Cerebral venous angioma and capillary hemangioma. Report of two cases

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Abstract: Cerebral venous angioma is the rarest vascular malformation of the brain. Most congenital intracranial vascular malformations fall into one of four groups: arteriovenous malformations, capillary telangiectasias, cavernous malformations, and venous angioma or developmental venous anomalies. The typical venous angioma is composed of a large parent vein that receives an array of radially-oriented tributary vein in a spoke-wheel configuration dubbed “the caput medusa”. Cerebral venous angioma is usually asymptomatic and may be found incidentally at autopsy or by cerebral angiography. Capillary hemangiomas are benign vascular tumors that are most commonly located superficially in the skin and soft tissue of the head and neck. There appears to be a female preponderance associated with the presence of these lesions. We report two cases of cerebral venous angioma and capillary hemangioma discovered after forensic autopsy.

Key words: nervous system venous angioma, vascular malformations, capillary angioma

Most congenital intracranial vascular malformations fall into one of four groups: arteriovenous malformations, capillary telangiectasias, cavernous malformations, and venous angiomas or developmental venous anomalies [1]. Yet, some intracranial vascular lesions fall outside of this classification and have been dubbed “mixed” vascular malformations since they possess features of more than one type of classic malformation; the hybrid venous angioma-cavernous malformation, venous angioma-arteriovenous malformation, cavernous malformation-arteriovenous malformation, and cavernous malformation-capillary telangiectasia have all been described [2].

The typical venous angioma is composed of a large parent vein that receives an array of radially-oriented tributary veins in a spoke-wheel configuration dubbed “the caput medusa” [3]. Prospective studies on venous angiomas have demonstrated a very low rate of both symptomatic hemorrhage [0.34% per year] and neurologic symptoms; bleeding, when it rarely occurs, has been hypothetically blamed on putative neighboring cavernous malformations [4]. Blood flow through venous angiomas is low and they are thought to drain normal brain. In contrast, the arteriovenous malformation is a true malformation formed from a nidus of tangled vessels in which high flow and pressure result from a direct communication between the arterial and venous system [3].

Cerebral venous angioma is usually asymptomatic and may be found incidentally at autopsy or by angiography [5, 6]. When cerebral venous angioma present symptoms such as cerebral hemorrhage, epilepsy, headache, cranial nerve paresis, and/or cerebral ataxia, surgical intervention has been carried out [7, 8, 9, 10]. However, the indications for surgical treatment remain controversial [11, 12, 13, 14].

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Case Reports

Case 1. A 63-year-old man died in a farm after a generalised convulsions crisis. The forensic autopsy revealed several pathological conditions: cardiomegaly (640g) with left ventricular hypertrophy (left ventricle wall thickness 20 mm), a cerebral hemangioma with intense calcification in the right temporal lobe (4 cm diameter) (fig. 1, 2), a hepatic hemangioma (3 cm diameter) (fig. 3).

Case 2. A 67-year-old women was found dead in his dwelling-house in June 6, 2009. The most relevant autopsy findings were represented by cardiomegaly (determine by dilatation of left ventricle), a cerebral hemangioma formation (0.5 cm diameter) in the left frontal lobe (fig. 4).

Discussion

Cerebral venous angioma is the vascular malformation of the brain. Angiographic demonstration of this lesion was first reported in 1967 [15]. Since then, angiographic findings have been presented in five additional cases [16, 17, 18]. Recently, three other cases have been evaluated, comparing radionuclide flow studies and contrast angiography [19]. Histologically, the lesion is characterized by many irregular and dilated venous channels. Smooth muscle or connective tissue is found in the walls of these vessels, but no elastic tissue. Both grossly and microscopically, a venous malformation resembles an arteriovenous malformation, except that arteries are absent. In contrast to the cavernous angioma, the dilated vessels in venous angioma are separated by neural tissue [20].

Vascular malformations of the brain are congenital anomalies [19]. The term “angioma” is widely used, but is not strictly correct because these lesions show no evidence of cellular proliferation and therefore are not true neoplasms. On the other hand, they do resemble neoplasms in their clinical behavior since they may grow and inflict progressive destruction on the adjacent brain [20].

Cerebral venous angioma is a well-known vascular malformations with a characteristic angiographic appearance of numerous dilated medullary veins converging centrally into a large draining vein in the late capillary or venous phase [5, 21, 22]. Cerebral venous angioma is not a rare vascular malformation, representing 2.6% of the population studied by Sarwar and McCormick in 1978: 165 of
4069 autopsied brains had one or more vascular malformations, and cerebral venous angioma was found in 105 of the 165 brains [6]. Cerebral venous angioma is usually clinically silent [6], but may cause neurological disorders [5] such as hemorrhage, epilepsy, cerebellar ataxia and/or cranial nerve paresis.

Some investigators [11, 13, 23] have stressed the clinical significance of cerebral venous angioma as a cause of intracerebral hemorrhage, while Hacker et al. [12] and Saito and Kobayashi [14] argued against this. These opposing opinions may be partly a result of the obscure etiology of cerebral venous angioma, both with and without hemorrhage. The absence of angiographic change on follow-up studies has been reported sporadically [24, 25]. These studies suggest that asymptomatic and most symptomatic cerebral venous angiomas are stable either clinically or angiographically over a long period. In other word, spontaneous disap-pearance of cerebral venous angioma cannot be expected [26].

Handa and Moritake [5] mentioned that cerebral venous angiomas are prone to bleed, when they are of medium size, located in the deep cerebrum and cerebellum, and dhow no early appearing vessels on angiography. Some investigators [11, 13] stressed the need for expeditious removal of hemorrhagic cerebral venous angioma that might be likely to bleed. If complete extirpation of cerebral venous angioma, widely distributed in the normal brain parenchyma, is difficult during operations. Secondly, cerebral venous angioma may play a part in the main draining system of the surrounding brain parenchyma [14, 27] so obstruction or resection of the draining vessels including the cerebral venous angioma may cause serious venous infarction [7, 27, 28] or acute brain swelling [26].

Capillary hemangiomas rarely occur in the central nervous system [29]. Most documented cases have been described in the spinal roots and the cauda equine [30, 31, 32]. Only sex cases of intracranial capillary hemangiomas has been previously reported in the literature. The prevalence of intracranial capillary hemangiomas may be underreported if they regress in size over time and remain asymptomatic.

Capillary hemangioma must be distinguished from more common intracranial vascular lesions such as cavernous hemagiomas and hemangioblastomas. Pathologically, cavernous hemangiomas consist of dilated hyaline vessels, and demonstrated thrombosis, perivascular hemosiderin deposition, and calcification [33]. The pathogenesis, behavior, and natural history of intracranial capillary hemangioma are not known. Unfortunately, capillary hemangiomas cannot be differentiated from other intracranial...
lesions based on imaging finding ale, thus warranting the examination of a tissue specimen for a definitive diagnosis [29].

Treatment modalities that have provided successful outcome in published case reports for symptomatic capillary hemangiomas have included resection [34], radiosurgery [35] and pharmacotherapy (steroid agents and interferon-α) [36]. If a tissue diagnosis of capillary hemangioma is confirmed intraoperatively, aggressive resection may not be necessary [29].

References