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Role of MRI in the Diagnosis of Dysembryoplastic Neuroepithelial Tumours (DNET): Two Case Reports

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Aims: Show the value of MRI in the diagnosis of DNET.
Presentation of Case: We report two cases of DNET, in two patients aged 17 and 14 years, with epilepsy resistant to medical treatment, with no neurological deficit on clinical examination. On MRI, the first case shows a temporal cortical lesion and the second a parietal lesion, with no mass effect or peri lesional edema.
Discussion and Conclusion: Dysembryoplastic neuroepithelial tumors (DNET) are benign brain tumors clinically revealed by partial seizures occurring in young patients under 20 years of age, with normal neurological examination. Imaging contributes to the diagnosis, it is a cortical sus-tentorial lesion, without mass effect or peri lesional edema. The temporal location is the most frequent, followed by the frontal location more rarely parietal and occipital. It is a generally stable tumor. Surgery is the only treatment for DNET, it allows epilepsy to be controlled in 85%, but confirmation remains histological.

Keywords: Epilepsy; MRI; DNET.

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1. INTRODUCTION

1.1 Introduction

Dysembryoplastic neuroepithelial tumours (DNETs) are very rare benign brain tumours, currently classified by the World Health Organization (WHO) as grade 1 mixed neuronal and neuronoglial tumours, [1,2] It is seen in children and young adolescents under 20 years of age, with a history of drug-resistant epilepsy, [3] the typical form is seen on MRI as a supratentorial, cortical, usually temporal lesion without mass effect or peri-lesional oedema [4]. The main differential diagnoses of DNET are gangliogliomas and oligodendrogliomas [1]. Surgery alone allows the control of epileptic seizures in 85% of cases [2]. We report two cases of DNET, in two young patients, with epilepsy resistant to medical treatment, without neurological deficits on clinical examination, through which we will focus on the radiological characteristics and the role of MRI in the diagnosis of these tumours.

2. PRESENTATION OF CASE

2.1 Case 1

Our first patient is 17 years old, followed for partial epilepsy. She presented with a recurrence of epileptic seizures that were becoming resistant to treatment. The clinical examination did not reveal any neurological deficit. She was referred by his neurologist for radiological evaluation exploration. A brain MRI was performed on a Siemens 1.5 Tesla machine. The following sequences were performed: sagittal T1-weighted sequence, axial T2-weighted sequence, axial Flair sequence, axial gradient echo sequence, and axial diffusion sequence with calculation of the Apparent Diffusion Coefficient (ADC) and 3D T1 sequence after injection of Gadolinium. It revealed a well-limited cortico-subcortical left temporal intra-axial lesion process appearing in T1-weighted hyposignal, heterogeneous T2-weighted hypersignal, and Flair, hyposignal on the diffusion sequence, with low ADC, absence of calcifications on the T2 guarded echo-sequence. There are two fleshy lesions T1-weighted hyposignal, T2-weighted hypersignal and diffusion with low annularly enhanced ADC after injection of Gadolinium, (Fig. 1). It measures 38x62x45 mm. The absence of a mass and peri-lesional oedema is noted, with an imprint on the scale of the temporal bone opposite. In view of these radiological images, DNET was evoked. The patient underwent surgery, which allowed control of her epileptic seizures. Anatomopathological examination confirmed the diagnosis of DNET (Fig. 2).

2.2 Case 2

Our second patient is 14 years old, with no particular pathological history. He consulted the emergency room for partial apyretic and drug-resistant seizures. The clinical examination was without abnormality. He was referred to us for a brain MRI. We performed the following sequences: sagittal T1-weighted sequence, axial T2-weighted sequence, coronal Flair sequence, axial gradient echo sequence, axial diffusion sequence with calculation of the Apparent Diffusion Coefficient (ADC) and 3D T1 and axial T1 sequences after injection of Gadolinium. It showed a left parietal cortico-cous cortical area, with T1-weighted hyposignal, heterogeneous T2-weighted hypersignal, surrounded by a Flair-weighted hypersignal border, without mass effect or peri-lesional oedema, this area is discretely hypointense in terms of diffusion signal, with an intermediate ADC, and is not enhanced after injection of gadolinium (Fig. 3). It measures 35.6x29 x 27 mm. The radiological images were in favour of DNET, which was confirmed histologically after surgical treatment.

3. DISCUSSION

Dysembryoplastic neuroepithelial tumours (DNETs) are very rare benign brain tumours belonging to the glial tumours, [5] (<1% of all brain tumours, <0.2% of neuroepithelial tumours in patients over 20 years of age) [6]. Discovered recently by Daumas-Duport et al, [7] it occurs before the second decade of life and manifests clinically as a drug-resistant partial epilepsy without neurological deficit [8]. Imaging contributes to the positive diagnosis. It is a cortical, supratentorial tumour, without mass effect or peri-lesional oedema [4]. The temporal lobe is the most frequent site (62%), followed by the frontal lobe (31%), [9] then the parietal and occipital lobes which are rarer (7%) [3]. Other much rarer locations have been described in recent years: the brain stem, cerebellum, caudate and thalamic nucleus [1,8]. However, focal enhancement may be noted (18%) [5]. Calcifications may be present, punctiform (37%) or nodular (63%), associated with an imprint on the adjacent bone table in 50% of cases [1,6]. As was the case in our first observation. The CT scan may be normal in 10% of cases, when the

tumour is isodense, it becomes difficult to specify its exact location and extension, hence the interest of MRI which allows a better characterisation of the tumour [5]. MRI allows three types of DNET to be distinguished: a pseudocystic or pseudopolycystic type, with T1-weighted hyposignal, T2-weighted hypersignal, without contrast after injection of gadolinium and without calcifications, a nodular or multi-nodular type with heterogeneous signal with the possibility of calcifications, annular or nodular contrast. And a dysplastic type with T1-weighted hyposignal, poorly limited with a blurred grey-white matter interface [10]. The MRI data of our patients allowed us to identify the nodular form in the first observation and the cystic form in the second.

Histologically DNETs form a specific glioneuronal component, glial nodules and cortical dysplasia, the degree of their association allows three forms to be distinguished: the simple form is composed solely of the specific glioneuronal component. The complex form is composed of the three basic components, and the non-specific form is marked by the absence of the glioneuronal component [5]. The simple and complex forms are shown on MRI as pseudo-cystic, the non-specific form as nodular or dysplastic [10]. The main differential diagnoses of DNET are gangliogliomas and oligodendrogliomas. The latter had similar radiological features to DNETs. However, they have some peculiarities that allow them to be differentiated. In particular, the preferential frontal location of oligodendrogliomas and the frequent calcifications and contrast enhancement of gangliogliomas. The combination of clinical and radiological signs and above all histology allows the diagnosis to be confirmed, which is essential, since the course of action is different, involving only surgery for DNETs, whereas adjuvant radiotherapy or chemotherapy may be necessary for the other tumours [1,3–5].

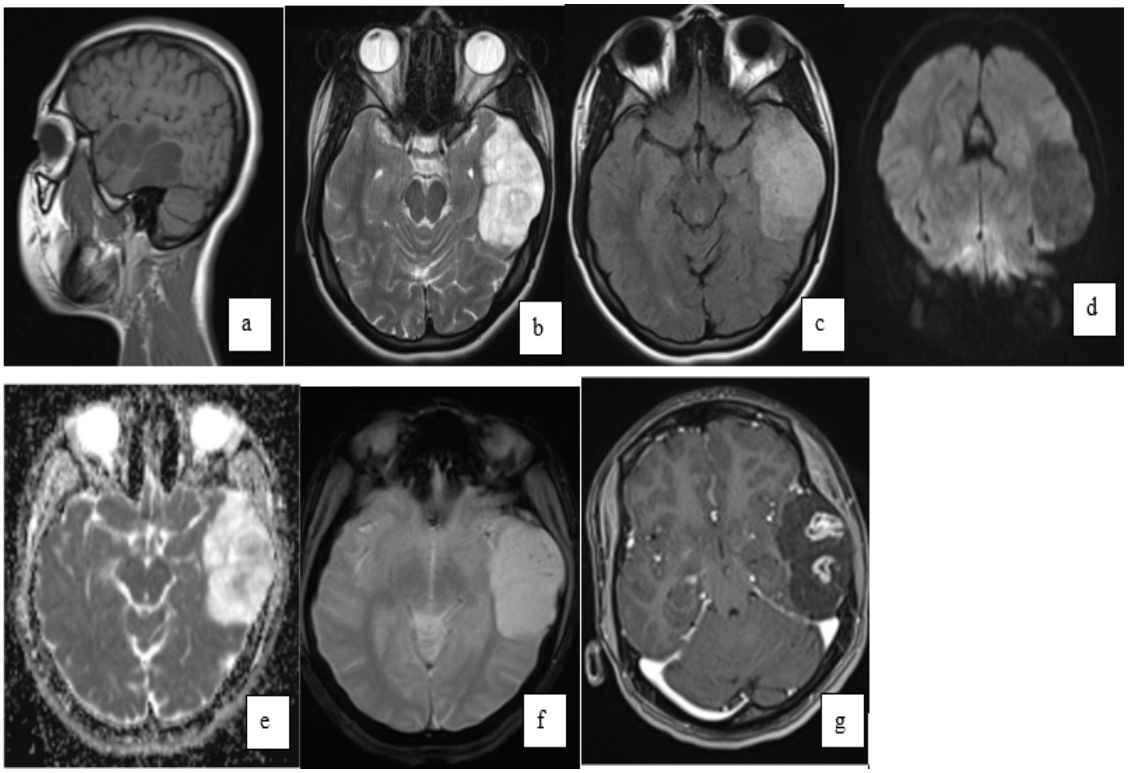

Fig. 1. Brain MRI in axial and sagittal section showing a hypo signal left temporal cortico-subcortical lesion with a fleshy area in T1-weighted hyposignal (a). The latter has a microcystic with the fleshy area in T2-weighted hypersignal (b) and Flair, with no mass effect or peri-lesional oedema (c), with hypersignal on the diffusion sequence(d) with low ADC (e) , Absence of a signal-free area on the gradient echo sequence indicates calcification or hemorrhage (f). On the 3D T1 sequence after injection of gadolinium, there is annular contrast of the two fleshy lesions (g)

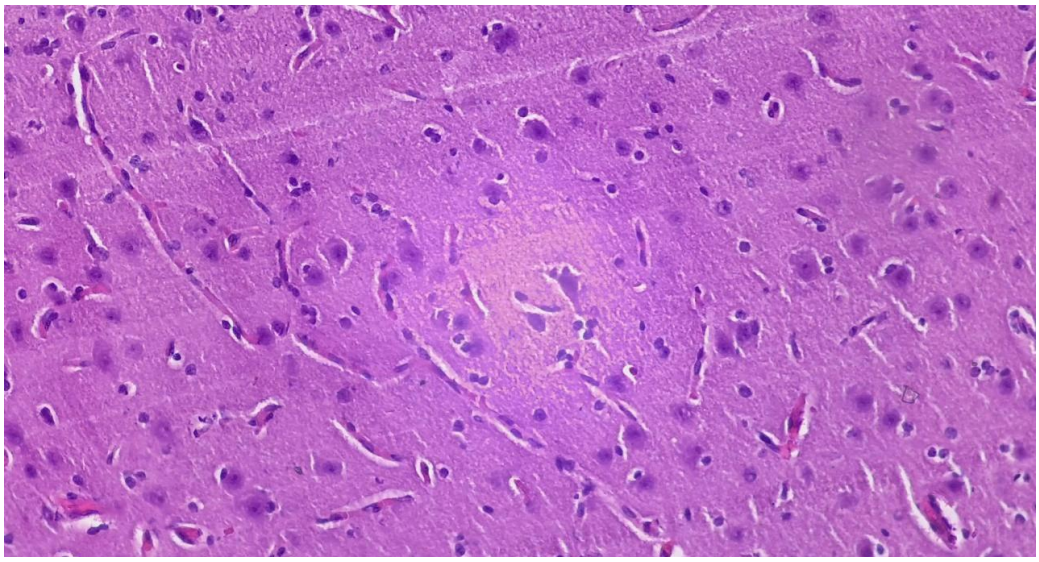

Fig. 2. Glial glial tissue, with a few regular looking neurons, arranged between the glial backgrounds which is luxoid, looking like "floating" neurons per place: morphological aspect of DNET

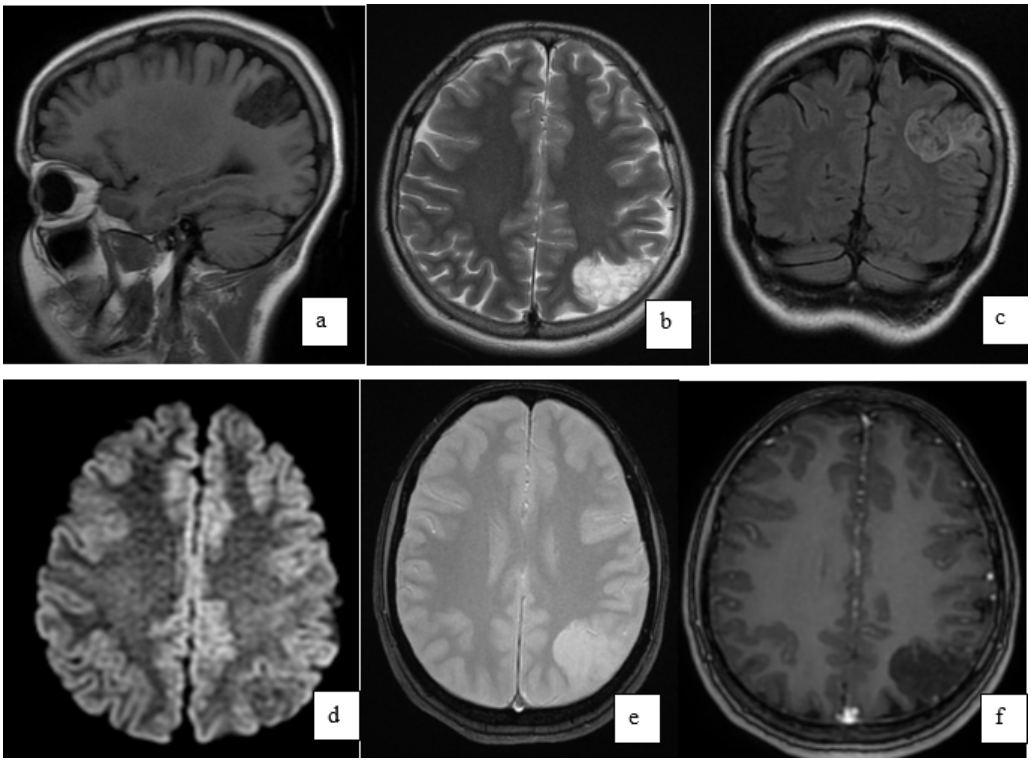

Fig. 3. Cerebral MRI in sagittal and axial section showing a cortico-subcortical, left parietal area in T1 hypo-signal (a), in T2 hyper signal (b), with no peri-lesional oedema effect, nor mass effect with the presence of a ring in hyper signal on the Flair coronal sequence (c). On the diffusion sequence, the latter is hypo-signal (d), with no visualization of calcification or stigmata of haemorrhage on the T2 gradient echo sequence (e).nor contrast on the 3D T1 sequence after injection of gadolinium

4. CONCLUSION

Modern imaging, including MRI, plays an important role in the preoperative diagnosis of DNET. The combination of clinico-radiological criteria in children and young adults, especially in the typical form, should allow early diagnosis and better management.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

ETHICAL APPROVAL

"All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki."

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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