveals tubular structures limited by a wall of electron-dense material with prominent irregular calcium deposits^[9].

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 as in most cases, surgical resection leads to complete cure^[11].

Conflict of Interest

All authors have none to declare

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