

Cerebral proliferative angiopathy: a rare cause of stroke and seizures in young

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DESCRIPTION

A 12-year-old boy presented with one episode of generalised tonic-clonic seizure and sudden onset left hemiplegia to the emergency department. He had two episodes of febrile seizures in childhood with no other significant history. His non-contrast CT of the head showed right frontal lobar haemorrhage with mass effect and an old infarct in the right anterior frontal lobe (figure 1A). He underwent decompressive hemicraniectomy. His digital subtraction angiography in the arterial phase (figure 1B) showed multiple areas of abnormal leash of small vessels in the right frontal and parietal regions fed by the cortical branches of the right Middle Cerebral Artery with no early draining vein appreciated in the venous phase (figure 1C) suggesting cerebral proliferative angiopathy. He was given physiotherapy and he improved with a modified Rankin Scale (mRS) score of 1 after 2 months on follow-up.

DISCUSSION

Cerebral proliferative angiopathy is a rare and distinct entity, which differs from classical cerebral arteriovenous malformation in epidemiological, clinical and angiomorphological features. It is more commonly seen in adolescent and middle-aged females. The common presentations are seizures, headache and progressive neurological deficits. Haemorrhages are less common in cerebral proliferative angiopathy than in classical arteriovenous malformation. The lesions are large with normal brain parenchyma between the vascular spaces.^{1 2} The angiography in cerebral proliferative angiopathy shows multiple arterial feeders, which are not enlarged or moderately enlarged without any dominant feeder. The draining vessels are scanty compared with the size of the nidus unlike classical arteriovenous malformation.¹ The treatment is often challenging as surgery and embolisation may cause permanent neurological deficit, due to damage of interspersed normal neural tissue in cerebral proliferative angiopathy.³

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Contributors AR drafted the manuscript and had role in organisation and execution of the project. CKA had role in critical review of the manuscript and assessment of the radiological



Figure 1 Cranial non-contrast CT (A) reveals right frontal lobe haemorrhage with mass effect. Catheter digital subtraction angiography of right Internal Cerebral Artery (RICA) in the arterial phase (B) demonstrates abnormal leash of vessels (white arrows) in the right frontal lobe fed by cortical branches of Middle Cerebral Artery (MCA) with no early draining vein appreciated in the venous phase (C).

Learning points

- ▶ Cerebral proliferative angiopathy is a distinct entity, which differs from classical cerebral arteriovenous malformation.
- ▶ It is a rare cause of stroke and seizures in young and its treatment is challenging.

findings. SA had role in organisation and execution of the project. RS had role in conception, organisation and execution of the report and critically reviewed the manuscript.

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