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SUBEPENDYMAL GRAY MATTER HETEROTOPIA

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Background: A 31-year-old woman presented at the department of neurology with a history of seizures since childhood. About five years ago her seize pattern changed from generalized atonic attacks with asphyxia to simple partial attacks characterized by left hemiparesis and speech difficulties. These seizures occurred multiple times a day and typically lasted about a minute. The patient had a negative family history of epilepsy.

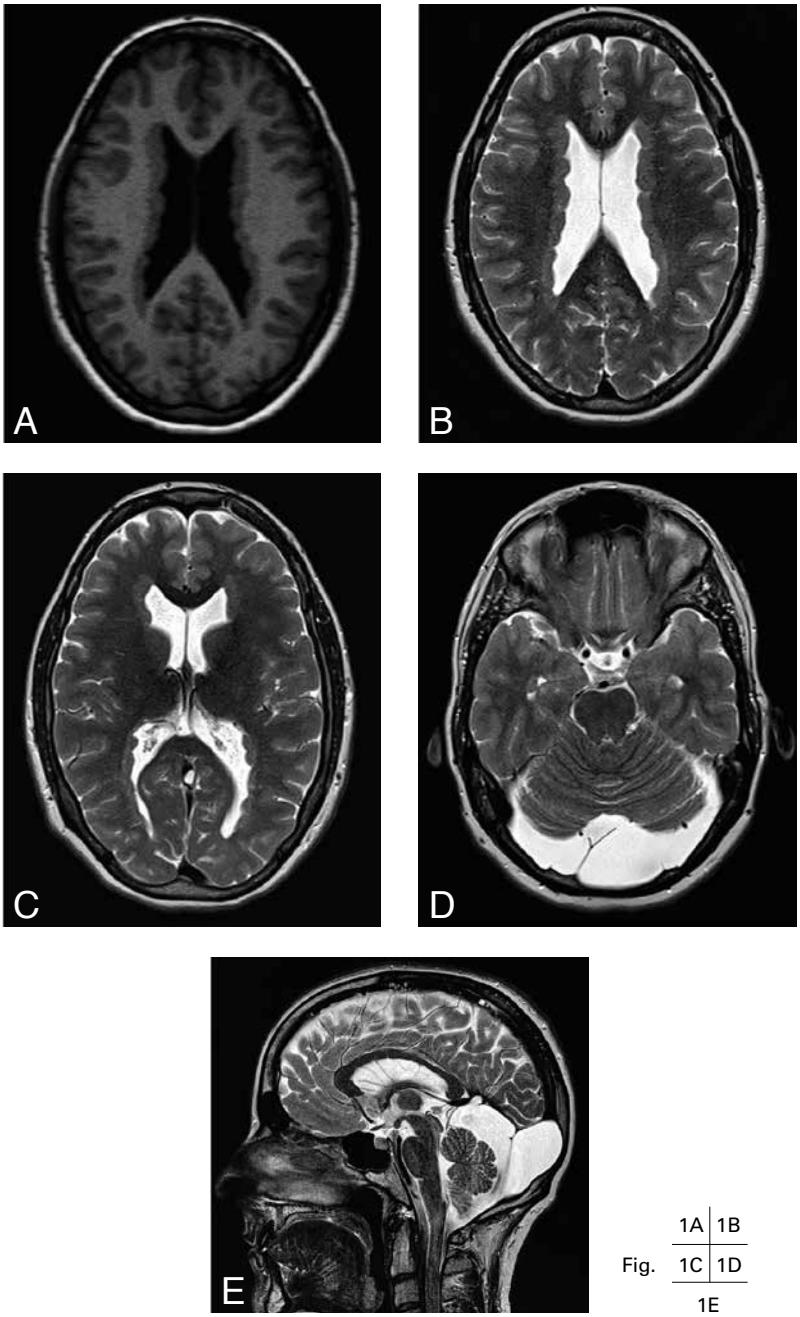

Fig. 1A | 1B / 1C | 1D / 1E

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Work-up

MRI of the brain (Fig. 1) consists of an axial MPRAGE image (A), axial T2-weighted images (B-D) and a sagittal T2-weighted image (E). Images at supratentorial level (section through the lateral ventricles) (Fig. 1a-c) show multiple diffuse continuous nodules lining the lateral margin of both lateral ventricles and protruding into the lumen of the lateral ventricles. The lesions are iso-intense to gray matter on all imaging sequences.

Images at infratentorial level and in the sagittal plane (D-E) show a large extra-axial non-enhancing retro- and supracerebellar cystic structure. A few thin internal septations are present. There is mild mass effect on the cerebellum and tentorium and scallopping of the occipital bone. Also notice the presence of a small periventricular nodule protruding into the lumen of the temporal horn of the right lateral ventricle.

Radiological diagnosis

Based on the characteristic MRI findings the diagnosis of *subependymal gray matter heterotopia* (also known as periventricular nodular heterotopia) was made. As an incidental finding a large arachnoid cyst was observed in the posterior fossa.

Discussion

Gray matter heterotopias are common malformations of cortical development and result from the failure of clusters of neurons to migrate away from the embryonic ventricular zone to the developing cortex. They can be discovered during the evaluation of adults and children with epilepsy, children with neurodevelopmental disorders, or as incidental findings. Only since the last two decades, with the advent of MRI, the presence of heterotopia is revealed much more frequently than before.

Clinically and radiologically distinction is made between subependymal, subcortical, and band (or double cortex) heterotopia forms. Subependymal gray matter heterotopia (SEH) is the most common form and consists of one or more gray matter nodules located immediately beneath the ependyma of the lateral ventricles. Five different groups of SEH are distinguished: (1) bilateral and symmetrical; (2) bilateral single-noduled; (3) bilateral and asymmetrical; (4) unilateral; and (5) unilateral with extension to neocortex. Bilateral and symmetrical SEH is the best known form of SEH and is characterized by familial occurrence, a clear female predominance and a positive family history of epilepsy. Therefore, genetic factors are believed to play a major role in the etiology, which is supported by further genetic research (genetic linkage analysis).

Bilateral single-noduled and asymmetric bilateral cases of SEH are also characterized by a striking gender prevalence and familial occurrence.

In unilateral periventricular nodular heterotopia no association with sex, nor familial occurrence are noticed, but it is presumed that acquired factors, damaging a limited region of the developing brain, may provoke the genesis of unilateral nodules.

Patients with bilateral symmetrical SEH are usually normal individuals affected by focal epilepsy, frequently drug-resistant but not characterized by high frequency seizures. Bilateral symmetrical SEH is associated with posterior fossa malformations such as cerebellar hypoplasia and megacisterna magna. The latter one is by far the most frequent associated anomaly. An association between posterior fossa arachnoid cysts and bilateral symmetrical SEH has not been reported in the literature. Therefore we consider the large posterior fossa arachnoid cyst in our patient an incidental finding.

SEH can be confidently diagnosed by CT scan and MRI. On CT scan irregularities along the lateral margins of the lateral ventricles may be the only clue to the diagnosis of SEH. MRI is superior in the imaging of SEH and can precisely delineate the morphology, distribution and extent of SEH. The signal intensities of the nodules are identical to those of gray matter on all sequences, including contrast-enhanced series, which, like normal gray matter, show no enhancement of the nodules.

The main differential diagnosis of SEH is tuberous sclerosis. The subependymal nodules can be observed in tuberous sclerosis, are often calcified (except in early childhood) and have a signal on T2-weighted images that is higher than that of normal gray matter. Other typical brain abnormalities such as cortical tubers and white matter lesions are often present in tuberous sclerosis and can guide the radiologist towards the correct diagnosis.

Bibliography

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