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UNCOMMON CLINICO RADIOLOGICAL MANIFESTATION OF CEREBRAL PROLIFERATIVE ANGIOPATHY

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ABSTRACT

Cerebral proliferative angiopathy (CPA) is a distinct entity unlike classical brain arteriovenous malformations (AVMs). CPA is reported as a rare entity, corresponding to 3.4% of all brain AVMs. CPA presented as seizures, headaches and neurological symptoms related to cerebral haemorrhage. Cerebral angiography is the gold standard for CPA diagnosis; however CT angiography and MR angiography can also be accurate in making the diagnosis among the other cerebral vascular malformations. There is absence of early venous drainage, which helps to differentiate CPA from a classical cerebral AVM, the nidus is fed by multiple arteries (absence of a dominant feeder), classical nidus appearance with scattered "puddling" of contrast which persisted into the late arterial and early venous phase.

Key Words: Cerebral proliferative angiopathy (CPA), Arteriovenous malformations (AVMs), Angiography.

INTRODUCTION

Cerebral proliferative angiopathy (CPA) is a rare entity, corresponding to 3.4% of all brain AVMs. It presented as seizures, headaches and neurological symptoms related to cerebral haemorrhage. Its diagnosed by CT angiography and MR angiography but cerebral angiography is the gold standard. We presenting a 42 year old male with an atypical angiopathy which is distinct from other brain AVMs [1].

CASE REPORT

A 42-year-old male presented with one episode of generalized tonic clonic seizure and right sided headache for 8 days. There was neither any history of headache, vomiting or any stroke like episode nor his neurological examination was suggestive of any focal neurological deficit.

Initially magnetic resonance imaging brain was done elsewhere which revealed multiple abnormal dilated vascular channels in right temporo-occipital region, right superior parietal region and left superior fronto-parietal region suggestive of abnormal proliferation of vascular channels [Figure 1]. Few of these vascular channels in the right posterior temporal region appears hyperintense on T1WI and shows significant blooming on GRE sequences suggestive of possible thrombosis. Brain parenchyma is normal. No evidence of any nidus is seen on MR images. Subsequently CT cerebral angiography and cerebral digital subtraction angiography (DSA) was done at our center. CT cerebral angiography and cerebral digital subtraction angiography (DSA) showed diffuse network of densely enhancing vascular channels in right posterior parietal, right temporo-occipital region with intervening normal brain parenchyma. Persistence of contrast in delayed phase was noted suggestive of absence of early venous drainage. No dominant feeder artery is seen. These findings were suggestive of cerebral proliferative angiopathy (CPA) [Figure 2]. Patient was managed conservatively with antiepileptic drugs with strict adherence to compliance. He was seizure free and doing well.

DISCUSSION

Our patient was having an atypical angiopathy which is distinct from other brain AVMs [1] and is

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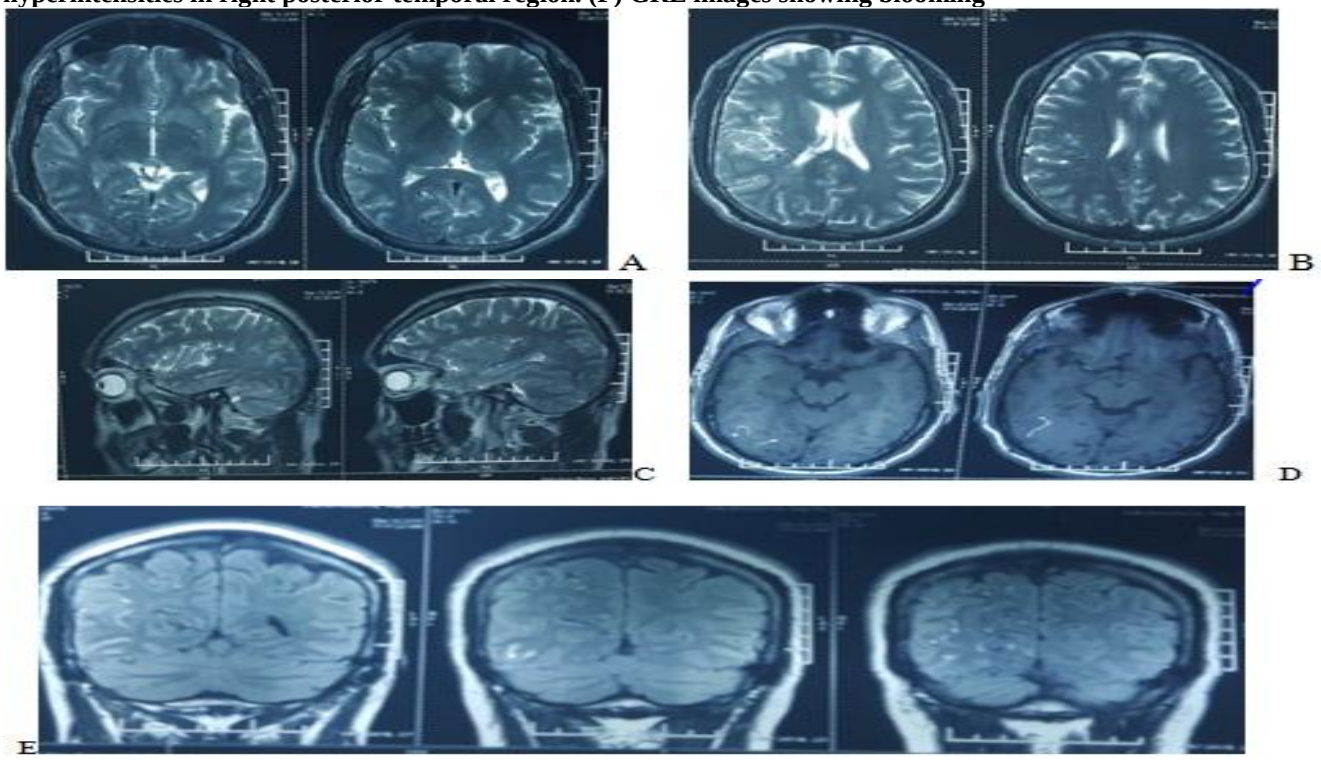
relatively rare comprising 3.4% of all brain AVMs in one series [2]. Only few cases of this rare clinical entity have been described in the world literature till now. The salient issues of cerebral proliferative angiopathy (CPA) which helps to discriminate it from classical brain AVMs. The absence of dominant feeders or flow-related aneurysms, the presence of proximal stenosis on the feeding arteries, the extensive transdural supply to both healthy and pathological tissues, the large size (which might be lobar or even hemispheric), the presence of capillary angioectasia and the only moderately enlarged veins (compared with the size of the nidus). In our patient cerebral DSA (Fig 2)showed diffuse angiogenesis, absence of a well-formed nidus and early venous phase suggestive of fast capillary transit favoring the diagnosis of CPA.

This entity has a significantly different clinical presentation and clinical course compared with classical brain AVMs. Patients (typically, young females 2:1) usually do not present with an acute neurological deficit or hemorrhage but more commonly with epileptic manifestations, headaches and progressive neurological deficits, however, when they do bleed, the risk of recurrent hemorrhage seems to be higher [2]. Similar was our observation never lead to hemorrhage or any focal neurological deficit but only served as an epileptogenic focus. Other particular features are the high rate of stroke-like symptoms. Seizures and disabling headaches also occur in the CPA population more than the AVM population. Treatment indications in the series of Lasjaunias *et al*, were set very strictly and were confined to hemorrhage, identifiable fragile angioarchitecture (such as intranidal aneurysmal ectasias), uncontrollable seizures and disabling headaches. Surgery, radiosurgery and large non-targeted embolization carry the risk of permanent neurological deficit attributable to the interspersed normal neural tissue. These kinds of treatment should therefore be reserved to patients with otherwise intractable headaches and epilepsy.

Because one of the major pathomechanisms of this disease is ischemia a therapy that enhances cortical blood supply can be indicated. Apart from targeted embolization, other treatment options are synangiogenesis and calvarial burr holes both of which act by increasing cortical blood supply but there is not sufficient data to establish their utility as a primary treatment modality [2, 3]. However, in patients presenting with hemorrhage, endovascular treatment should be performed, aiming at fragile areas that may be identified during angiography [4]. Our patient, neither presented with hemorrhage nor was there was any fragile area or any intranidal aneurysm identifiable on DSA, making target embolization useless and leaving us with conservative treatment as the best possible option.

Fig 1. Magnetic resonance imaging (A,B,C) brain axial and sagittal T2-weighted images showing multiple flow voids in right in right temporo-occipital region, right superior parietal region. D, E T1-weighted images showing hyperintensities in right posterior temporal region. (F) GRE images showing blooming



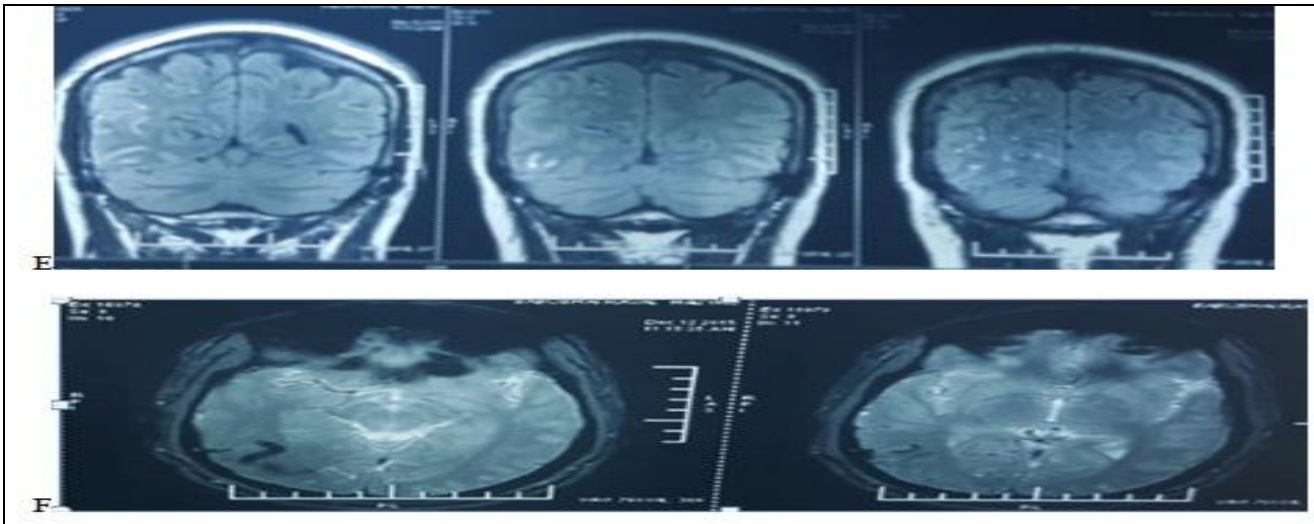
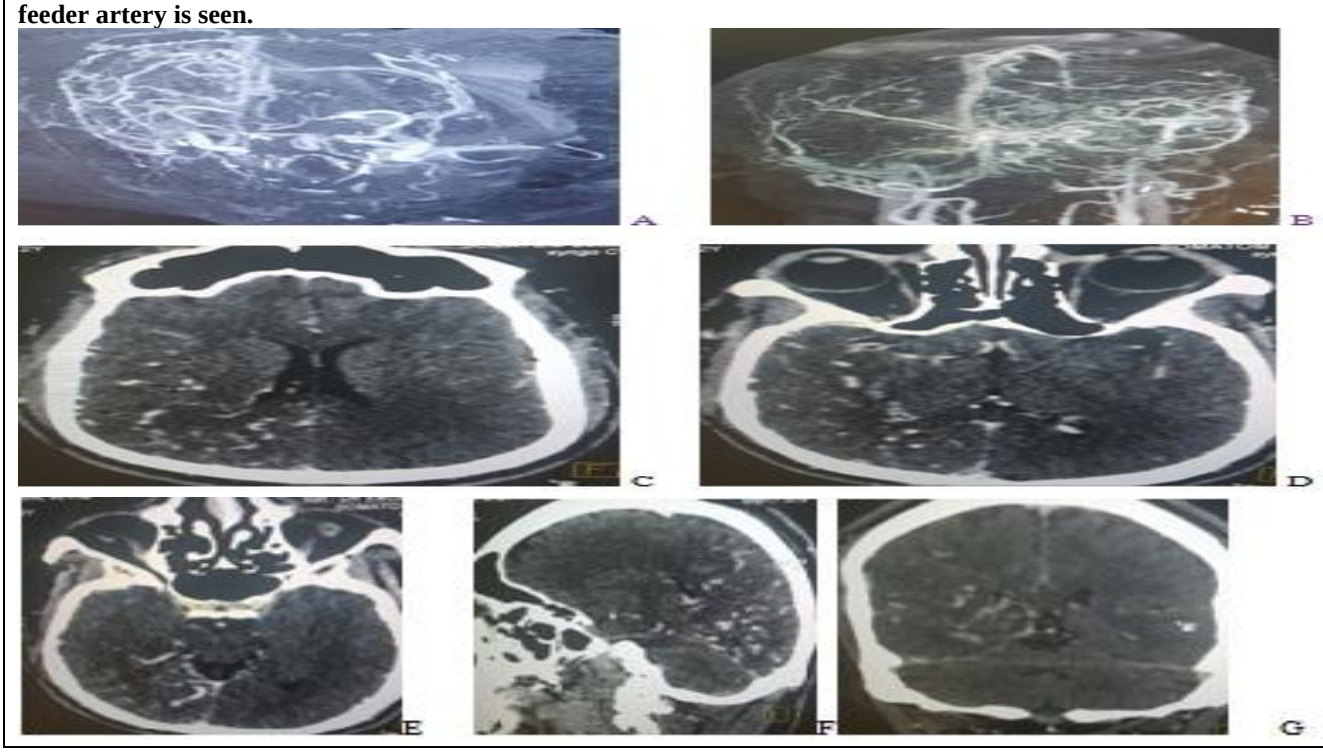


Fig 2. CT Cerebral digital subtraction angiography (A,B) showing diffuse proliferative angiogenesis. Post contrast CT (C,D,E,F,G) brain axial, sagittal and coronal images showing diffuse network of densely enhancing vascular channels in right posterior parietal, right temporo-occipital region with intervening normal brain parenchyma. No dominant feeder artery is seen.



CONCLUSION

It is important to recognize this rare entity of CPA from the various features as described above. Treatment of this disease is challenging and management of such patients is best done at centers with expertise in cerebrovascular diseases.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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