Unusual case of giant cerebral infiltrative cavernoma causing obstructive hydrocephalus

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DESCRIPTION

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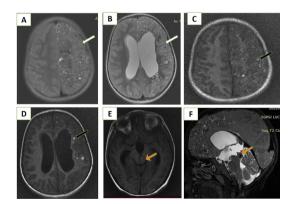
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A teenage girl presented with reports of recurrent headache, recurrent episodes of generalised seizures and weakness of the right upper and lower limbs for 1 year. Her previous history and family history were unremarkable. She was conscious and oriented to time, place and person on examination. Mild sensory and motor deficits were present in both the right upper and lower limbs (motor strength grade 4/5).

The MRI of the brain (figures 1 and 2) revealed a spongiform mixed signal intensity lesion involving the left frontal-parietal lobe (measuring $12.2 \times 5.3 \times 7$ cm) involving both white and grey matter. The lesion consists of multiple tiny (less than 1 cm) T1-weighted (T1W) and T2-weighted (T2W) hyperintense cystic areas surrounded by T1W/ T2W hypointense rim representing haemosiderin. Associated mild volume loss was seen in involved lobes, leading to left lateral ventricle undulated appearance. Few similar smaller lesions were seen in the midbrain extending into the floor of the third ventricle and cerebral aqueduct, causing obstruction and mild upstream hydrocephalus (figure 1E and F). Another similar small lesion was seen in the left thalamus. On susceptibility-weighted imaging (SWI) (figure 2), diffuse blooming was seen in the entire left frontal-parietal lobe, with small foci of



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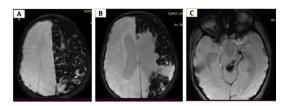


Figure 2 MRI brain. Susceptibility weighted imaging (SWI) images in the axial plane (A, B, C) show diffuse blooming in the entire left frontal-parietal lobe, with small foci of blooming in the midbrain.

blooming in the midbrain and left thalamus. No diffusion restriction was seen on diffusion weighted imaging. A diagnostic digital subtraction angiography revealed mild irregularity of cortical branches of the left middle cerebral artery and cortical veins with no developmental venous anomaly, arteriovenous malformation or fistula.

Based on these imaging findings, a diagnosis of diffuse and multifocal brain cavernoma was made. Lesions in the midbrain were causing compression at the level of the third ventricle and cerebral aqueduct, leading to mild upstream hydrocephalus. Near-complete (more than 90%) surgical resection of the cavernoma was done using a microsurgical approach and functional mapping. A thin rim of the residual lesion was left adjacent to the motor cortex, left thalamus and midbrain. A ventriculoperitoneal shunt was placed in the left lateral ventricle. She recovered gradually with mild residual haemiparesis on the right side. Histopathological examination of the lesion showed multiple vascular channels lined by single layer of dysplastic epithelium interspersed with minimal astroglial brain tissue. These findings were consistent with radiological diagnosis of gaint infiltrative cavernoma.

Cerebral cavernoma refers to intracranial vascular malformation of developmental origin.

Patient''s perspective

My daughter has been unwell for a long time. Previously her seizures were controlled with medications, but gradually they were uncontrollable. Therefore, we decided to take her to a higher centre. Doctors in the hospital have done a few investigations where they found multiple blood-filled lesions in the brain. They discussed different treatment options and all the risks and benefits of treatment. Finally, she underwent surgery. Her seizures are well controlled now, although there is some weakness in her limbs.

Learning points

- Cerebral cavernoma is usually round, lobulated hamartomatous lesions with normal brain parenchyma between them. On imaging, they are well-defined lesions showing blood in different stages producing popcorn appearance with surrounding T2 hypointense rim.
- Infiltrative giant cavernoma is a rare and atypical type of cavernoma showing diffuse infiltration of brain parenchyma involving multiple lobes.
- Mass effect and obstructive hydrocephalus can be rarely seen in infiltrative cavernomas.

These malformations consist of clusters of variable-sized abnormal capillaries containing slow-flowing blood.¹ The incidence of cavernous angioma is around 0.5% in the population.¹ Both sporadic and familial forms are described. Giant cerebral cavernoma refers to cavernoma of more than 6 cm.² On imaging, cavernoma usually appears as a well-defined round lobulated lesion with large cystic cavernous areas showing mixed signal intensity with a peripheral hypointense rim.³ However, in the present case, lesion was diffuse, infiltrative and large (12 cm in maximum diameter). Cystic spaces were tiny with no normal intervening brain parenchyma visualised. However, the presence of dense and confluent blooming favoured the diagnosis of cavernoma.⁴ Another unusual feature in this case was presence of obstructive hydrocephalus caused by cavernoma in mibrain

bulging into the aqueduct. Previously, only one similar case was reported in the literature where lesions showed an infiltrative pattern in the right cerebral haemisphere.²

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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