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Case Report

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DYSEMBRYOPLASTIC NEUROEPITHELIAL TUMOR AND MEDICALLY INTRACTABLE SEIZURES: A CASE REPORT

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ABSTRACT

Dysembryoplastic Neuroepithelial Tumors (DNT) are a group of rare low-grade brain tumors, that were first described in 1988. DNTs are frequently associated to medically intractable seizures and are mainly reported in children and young adults. In current paper, the authors present the case of a 38 years old patient with a 20 years history of complex partial seizures with secondary generalization. Our patient underwent a craniotomy with complete resection of the tumor with a favorable outcome after surgery. The aim of our study was to better characterize the clinical-radiologic and pathologic spectrum of DNTs throughout a case report and show the importance of raising awareness about early diagnosis in our local context in Morocco.

INTRODUCTION

First described in 1988 by Daumas-Duport.[1] Dysembryoplastic Neuroepithelial Tumors (DNTs) are benign lesions that should be considered when all the following criteria are associated: partial seizures, with or without secondary generalization, beginning before the age of 20 years, no neurological deficit or stable congenital deficit, cortical topography of the lesion as better demonstrated by MRI and no mass effect on imaging.[2] We should highlight that the differentiation of DNT from oligodendroglioma is of crucial importance in order to avoid unnecessarily aggressive treatments.

CASE REPORT

A 38 years old woman, righthanded, with a medical history of complex partial secondary generalized temporal seizure since the age of 17. The patient used traditional medicines for many years without any beneficial outcome. Eventually, she was seen by a neurologist, put under tow high dose antiepileptics without any improvement. The medical examination reported a form of psychomotor retardation without any motor nor sensitive deficits. In April 2018, the first CT scan with iodinated contrast injection was performed and showed a hypo-attenuated cortical lesion on the left temporal lobe. On the preoperative MRI, the lesion appeared to be hypointense on T1 sequences and was of high signal intensity on T2 weighted images with cystic component and no tumor enhancement after Gadolinium injection nor peri-lesion edema.

The patient underwent a temporal craniotomy with macroscopic complete resection of the tumor. The histological examination revealed a tumor proliferation of diffuse and colonic organization on a fibrillary stroma. The tumor cell population consisted of three cell types: oligodendroglial-like cells mixed with dystrophic astrocytic cells and regular neurons. The Immunohistochemistry findings showed that the floating neurons expressed neuronal markers including: Synaptophysin neurofilament, neuron specific enolase, very low nuclear expression of Ki67 (1%) and GFAP-positive astrocytes confirming, thus, the diagnosis of complex type of DNT.

On the one-year follow up, the MRI scan didn't show any lesion anymore and the seizures have disappeared with a progressive regression of antiepileptics intake, however, we didn't report any improvement in the psychomotor retardation.

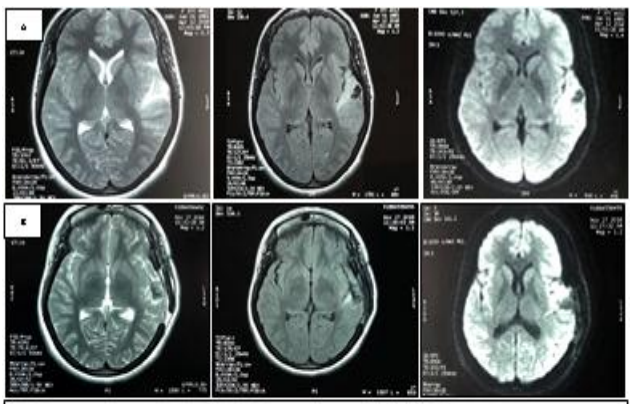

Figure 1: Preoperative (A) and postoperative (2) MR imaging, from left to right axial T2, T2 flair and diffusion sequences

DISCUSSION

DNTs accounts for 0.6 to 0.8% of brain tumors in children,[3,4] their diagnosis is based on a combination of clinical (partial epilepsy before age of 20, absence of neurological deficit), histological (glial tumor aspect) and radiological features (cortical lesion in general temporal without mass effect or perilesional edema). It's a benign group of tumors and the surgery alone allows recovery from epilepsy in 80% of cases.[5] Their differentiation from classic gliomas is crucial and helps to avoid the side effects of radiotherapy or excessive chemotherapy for young people with a normal life expectancy.

Partial complex seizures are the most common clinical manifestations, followed by generalized tonic-clonic, and partial seizures with secondary generalization, generally no neurologic deficit is found.[6]

This tumor was originally recognized in patients who underwent surgery for treatment of medically intractable seizures, however, recent progress in neuroimaging has allowed an increased detection of dysembryoplastic neuroepithelial tumors in patients with a single episode of epilepsy or in older patients. Recently, MRI features in histologic variants of DNTs were classified into three types as follows: type 1 which is a cystic/polycystic-like, well-delineated, strongly hypointense on T1 tumor, type 2 appears as a nodular- like, heterogeneous signal lesion, or type 3 more a dysplastic-like, isosignal/ hyposignal T1, poor delineation and gray-white matter blurring lesion.[7,8]

The original report in 1988 by Daumas-Duport et colleagues. described unique morphological features, including intracortical multi-nodularity, a specific glioneuronal element associated with cortical dysplasia all along with high expression of neuronal markers by the floating neurons including synaptophysin neurofilament, NeuN, neuron specific enolase (NSE) and GFAP-positive astrocytes.[1,9]

The differential diagnosis of DNT includes low-grade gliomas such as astrocytomas, oligodendrogliomas, oligoastrocytomas, gangliogliomas, and pleomorphic xanthoastrocytomas. The gangliogliomas are the most important differential diagnosis to be evoked mainly because of their frequent temporal location and their characteristics in imaging that are close to those of a DNT.

In general, DNTs are stable lesions, but complications like, intralesional hemorrhage,[11] necrosis or even malignant transformation,[12] were recorded in the literature. Surgical excision is the only treatment for DNTs, it allows a good control of seizures. Radiotherapy and post-operative chemotherapy have no evidence to be used especially in a population of children and young adults.

Despite that our patient presented all criteria for DNT, the diagnosis took years to be established. Therefore, this delayed diagnosis resulted in unusual neurological complications.

CONCLUSION

In the case of DNT-like cortical epileptogenic lesions, we need to combine clinical, neuroradiologic and pathologic data in order to achieve an accurate diagnosis, enhance the management of the patient and avoid unnecessary treatments. this case shows us that further efforts are needed, in our context, to change the cultural prejudices about epilepsy but also to generalize the multidisciplinary approach for such uncommon tumor in our hospitals.

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