

Soap Bubble Appearance: Dysembryoplastic Neuroepithelial Tumor “DNET”

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Abstract

Dysembryoplastic neuroepithelial tumors are rare benign brain tumors preferentially affecting epileptic children. The diagnosis is evoked by the association of clinico-radiological criteria: partial epilepsy beginning before the age of 20 years, absence of neurological deficit, cortical and temporal topography of the lesion and absence of mass effect or peri-lesional edema. The soap bubble appearance is a characteristic MRI sign of this tumor whose reference treatment is surgical excision allowing to palliate epileptic seizures in an efficient way.

Keywords: Neuroglial Tumour; Brain MRI; Soap Bubble

Introduction and Case Presentation

A 2-year-old male child from a well-monitored pregnancy, carried to term, with no particular history, who presented with complex partial epilepsy resistant to treatment. A brain MRI was performed showing a left temporal cortical lesion, multimicrocystic in T2 hypersignal, FLAIR hyposignal realizing the aspect of “soap bubbles”, with a peripheral hyperintense corona in FLAIR realizing the aspect of “Bright rim sign”, without enhancement after injection of PDC nor peri-lesional oedema or mass effect (Figure). A lesionectomy was performed by temporal flap, with simple operative follow-up without complications.

Discussion

DNETs are rare, slow-growing, benign neuroglial tumors. They affect the temporal lobe most often followed by the frontal lobe. Rarely, they may localize in the caudate nucleus, cerebellum and pontine. These tumors preferentially affect children with epilepsy. MRI shows a pseudocystic cortical mass, most often multimicrocystic giving a characteristic “bulla” appearance with thin septa isosignal to the cortex [1]. These lesions have the same signal as the cerebrospinal fluid with a T1 hyposignal, T2 hypersignal cancelled on FLAIR, without diffusion restriction, do not enhance after injection [1]. A particularly characteristic appearance on FLAIR is the bright rim sign [2].

The diagnosis of DNET should therefore be made when these clinico-radiological criteria are combined: partial epilepsy with onset of seizures before the age of 20, absence of neurological deficit, cortical and temporal topography of the lesion, and absence of perilesional edema or mass effect on the surrounding structures [3,4].

The differential diagnosis is mainly with ganglioglioma, multinodular and vacuolar neuronal tumor (MVNT), neuroepithelial cyst, cystic encephalomalacia, and Virchow robin space dilatation [1,2].

Surgical removal is the only treatment for these tumors, effectively palliating epileptic seizures [5].

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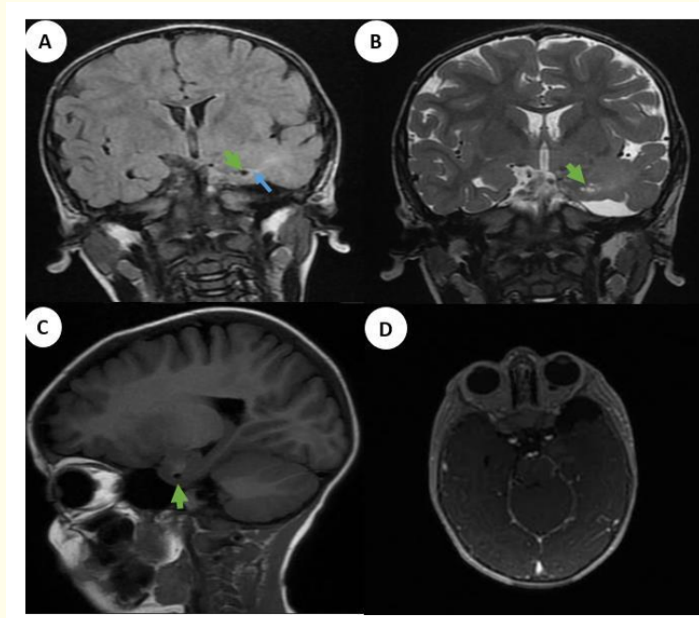


Figure: Brain MRI cross-sectional images: coronal FLAIR (A), coronal T2 (B), sagittal FLAIR (C) and axial T1 with PDC injection (D) showing a multimicrocystic aspect in T2 hypersignal, FLAIR hypointense realizing the “soap bubble” aspect (green arrow), with a hyperintense peripheral corona in FLAIR giving a “Bright rim sign” (blue arrow), without enhancement after PDC injection (D), nor peri-lesional edema or mass effect.

Conclusion

MRI is an effective imaging modality to make the diagnosis of dysembryoplastic neuroepithelial tumor in children with epilepsy. In this case, MRI demonstrated the soap bubble appearance of temporal topography consistent with a dysembryoplastic neuroepithelial tumor and the child underwent temporal flap lesionectomy with good clinical improvement. The diagnosis of DNETs, whose frequency is underestimated, is retained on a cluster of arguments and surgery can change the life of the patients.

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