Clinical History

A 23-year-old female patient was admitted with a 2-month history of repeated attacks of generalized convulsions occurring during sleep. No past history of head trauma nor epilepsy was noted. Physical examination did not reveal any abnormality, particularly no focal neurological deficit. Routine laboratory tests were normal and computerized tomography of head was performed.

Questions

1. What are the radiographic findings?
2. What is the most likely diagnosis?
Figure 1 - Axial CT scan before contrast injection. Left pre-rolandic area solid round shaped well demarcated hypodense cortical mass measuring 2.7 x 2.3 cm without significant enhancement.

Figure 2 - Axial CT scan after IV contrast injection. Left frontal superficial hypodense well delineated solid mass showing no contrast uptake.

Figure 3 - MRI, transverse T2W1. Left frontal well demarcated mass displaying lobulated outlines and high signal intensity. Remodeling of the adjacent inner table and diploe is seen. Note absence of edema.

Figure 4 - Post contrast T1W1 scan. Left frontal hypointense well defined cortical mass with mild subcortical extension.
CT scan (Figures 1 & 2) demonstrates in the left pre-rolandic area a solid round well demarcated hypodense cortical mass measuring 2.7 x 2.3 cm without significant enhancement seen after IV contrast injection. Further characterization of this mass was performed by MR scan. The tumor shows homogenous high signal intensity on T2W (Figure 3) and low signal intensity on T1W1. No detectable contrast uptake was seen after Gadolinium IV injection (Figure 4). The mass is mainly cortical, without any surrounding edema or mass effect on the midline structures. Note also the thinning of the adjacent inner table. These features are suggestive of the diagnosis of low-grade glioma and dysembryoplastic neuroepithelial tumor (DNET).

Diagnosis

The patient had complete surgical resection. The diagnosis of DNET was confirmed by histological findings. No seizure as reported 6 months after surgery.

Discussion. Dysembryoplastic neuroepithelial tumor is a recently recognized entity, first introduced into literature by Daumas Duport in 1988. It retained attention because of its high association with intractable seizures and its favorable prognosis. It is a benign neuroglial tumor located generally in the cortex, but may extend into the adjacent subcortical white matter. It constitutes approximately 10% of all tumors from patients with intractable epilepsy. More than 60% of DNET are found in the temporal lobe, 30% in the frontal lobe and the remainder are scattered through the parietal and the occipital lobes, the deep cerebral nuclei and the brain and cerebellum. The presenting symptom is almost invariably complex partial seizures, often intractable. The tumor is commonly seen in young patients and the ratio of male-female is variable. Macroscopically the tumor is mainly solid seated in the cortical area, but often has cystic or macrocystic components. It is found to involve both cortical surface and the white matter. Microscopically, the tumor is characterized by a high degree of cellular pleomorphism including astrocytes and oligoastrocytomas. Daumas Duport considered this tumor as a benign lesion because of the excessive cellular proliferation, its symptoms chronicity and the absence of recurrence after resection. The imaging findings in patients with DNET are highly suggestive. Despite their histological heterogeneity the majority of DNET appear on CT as well demarcated lobulated cortical masses that are hypodense and do not enhance after IV contrast. But in certain patients, the CT scan has been reported to be normal despite the positivity of MRI. Intra-tumoral calcification was detected in approximately 25-30% of cases. Magnetic resonance imaging details the anatomical location and extent of the tumor. Dysembryoplastic neuroepithelial tumor exhibits low signal intensity on T1W1 and high signal intensity on T2W1, without peritumoral edema and mass effect. Gyriform tumoral configuration on T1 or T2W1 was a prominent sign observed in all patients of Kuroia series. Because the lesions are well demarcated showing lobular margins, the lesions are primarily cortical, DNET of the convexity are frequently (up to 60%) associated with bone remodeling of the adjacent calvarial inner table. Subcortical extension is seen in about 30%. Contrast enhancement is observed in up to 20-40% and may be diffuse or partial.

Differential diagnosis, based on MR features, includes ganglioma and other low grade gliomas, but unlike DNET these neoplasms do not have propensity for cortical involvement. Recognition of DNET is important, because surgical resection is almost always curative and earlier intervention can prevent the physical and psychological damage resulting from chronic seizures.
References