de Monro sans autres signes associés, les antécédents ainsi que l’exploration endoscopique permettent de confirmer le diagnostic.

Conflit d’intérêt

Aucun.

Références


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MR perfusion of intracranial Rosai-Dorfman disease mimicking meningioma

IRM de perfusion d’une atteinte intra-cranienne de Rosai-Dorfman ressemblant à un méningiome

A 28-year-old woman presented with headaches and seizures for a few months. Physical examination disclosed no neurological deficit, nor any palpable extracranial lesion. Magnetic resonance (MR) imaging performed on a 3T magnet revealed a 4-cm, extra-axial, well-defined left frontal mass lesion that demonstrated T1 hypointensity, T2 isointensity, and enhanced intensely after gadolinium administration. It was surrounded by substantial vasogenic oedema. Dynamic susceptibility contrast (DSC) MR perfusion imaging showed an 8-folds increased relative cerebral blood volume (rCBV) in the lesion compared to the contralateral normal appearing whitematter, with a percentage of signal-intensity recovery of 89% (Fig. 1). Based on these criteria, pre-operative diagnosis was meningioma. Subsequently, complete resection was achieved.

On histological examination, there were nodules of histiocytes and significant cellular inflammatory infiltrate, without substantial elevation of mitotic activity. The histiocytes occasionally showed emperipolesis (lym- phocytophagocytosis). On immuno-histochemical analysis, histiocytes were positive for S100 protein and CD68, but negative for CD1a, thus strongly suggesting Rosai Dorf- man disease (RDD), and also positive for CD34 and CD31. RDD is a benign non-Langerhans histiocytic proliferative disorder, characterized by massive, painless, cervical lymphadenopathy accompanied by fever, weight loss and hypergammaglobulinemia. RDD also involves extranodal sites in about 40% of cases with a predilection for the skin, orbit, upper respiratory tract and bone marrow [1,2]. Central nervous system involvement is rare and an isolated intracranial RDD lesion even rarer. The majority of such cases appear as intracranial tumor-like lesions, solitary or multiple, presenting as dural-based extra-axial masses. They may involve the epidural or subdural space at the parasagittal convexity, the suprasellar area or the cerebel- lopontine angle, with a predilection for the skull base where they often erode the adjacent bone [2]. Imaging studies usually show a meningeval-based mass demonstrating iso- to hyperintensity on T2-weighted images and low signal intensity on T1-weighted images. The masses homogeneously enhance after gadolinium administration, thus mimicking lesions from a wide spectrum of dural-based masses, including meningioma [2,3].

DSC-MR perfusion is a technique that may help in distin- guishing meningiomas from dural metastases or lymphoma, by usually showing significant higher mean rCBV in the former [4]. On the other hand, overlapping rCBV values preclude differentiating meningiomas from other hypervascular dural lesions such as some metastases, hemangiopericy- tomas and solitary fibrous tumors of the meninges [5], or our case of meningeval RDD. Hyperperfusion was an unexpected finding because, in contrast with meningioma, blush at external carotid artery digital subtracted angiography is usually absent in meningeval RDD [3]. However, high positivity for CD34 and CD31 antibodies, that are surrogate markers of the intrinsic vascularization of lesions, may correspond to the high rCBV, as observed in meningiomas. Indeed, in menin- giomas demonstrating immunostaining with anti-CD31 and anti-CD34, significant correlation has been demonstrated between cerebral blood flow values, as assessed by perfusion MR imaging, and microvessels, as determined by the expression degree of CD31 and CD34 [6]. In conclusion, our case is the first to report the elevated rCBV value in meningeval RDD. Knowledge of this phenomenon may be of interest in the differential diagnosis of extra-axial mass lesions.
Figure 1  Axial FLAIR (A) and coronal T2 (B) images show a left frontal extra-axial lesion, isointense to the grey matter, with mass effect and surrounding edema. C. Axial T1 with gadolinium shows an important and homogenous enhancement. D. Axial map of rCBV reveals its hypervascular nature (with a maximal ratio of 8, relative to the contralateral normal appearing white matter). E. T2* signal-intensity time curve demonstrates high rCBV and percentage of signal-intensity recovery.

References