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(CASE REPORT)

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Dysembryoplastic Neuroepithelial Tumors (DNETs): A rare cause of epilepsy in children

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Abstract

A dysembryoplastic neuroepithelial tumor (DNET) is a low-grade, slow-growing brain tumor. It is a glioneuronal tumor, which means it contains properties of both glial and neuronal cells. DNETs can manifest as seizures that may be drug-resistant. The initial diagnosis is made by MRI or the radiological appearance is often suggestive. In this work we report the case of a patient in whom the DNET type tumor was revealed by convulsive seizures.

Keywords: Brain Tumor; DNET; Pediatrics; Spectro-MRI; Epilepsy; Drug resistance

1. Introduction

Brain tumors are the most common solid tumors in children (1). Approximately half of these are located in the supratentorial region (2). They are not always easy to diagnose, as they may initially present only with vomiting, abdominal pain, or developmental delays. Subsequently, other signs of increased intracranial pressure may appear, such as headaches and focal signs like cranial nerve palsies, motor or balance deficits, and seizures. Indeed, the clinical manifestations associated with these tumors depend on the location and the potential for growth of the lesion. Dysembryoplastic neuroepithelial tumor (DNET), a hemispheric cortical tumor, is thought to cause focal epilepsy in 14% of cases (2,3). We report the case of a patient diagnosed in our department following a presentation of an epileptic seizure.

2. Case Report

M.A., a 10-year-old girl with no significant past medical history, was admitted to our department for myoclonic seizures that had been occurring for four months prior to admission, at a rate of two episodes per day in the absence of fever. The patient was initially prescribed magnesium by her primary care physician. The condition worsened, with the seizures becoming hemi-tonic and affecting more than seven times a day, accompanied by urinary incontinence; hence her hospitalization in our department. On admission, the child was conscious, respiratory and hemodynamically stable, with no neurological deficits and a completely normal clinical examination.

On the paraclinical level, the waking electroencephalogram (EEG) was normal; while the sleep EEG revealed right frontotemporal interictal epileptic abnormalities in the form of spike-wave discharges sometimes spreading to the left, as well as bursts of slow waves in the same location. The brain CT scan (image 1) noted the presence of a right temporal intra-axial lesion. A brain MRI (image 2) revealed the presence of a well-defined, rounded, heterogeneous right temporal intra-axial cortical lesion, hypointense on T1-weighted images, hyperintense on T2-weighted images/FLAIR, enhancing after gadolinium injection, containing cystic locules and areas of T2* signal, measuring 27 x 20 x 21 mm, associated with

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perilesional edema in a "glove finger" pattern; Spectroscopy revealed an inversion of the NAA/Cho ratio, with an increase in choline and a decrease in NAA, and a slight increase in inositol and lactate. These radiological findings suggested a dysembryoplastic neuroepithelial tumor (DNET). Furthermore, the patient's ophthalmological and laboratory tests were normal. The patient was started on sodium valproate (Micropakine) at a dose of 30 mg/kg/day and is scheduled for tumor resection.

3. Discussion

DNETs are low-grade neuroglial tumors (WHO Grade I) with very low potential for progression and minimal oncological risk. They represent 14% of epilepsy-related tumors and 1.2% of all resected tumors in patients under 20 years of age (1,2,3,4). Initially described by Daumas-Duport in children treated with surgical resection of epilepetogenic tumors (5), they are most often discovered during the etiological workup of focal epilepsy refractory to antiepileptic treatments, but their prevalence as a cause of childhood epilepsy remains unknown. Several studies agree on an average age of symptom onset between 6 and 10 years and note a slight male predominance (1,6).

3.1. Location of DNETs

The location is cortical/subcortical, most often in the medial temporal lobe (62%), as described in our patient, or frontal lobe (31%) (7). Rare intraventricular forms or forms developing within the basal ganglia and thalamus, in the insular cortex, brainstem, cerebellum, and occipital lobe exist (1,8,9).

3.2. Histology and Tumor Markers

The most characteristic histological feature of DNETs is the specific glioneuronal component. They consist of several nodules containing columnar structures (axons) closely associated with pseudo-oligodendroglial cells and a few scattered stellate astrocytes. The background has a mucoid appearance and contains non-dysmorphic "floating" neurons. The pseudo-oligodendroglial cells express OLIG2, while the neuronal component expresses neuronal markers (10).

According to the Daumas-Duport classification, there are three histological types:

- Simple specific DNET with a specific glioneuronal component associated with a pseudocystic appearance;
- Complex specific DNET associated with a pseudomulticystic appearance (glioneuronal component and multinodular architecture with cortical dysplasia); Association with cortical dysplasia is frequent in this form and is described in 50 to 83% of cases; conversely, the absence of cortical dysplasia on histopathological examination is thought to be linked to a poor prognosis (5,6).
- Nonspecific DNET associated with a homogeneous or heterogeneous nodular or pseudodysplastic appearance (no glioneuronal component) (2,11).

3.3. Molecular Biology

DNETs are frequently associated with an alteration of the FGFR1 gene (partial duplication, point mutation, or, more rarely, FGFR1-TACC1 fusion), leading to activation of the MAP kinase pathway. BRAF V600E mutations, as well as other alterations of tyrosine kinase receptors, have been less frequently encountered (5-10% of cases) (12,13,14).

3.4. Clinical Presentation

From a clinical perspective, the location of DNETs explains their clinical presentation; indeed, they most often manifest as focal epilepsy (as in our patient), drug-resistant with or without secondary generalization, which can appear in childhood or adolescence, but diagnosis can be delayed until adulthood if imaging is not performed promptly (15).

It should be noted that DNETs are highly epileptogenic tumors, and the type of seizure depends on the tumor's location, which often affects the mesiotemporal cortex (1). Generalized tonic-clonic seizures and, more rarely, myoclonus have been reported, as was the case in our patient. Furthermore, the tumor has been revealed in some patients by headaches and, more rarely, by congenital focal neurological deficits (16).

3.5. Radiology

Radiological examination helps guide the etiological diagnosis of DNETs. Indeed, DNETs often have a typical appearance on brain CT and MRI scans. On brain CT, DNET presents as a well-defined, hypodense, triangular lesion, without edema or mass effect on adjacent structures (even in cases of large lesions), sometimes leaving an indentation on the cranial

vault. In some cases, DNETs may present as multilobulated lesions with septa composed of multiple pseudocysts. Bone scalloping is common, and calcifications are present in one-third of cases. There is an adjacent bone deformity in 40% of cases (2, 7, 15).

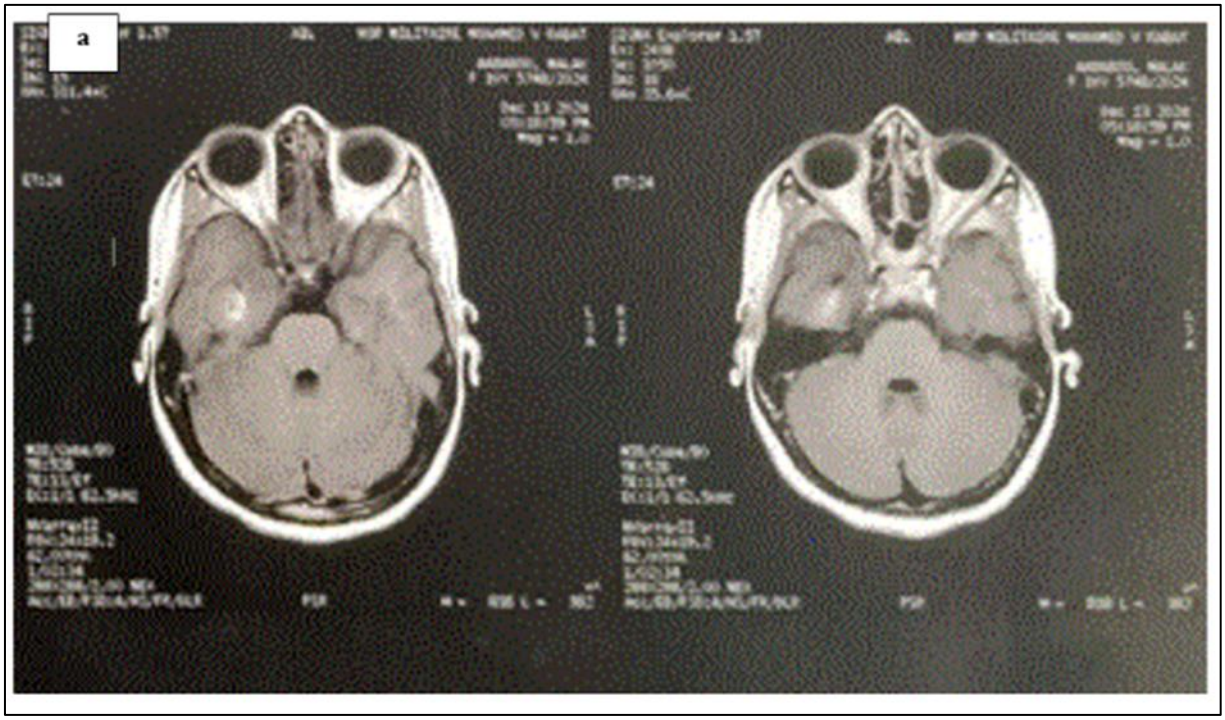

Figure 1 Cross-sectional brain scan showing a right temporal intra-axial lesion

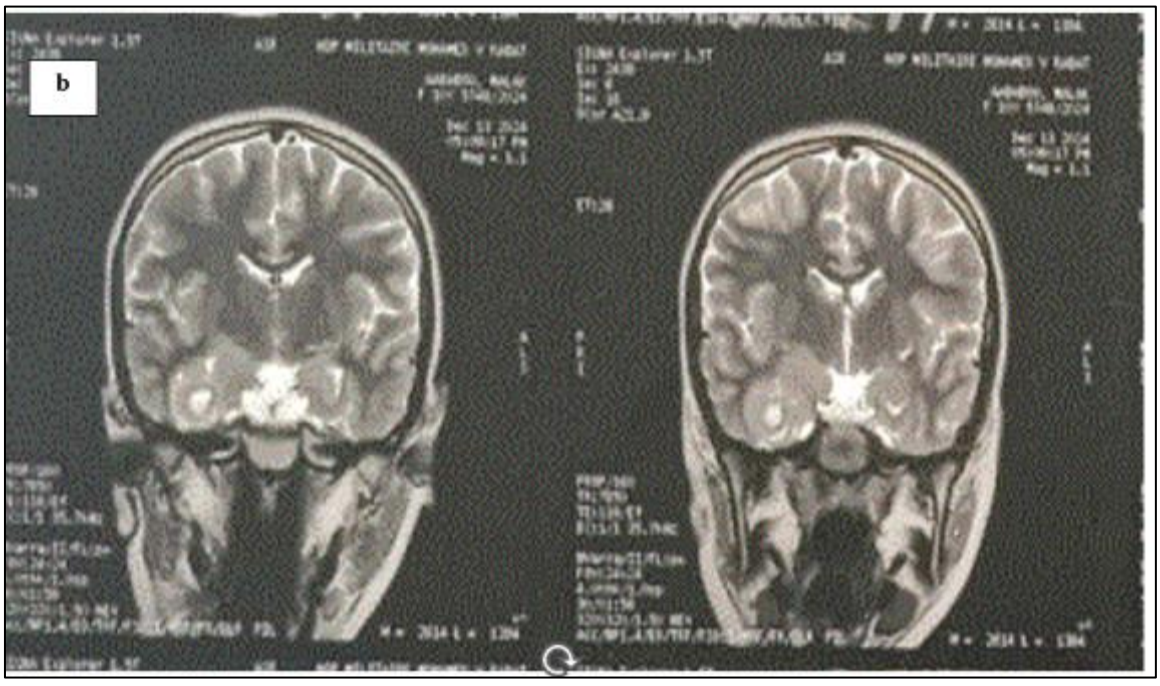

Figure 2 Transverse (a) and sagittal (b) brain MRI showing a well-defined, rounded, heterogeneous intra-axial lesion in the right temporal cortex, hypointense on T1-weighted images and hyperintense on T2/FLAIR

On MRI, DNETs are multinodular (soap-bubble appearance), cortical, appearing hypointense on T1-weighted images, with a distinct pseudocystic, multiloculated hyperintense signal on T2-weighted images, and hyperintense on FLAIR images. This appearance was indeed noted in our patient. Furthermore, contrast enhancement is exceptional in children. When present, it may be peripheral and subtle, or sometimes more nodular. The presence of contrast enhancement is associated with a greater risk of tumor progression and should initially suggest the diagnosis of ganglioglioma (2, 18, 19). However, it is important to note that DNETs can exhibit spontaneous changes in their signal or contrast enhancement during follow-up. MR spectroscopy, on the other hand, reports an elevation of myo-inositol and a non-significant decrease in NAA, as well as an elevation of choline (found in our patient), less pronounced than in gangliogliomas (19, 20).

3.6. Treatment

The therapeutic management of DNETs is medical and surgical; anticonvulsant treatment is indicated in cases of epileptic seizures and is provided by sodium valproate, benzodiazepines, or carbamazepine, either as monotherapy or in combination.

Surgery by lumpectomy cures the epilepsy and is indicated when the tumor is large, causing seizures that become drug-resistant, although the potential for progression of DNETs is low (3, 21, 22,). The prognosis is generally excellent after surgical resection, with the disappearance of epileptic seizures (10).

4. Conclusion

DNETs are among the most frequent benign tumors in children with a low potential for progression; their main concern lies in their convulsive nature. Imaging also allows for preliminary diagnosis to differentiate them from gangliomas or astrocytomas. Surgical treatment of these tumors allows for a better post-operative prognosis through the almost total disappearance of epileptic seizures.

Compliance with ethical standards

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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