Acta Neurol Belg (2013) 113:523–525
DOI 10.1007/s13760-012-0157-3

LETTER TO THE EDITOR

A cortical cystic epileptogenic lesion: tanycytic ependymoma

Luigi Rigante · Mariangela Novello · Luca Massimi · Massimo Caldarelli

Received: 24 September 2012 / Accepted: 31 October 2012 / Published online: 16 November 2012

Introduction

Ependymomas are neuroepithelial tumors usually found in the fourth ventricle, spinal cord and filum terminale, with higher incidence in children and young adults and two main histological types (classic cellular and myxopapillary). Tanycytic ependymoma is a rare variant with cystic appearance, occasionally found outside the ventricular system and usually presenting with seizures. Histopathological analysis is essential to make differential diagnosis with DNETs, astrocytomas and brain abscess, since treatment of choice is surgery followed by radiation for anaplastic variants, with favourable prognosis, low recurrence rate and seizures control.

Case report

A 14-year-old left-handed male patient presented with simple partial clonic seizures in left upper limb evolving to generalized, triggered by light stimulus and managed with Diazepam i.v. Neurological examination on admission was unaffected. Basal EEG showed bilateral asymmetric alpha rhythm waves, better organized on the left hemisphere. Carbamazepine (200 mg oral 2 times/day) was given with complete seizure control. Brain Magnetic Resonance Imaging (MRI) with gadolinium showed a cortical small cystic mass in the right temporo-parietal lobe, with fluid signal intensity on T2-weighted images and mild contrast enhancement of the cystic wall. Perfusion imaging showed no intra-/perilesional cerebral blood volume (CBV) increase (Fig. 1). MRI spectroscopy showed a lipid-lactate peak without increased cellular turnover. Functional MRI confirmed left hemisphere dominance (left/right laterality index = 0.33) of Broca's and Wernicke's areas and relative distance of the lesion from the right arcuate fasciculus (Electronic supplementary material, Fig. 1). Craniotomy and lesion removal was thus performed. Postoperative course was uneventful with complete seizure control with antiepileptic therapy at 12 months-follow up.

Histologic examination showed a neoplasm prevalently composed by elongated spindly bipolar cells with fascicular architecture and focal evidence of perivascular pseudorosettes, consistent with tanycytic ependymoma. True ependymal rosettes were not observed. The neoplastic cells showed immunolabeling with EMA, with diffuse or intracytoplasmic dot-or ring-like pattern, and GFAP. Proliferation index (Ki67) was about 1 %. Mitoses were less than 2 × 10 HPF and necrosis was not found (Fig. 2).

Discussion

Ependymomas are neuroepithelial tumors commonly found in the fourth ventricle and the cervico-thoracic segment of the spinal canal, affecting mostly children and young adults [1]. Two main histological types are recognized: classic cellular (WHO, grade II or III), localized in the brain or spinal cord, and myxopapillary ependymoma

Electronic supplementary material The online version of this article (doi:10.1007/s13760-012-0157-3) contains supplementary material, which is available to authorized users.

L. Rigante (✉) · L. Massimi · M. Caldarelli
Neurosurgery Institute, Catholic University School of Medicine, Largo Gemelli n. 8, 00168 Rome, Italy
e-mail: luigirigante@gmail.com

M. Novello
Pathological Anatomy Institute, Catholic University School of Medicine, Rome, Italy

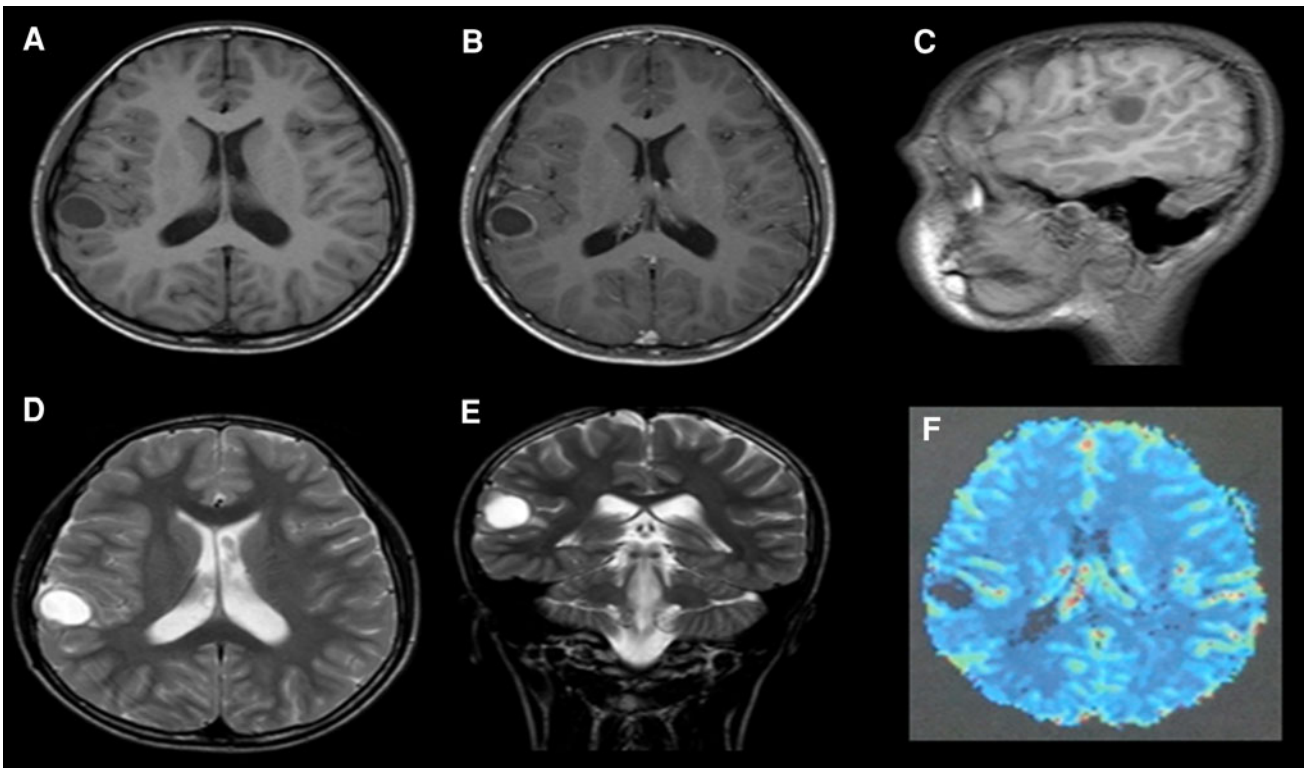

Fig. 1 Brain MRI with gadolinium showing a cortical cystic mass in the right temporo-parietal lobe, hypointense on T1-weighted sequences, with fluid signal intensity on T2-weighted images and mild contrast enhancement of the cystic wall, **a** axial T1-w, **b** axial T1-w with contrast, **c** sagittal T1-w, **d** axial T2-w, **e** coronal T2-w sequence, **f** perfusion MRI imaging displaying no intralesional cerebral blood volume increase

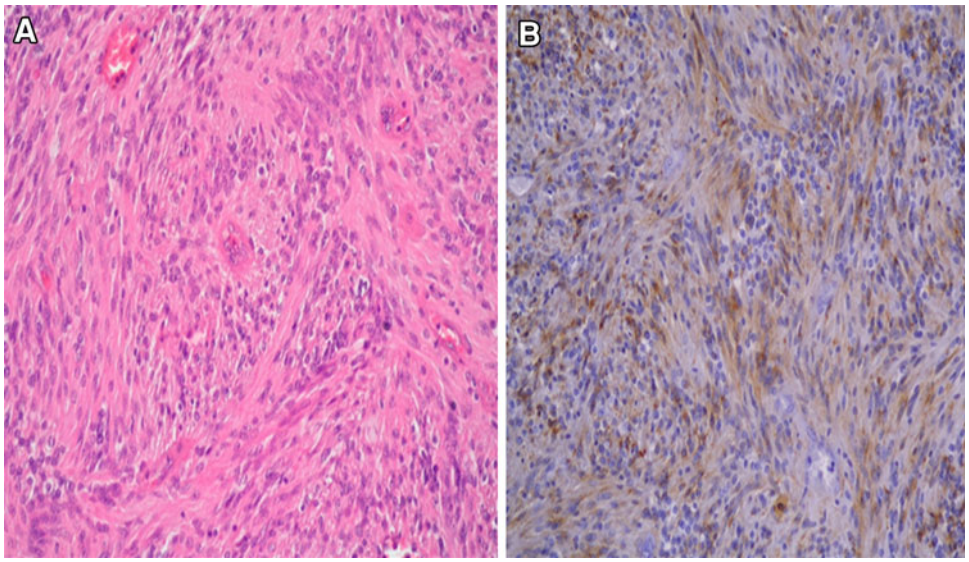

Fig. 2 Photomicrographs showing **a** fascicular pattern of spindle cells with perivascular arrangement (E&E, ×100), **b** EMA immunoreactivity in neoplastic cells (×100)

(WHO, grade I), usually found in the filum terminale. Tanycytic ependymoma is an unusual variant occurring in either the brain or more often in the spinal cord, characterized by spindled, elongated cells, reminiscent of a specialized form of ependymal cell [2]. Ultrastructural analysis may help detecting specific features of ependymal differentiation, such as junctional complexes between cells. Unfortunately, electron microscopy was not available for this case.

They can also be found outside the ventricular system in the central nervous system. Supratentorial cortical ependymomas are rare tumors (16 reported cases), usually

presenting with seizures [3]. They are located in the cerebral cortex and exhibit cystic or multicystic appearance with hypointensity on T1-weighted and hyperintensity on T2-weighted MRI sequences, low contrast enhancement mostly on the cystic wall (unlike brain abscesses) and without CBV increase on diffusion imaging. Spectroscopy determines their neoplastic nature (decrease in *N*-acetyl aspartate, increase in lactate, lipids and moderate increase of cholin, suggestive for low grade tumor, such as pilocytic astrocytoma). Like DNETs, cortical ependymomas have cortical location, present T1-weighted hypointensity and T2-weighted hyperintensity on MRI and lack increased cellular turnover. However, DNETs usually appear nodular and less cystic.

Recently, the angiocentric glioma, another low grade rare tumor presenting with epilepsy and showing signs of ependymal differentiation, has been added as a new entity to the WHO classification. Like tanycytic ependymoma, this is a non-enhancing lobar lesion with occasional overlying calvarial remodeling on MRI. Similarly, histopathological examination reveals bipolar spindle cells centered on cortical blood vessels, forming pseudorosettes with ependymomatous appearance and immunohistochemical staining shows GFAP positivity and "dot-like" cytoplasmic staining with the antibody against EMA [3].

Debate is ongoing as to whether cortical ependymoma represents a distinct entity or it is rather an example of misdiagnosed angiocentric glioma [3, 4].

Pathological analysis is essential to make diagnosis and ultrastructural analysis may indeed be helpful in the detection of specific ependymal features. Treatment of choice is surgery for low-grade tumors and resection followed by radiation for anaplastic ependymoma, with favourable prognosis, low recurrence rate and seizures control [3].

Conflict of interest None.

References

1. Massimino M, Buttarelli FR, Antonelli M, Gandola L, Modena P, Giangaspero F (2009) Intracranial ependymomas: factors affecting outcome. Future Oncol 5:207–216
2. Kawano N, Yagishita S, Oka H, Utsuki S, Kobayashi I, Suzuki S, Tachibana S, Fujii K (2001) Spinal tanycytic ependymomas. Acta Neuropathol 101(1):44–48
3. Van Gompel JJ, Koeller KK, Meyer FB, Marsh WR, Burger PC, Roncaroli F, Worrell GA, Giannini C (2011) Cortical ependymoma: an unusual epileptogenic lesion. J Neurosurg 114:1187–1194
4. Lehman NL (2008) Central nervous system tumors with ependymal features: a broadened spectrum of primarily ependymal differentiations? J Neuropathol Exp Neurol 67:177–188