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Case report

Isolated cerebral Rosai–Dorfman disease presenting as a sole mass protruding into the fourth ventricle: A case report ☆☆☆

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ABSTRACT

Rosai–Dorfman disease is a non–Langherans cell histiocytosis typically revealed by a lymphadenopathy. Central nervous system involvement is rare, exceptionally isolated, and usually consists of dural masses mimicking meningioma. Very few reports have described non-dural-based lesions, especially with an intra-ventricular development. We report hereby the case of a Rosai–Dorfman disease in a 30-year-old man presenting as an isolated mass arising from the right cerebellar peduncle and protruding into the fourth ventricle. We provide the results of the MRI examination with a special focus on advanced MRI features. As the diagnosis relies on pathological examination, we also detail the results of the analysis that followed the surgical resection of the mass including the immunohistochemical profile. This report highlights the necessity to consider Rosai–Dorfman disease as a potential diagnosis in case of an infra-tentorial mass and/or intra-ventricular mass.

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Introduction

Rosai–Dorfman disease (RDD) is a rare non–Langherans cell histiocytosis typically revealed by a lymphadenopathy, mostly

cervical, in adolescents and young adults [1]. Extra-nodal disease is identified among 40 % of the patients [1,2] and can involve skin and soft tissues, oral cavity, genitourinary and respiratory tracts but also central nervous system [1].

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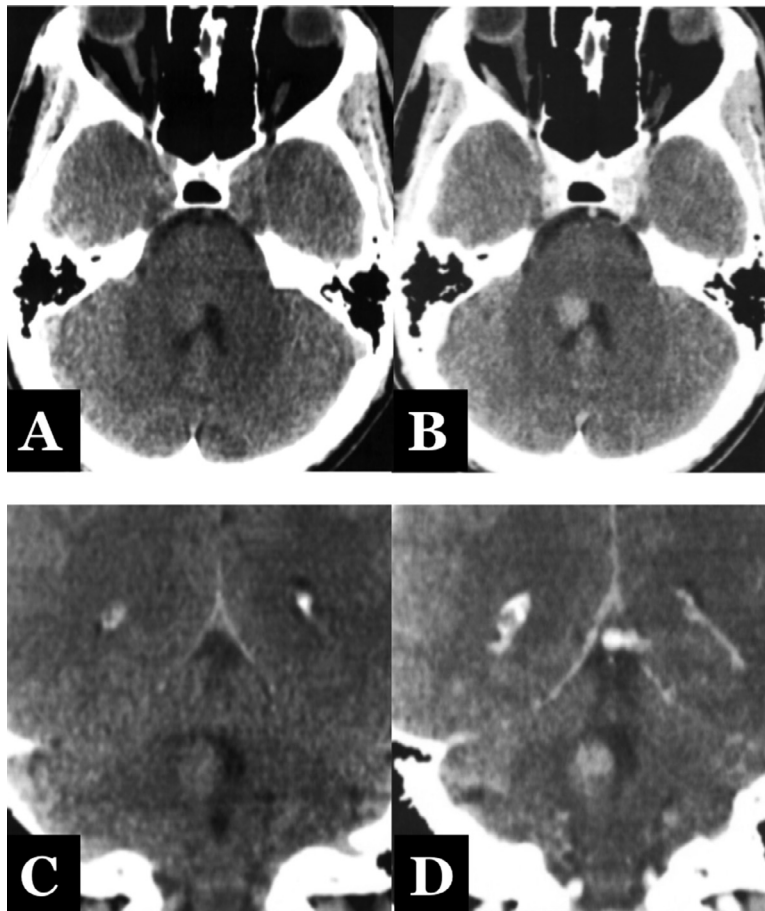


Fig. 1 – Computerized tomography Axial images: before (A) and after (B) contrast agent administration. Coronal images: before (C) and after (D) contrast agent administration.

Central nervous system Rosai-Dorfman Disease (CNS-RDD) occurs in 5 % of the cases [1,3,4] and is exceptionally isolated [3,4]. CNS-RDD typically consists of dural masses mimicking meningioma [5,6]. Non dural-based and intra-ventricular lesions have been rarely described [5,7,8] and exceptionally with perfusion-MRI and MR-spectrometry. We report here a case of isolated intra-ventricular CNS-RDD with a special focus on advanced MRI features.

Case report

Clinical presentation

We report the case of a Caucasian 30-year-old man, without any previous medical history, admitted in May 2019, who complained of vomiting, weight loss, dysphagia and vertigo for three weeks. Neurological examination found a nystagmus and a dysgeusia limited to the right half of the tongue.

Imaging

An encephalic computed tomography was performed (in a peripheral hospital immediately before the patient's transfer to

our institution), revealing a slightly hyperdense, enhancing, well-circumscribed, non-calcified mass protruding inside the fourth ventricle and measuring $12 \times 10 \times 14$ mm (Fig. 1).

MRI examination (Siemens Area 1,5 T) confirmed the presence and the location of this mass which appeared homogeneous, isointense to the cerebral cortex on T1-weighted images and slightly hyperintense on T2-weighted images (Figs. 2A and B). There was a homogeneous enhancement following intravenous administration of gadolinium (Dotarem) (Fig. 2C). Diffusion-weighted MRI showed a relative apparent diffusion coefficient (rADC) close to 1 (Fig. 2D). Perfusion-weighted MRI revealed no significant elevation of the relative cerebral blood volume or flow (rCBV / rCBF) (Fig. 3A) and the perfusion curve profile was suggestive of blood-brain barrier disruption (Fig. 3B). Magnetic resonance spectroscopy showed decreased NAA peak, slight lipid peak, and no alanine peak (Fig. 3C). Tumoral edges were regular and smooth. Minimal edema was visible in adjacent parenchyma. There was also no hydrocephalus.

Pathology

Surgical resection of the mass was performed. Microscopically, the lesion was mostly constituted of large macrophages having an eosinophilic or foamy cytoplasm with a large

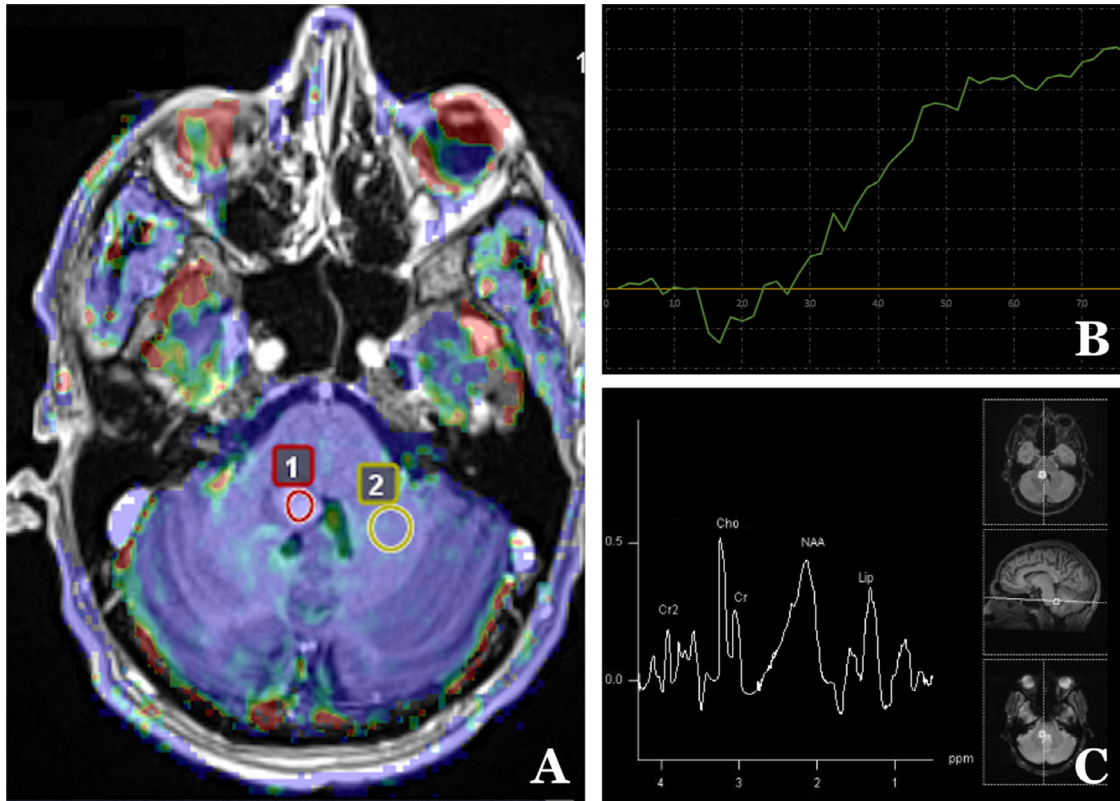


Fig. 3 – Advanced MRI examination.(A): axial fusion image consisting of cerebral blood volume cartography (rainbow scale) superimposed on enhanced-T1 weighted image (greyscale) showing no lesional hyperperfusion.(B): perfusion curve showing an overshooting of the baseline evocative of blood-brain barrier disruption (C): short echo time MR spectroscopy showing decreased NAA-peak, no alanine peak and a slight lipidic peak.

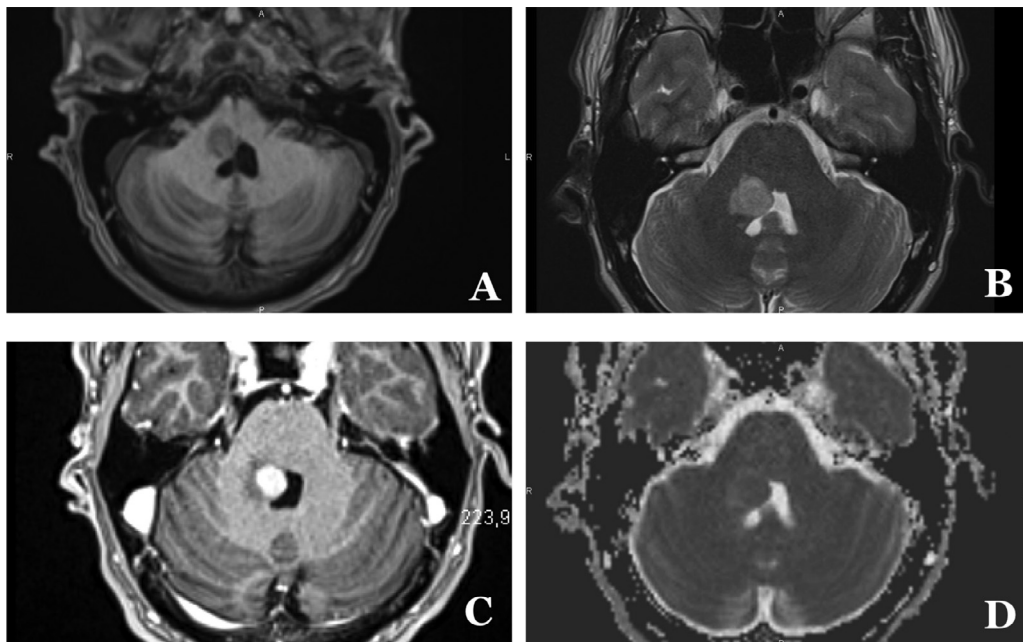


Fig. 2 – Conventional MRI examination Axial MRI examination showing the lesion protruding into the fourth ventricle. The lesion is iso-intense to grey matter on T1-weighted image (A), hyperintense with a slight adjacent edema on T2-weighted image (B) and homogeneously enhanced on T1-weighted image following administration of gadolinium (C) Apparent diffusion coefficient map (D) shows no abnormality.

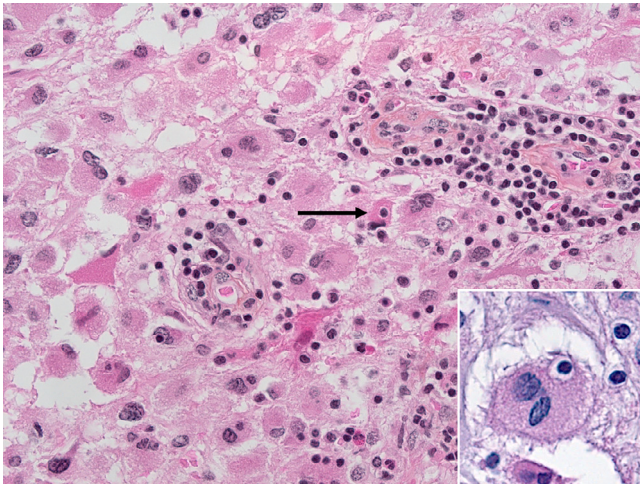


Fig. 4 – Pathological examination.Infiltration of the cerebral tissue by large eosinophilic or foamy macrophages; presence of an emperipolesis (arrow); Hemalum Phloxine - Inset: detail of emperipolesis (phagocytosis of a lymphocyte); Periodic Acid Schiff stain. Original magnification $\times 400$.

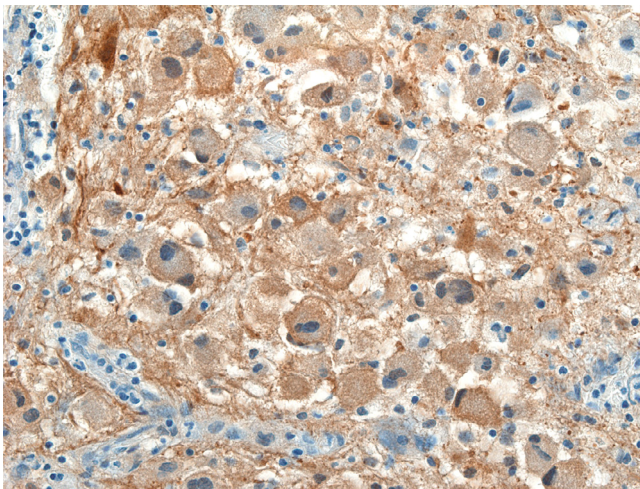


Fig. 5 – Immunohistochemistry.S100 protein expression by the macrophages. Original magnification $\times 400$.

vesicular nucleus (Fig. 4). Neovascularization with a dense peri-vascular lymphocytic infiltrate extending inside the histiocytic proliferation was also noted. Some macrophages had within their cytoplasm intact lymphocytes, designated as emperipolesis or lymphocytaphagocytosis, that was highly suggestive of a Rosai-Dorfman disease (Fig. 4). This hypothesis was confirmed by the immunohistochemical profile of the histiocytes: S-100 protein+ (Fig.5), CD68+, CD163+, CD1a- and IgG4-. Immunostaining for BRAFV600E was negative. Special stains (Ziehl-Neelsen, Gram, Grocott, Periodic Acid Schiff) did not reveal any underlying pathogenic agent.

Outcomes

No other localization of the Rosai Dorfman disease was found both clinically and radiologically. Thoracic, abdominal and pelvic CT scan as well as PET-CT were normal. Bone scintigraphy found no evidence of skeletal involvement.

After surgery, intra-venous corticotherapy was introduced (2 mg/kg per day), followed by a *per os* relay (1 mg/kg/j) in association with Cobimatenib (40 mg per day).

Discussion

Radiological features

Non-dural based lesions in CNS-RDD are very uncommon as demonstrated by the thorough review of the literature performed by Hong et al. who reported only 23 cases, whom only 8 concern the posterior fossa [8].

Ventricular involvement is exceptional. Jiang et al. [9] in a literature review of CNS-RDD locations identified 4 reports (over a total of 18 reports) of intraventricular involvement (none of them isolated) [2,10–12]. We identified only three supplementary reports [5,7,13], two of them consisting of an isolated disease, one arising from the pons and developing into the fourth ventricle [7] and the other one strictly located inside a lateral ventricle [5].

Reports concerning computerized tomography features of CNS-RDD are scant and mostly agree with our observation of a slightly hyperdense and enhancing lesion [8,14].

Conventional MRI features described in numerous reports summarized by the reviews of Hong and Jiang [8,9] are very concordant with our observation. Lesions are generally characterized as well-circumscribed and homogeneously enhanced after gadolinium administration. They tend to be iso- or slightly hypointense to cerebral cortex on T1-weighted images and iso- or slightly hyperintense on T2-weighted images.

As well, only a few reports described advanced MRI techniques in CNS-RDD lesions. Camp et al. [15] observed a non-specific choline peak in a non-dural based lesion. Doai et al. [16] and Symss et al. [17] both in case of dural-based mass found a lipid peak and no alanine peak, helping to question the diagnosis of meningioma. Perfusion-weighted images were acquired in non-dural based lesions by Camp et al. [15] as well as Varrassi et al. [14] who unlike us found a slightly elevated rCBV. Nevertheless, in a dural-based lesion, Hingwala et al. [18] found no elevation of rCBV leading to assume that the enhancement they observed was also related to a blood-brain barrier disruption. Diffusion-weighted images do not help to suspect the diagnosis as reports mention either - as in our case - no abnormality [10,11,14] or a restricted diffusion [14,18] in dural and non-dural based lesions.

Eventually, findings of advanced MRI techniques are not concordant and non-specific but could help to exclude differential diagnosis and especially meningioma.

Pathology

Radiological examination remains unable by itself to establish the definitive diagnosis of CNS-RDD which is brought by histopathological analysis [6]. CNS-RDD is characterized by a proliferation of histiocytes with a specific immunohistochemical profile: immunoreactive for S-100 protein and CD68 but negative for CD1a [4,6]. The presence of inflammatory cells with a predominance of lymphocytes is also suggestive [6]. In contrast to Langherans cell histiocytosis, eosinophils are rare. Engulfment of these inflammatory cells by the histiocytes, called emperipolesis, is the hallmark of RDD [4,6].

Conclusion

This observation of a rare and isolated location of CNS-RDD highlights the necessity to consider this disease as a potential diagnosis in case of an infra-tentorial mass and/or an intra-ventricular mass. Clinical manifestations of RDD should therefore be carefully searched during clinical examination.

MRI perfusion and spectroscopy are adjunct techniques that could be helpful to exclude differential diagnosis. Nevertheless, radiological examination remains unable by itself to establish the definitive diagnosis of CNS-RDD which is brought by histopathological analysis.

Patient consent

Written informed consent for publication of clinical details and images was obtained from the patient.

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