Cerebral proliferative angiopathy

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Since Cushing and Dandy reported their cases about “monstrous” vascular malformations with high mortality index in 1928, neurosurgery has been devoting efforts to study such complex pathology among the most important ones in the modern medicine¹. Cerebral vascular malformation is a relatively rare pathology and its prevalence is difficult to estimate since a few number of affected individuals remain symptomatic. The classification contemplates size, local and venous draining territory². Those malformations are congenital results of an anomalous cerebral development. Physiopathology, angioarchitecture and natural history distinguish many types of malformations such as arteriovenous malformations (AVM), venous angiomias, cavernous malformations, dural arteriovenous fistulas and capillary telangiecasy¹,². The AVMs are dilated tortuous vessel masses characterized by direct connections between arteries and veins (shunts) without a capillary net interposition¹. A peculiar type of large brain AVM with multiple non dominant arterial feeders to a lobar or hemispheric nidus, relatively small draining veins associated to intermingled normal brain and angiographic evidence of capillary angioectasia and angiogenetic activity determines an atypical entity named cerebral proliferative angiopathy (CPA)³. Rare vascular pathologies associated with skin and other organ lesions designate neurocutaneous angiomatosis syndrome³. Sturge Weber syndrome (SWS) and Rendu-Osler-Weber syndrome (ROWS) are examples of it⁴,⁵.

We report a female with an atypical diffuse lobar cerebral proliferative angiopathy (CPA).

CASE

A 39-years-old white female was admitted to our department presenting a chronic left, pulsatile, highly intense hemicrania headache. Physical practice increased it. She presented recurrent episodes of left hemiparesis and hemihyposthesia stopping completely in one to three hours. Left hemiparesis was permanent. She smokes chronically. Neither was hypertension related nor convictions.

Computed tomography (CT) revealed a high density image in the left frontal cortex with contrast enhancement suggesting an AVM nidus.

Magnetic resonance image (MRI) did not characterize a typical AVM nidus, but a diffuse network of densely enhancing vascular spaces with intermingled normal brain parenchyma among different vascular territories (watershed zones) on the left frontal lobe, lower and medium frontal gyri, pre-central gyrus and basal ganglia (Fig 1).

MRA (Fig 2) and angiography showed a spread angiopathy supplied by numerous arterial branches leading to late venous filling (Fig 3), without markedly dilated superficial or deep cerebral veins, draining to superior sagittal sinus and Rosenthal basal veins (Fig 4). There was a diffuse cortical-subcortical capillary angioectatic net spread to the whole left frontal lobe (Fig 4).
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with intra orbital extension (Fig 5). Irrigation did not reveal a dominant arterial feeder, predominating over the left medial cerebral artery, left anterior choroid artery, left ophthalmic artery, anterior cerebral arteries and perforating branches to basal nuclei (Fig 3). Irrigation contribution by falcine anterior artery, from ophthalmic artery, and right anterior cerebral artery, is a typical pattern of hemodynamic steal phenomenon (Fig 6). Left internal carotid aneurysms are evident (Fig 5).

Symptomatic medical treatment failed. Patient’s six-month outcome revealed permanent headache, dizziness, left parestesis and paroxysmal weakness without epileptic seizures.

DISCUSSION

Our patient suffers from an atypical angiopathy which differs from other AVM in their angiomorphology, histology, presumed mechanism, epidemiology, natural history and clinical presentation. It may therefore be classified as a distinct group of AVM.

Efforts made towards specific diagnosis include careful analysis of angiomatous syndromes.

Hereditary hemorrhagic telangiectasia, or Rendu-Osler-Weber syndrome, Sturge Weber syndrome, the Wyburn Mason syndrome and the angiomatosis of Divry Van Bogaert (ADB) are some vasculopathies that should be among differential diagnosis.

Although these rare syndromes share some features with our patient, none of them entirely resembles.

Cerebral proliferative angiopathy appears as a presumed diagnosis for a peculiar type of large brain AVMs, predominating at 3.4% of them, 67% being females.

Clinical aspects point to small chance of hemorrhage 12%, 45% of seizures, 41% of disabling headaches and 12% of neurological progressive deficits, with stroke-like symptoms and even transitory ischemic attacks.

Despite the absence of seizures, our patient is a young female with relevant complaints of headache, left paresthesia and intermittent paresis, totally related to CPA physiopathology, concerning “nidus” hyperperfusion associat-
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ed with hemispheric hypoperfusion, characteristic signals of hemodynamic steal phenomenon.

The main differences between CPA and typical AVMs are the numerous non dominant feeders including transdural supply to lobar or even hemispheric “nidus”, moderately enlarged draining veins and normal brain tissue intermingled between vascular spaces. Increased blood volume within the nidus and delayed blood transit time are predictive of the ectatic capillary net formation.

In fact, there is a progressive vicious cascade in which arterial supply associated with venous ectasia creates an environment of local increased blood volume and perinidal areas of severely hypoperfusion triggering an uncontrolled progressive angiogenic response to this normal brain with abnormal blood demand.

Treatment of AVMs, based on the Spetzler-Martin’s classification, may be done by surgical resection, embolization and radiosurgery. In CPA however, the normal brain tissue between vascular spaces carry the risk of permanent neurological deficit. Headache may be dramatically alleviated by limited arterial embolization in nonelusive areas. As the major pathomechanism of this disease is ischemia owing to incompetent angiogenesis; seizures, headaches and steal phenomenon, similar to Moya-Moya-like diseases, may be treated with calvarial burrholes that increases cortical blood supply by recruiting additional dural blood supply.

The main propose of this case report is indeed the knowledge of the CPA, as a differential diagnosis of AVM syndromes, once an AVM diagnose mistake implicate in potentially lethal results if surgery or embolization would be performed.

Vast bibliographical revision makes evidence of the extreme rare incidence of cerebral proliferative angiopathy which enhances the paramount importance of the case.

REFERENCES