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# Neuroimaging Highlight

Editors: Richard Farb, David Pelz

## Cerebral Proliferative Angiopathy

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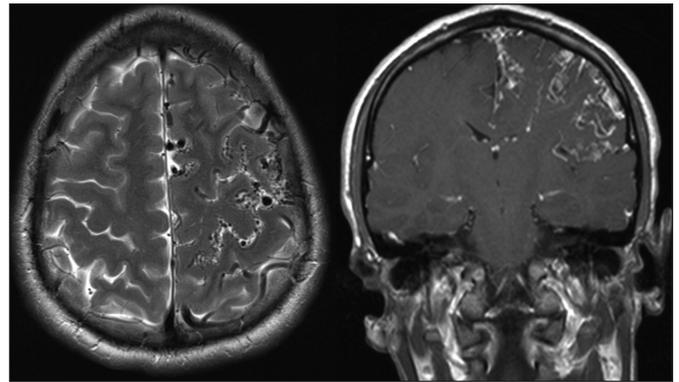
### CASE REPORT

A 31-year-old male presented with transient numbness of the right hand, progressing to involve the forearm. The symptoms lasted for around 30 minutes without any associated weakness or positive motor phenomena. An evaluation in the emergency room revealed no focal deficits. A computer tomogram (CT) scan performed in the emergency room suggested a few abnormal vascular structures in relation to the left hemisphere, with no evidence of hemorrhage. The patient was referred for magnetic resonance imaging (MRI) and conventional angiography.

The MRI showed a diffuse network of densely enhancing vascular spaces involving the left middle cerebral artery (MCA) and anterior cerebral artery (ACA) territories without a clear nidus or abnormal parenchymal signal change (Figure 1). There was no involvement of the basal ganglia or thalami. Given the size and extent of the abnormality, there was a relative paucity of draining vessels, which were only moderately enlarged.

A catheter angiogram revealed moderately enlarged MCA and ACA feeders supplying a diffuse area of abnormal vasculature without a well circumscribed nidus (Figure 2). Although there was a large area of rapid arterio-venous transit, given the extent of the abnormality it was felt not to be of a high-flow fistulous nature. There was no supply from posterior cerebral artery (PCA) branches or from the contralateral side. These features were consistent with cerebral proliferative angiopathy.

Cerebral proliferative angiopathy is a distinct vascular lesion which has some features distinct from other parenchymal brain arteriovenous malformations (AVMs)<sup>1</sup> and is relatively rare comprising 3.4% of all brain AVMs in one series<sup>2</sup>. Angiographically these lesions do not have a large dominant feeding vessel or flow related aneurysms and show the presence of capillary angioectasia without a well circumscribed nidus and only moderately enlarged veins relative to the extent of the vascular abnormality<sup>2</sup>. From a histological standpoint, unlike 'classical' AVMs, these lesions have normal brain parenchyma interspersed with proliferative vascular channels<sup>2,3</sup>. When comparing the two major types of vascular malformations in general described by Mulliken et al<sup>4</sup>, these lesions appear to be



**Figure 1:** Axial T2W and coronal gadolinium enhanced MR scans show areas of abnormal vascular flow voids involving the left MCA and ACA territories, showing enhancement on the post-gadolinium scans. There is a relative paucity of draining veins, which are only moderately dilated relative to the area of involvement.

intermediate between those lesions that demonstrate a purely proliferative endothelial component such as hemangiomas and those with a structural abnormality of the arterial, capillary or venous system such as the 'classical' AVMs<sup>5</sup>.

There is also the hypothesis that a hypoperfusion trigger to the involved brain might lead to uncontrolled angiogenesis that contributes to the proliferative response, to the extent that it may also lead to the recruitment of abnormal transdural supply. This was however absent in our case. There have been studies using perfusion MRI in these patients which showed an increase in mean transit time with areas of hypoperfusion to the affected hemisphere, extending even beyond the boundaries of the morphological abnormality<sup>6</sup>.

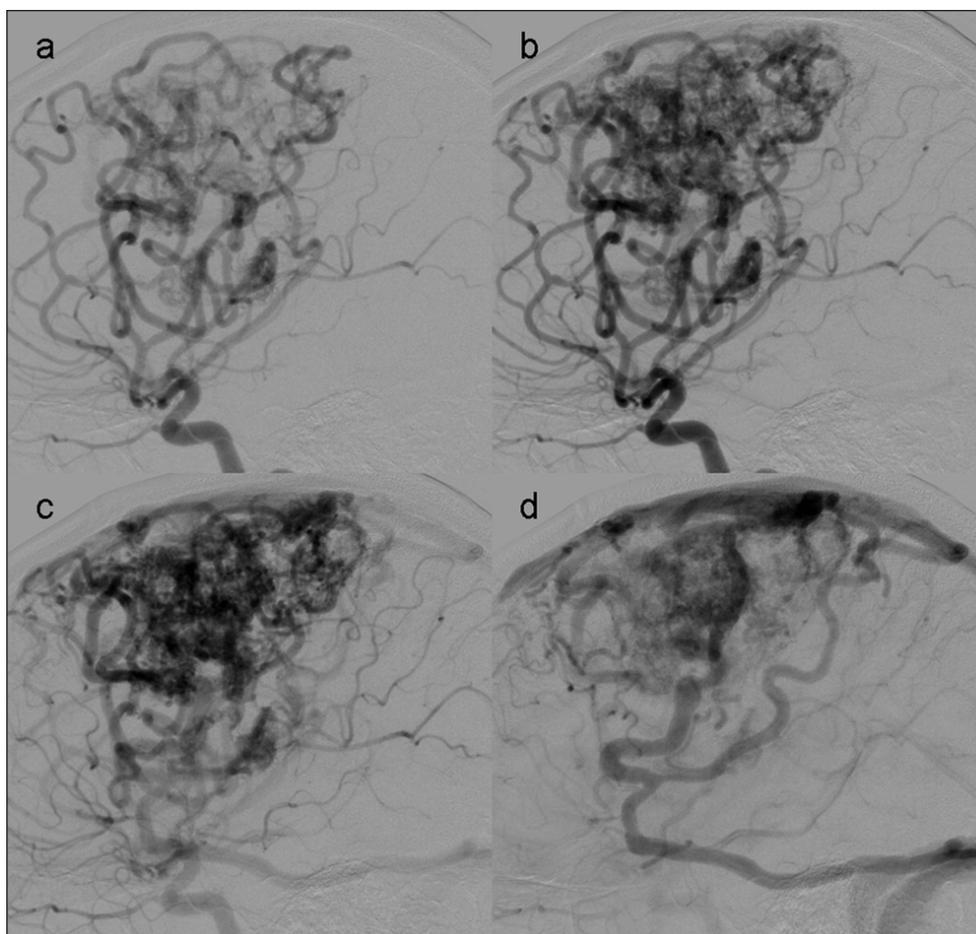
Clinically this disease seems to present in a less aggressive fashion, with a relatively low risk of hemorrhage at presentation

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**Figure 2:** Lateral views of serial phases of a catheter angiogram show dilated MCA and ACA feeding arteries supplying a diffuse area of abnormal vasculature with capillary angioectasia without a circumscribed nidus. There is rapid arteriovenous transit without high flow and only moderately sized draining veins. There are no flow related aneurysms proximally.

and usually presenting with seizures or headaches. When they do bleed, however, the risk of recurrent hemorrhage seems to be higher<sup>2</sup>.

Treatment of this entity is extremely challenging and indicated only in cases with intractable seizures or angi-morphological evidence of zones of weakness or evidence to suggest inadequate perfusion to eloquent brain<sup>2,3</sup>. In some patients persistent headaches are dramatically alleviated by a partial and limited arterial embolization in non-eloquent areas, without treatment of the dural component<sup>2</sup>. As with embolization, surgery and radiation may also have detrimental effects as normal, eloquent intervening brain may be affected.

It is important to recognize this rare entity from the various features as described above and to consider that the clinical profile and natural history of this disease are different from classical brain AVMs. Treatment of this disease is challenging and management of such patients is best done at centres with expertise in cerebrovascular diseases.

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