CASE REPORT

Advanced MRI in Rosai–Dorfman disease: Correlation with histopathology

Séquences IRM avancées dans la maladie de Rosai-Dorfman : corrélation avec les données histologiques

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Summary Rosai–Dorfman disease is an idiopathic benign lymphoproliferative disorder that can, on rare occasions, cause intracranial or intraspinal lesions with non-specific features on conventional imaging. For this reason, its diagnosis is based on the classical pathological findings of histiocyte proliferation and emperipolesis. In this case report, we describe the imaging features of Rosai–Dorfman disease as visualized by newer types of MRI sequences, such as diffusion tensor imaging (DTI), susceptibility-weighted imaging (SWI) and perfusion-weighted imaging (PWI). In fact, combining the findings of conventional cross-sectional imaging with high fractional anisotropy (FA), a low apparent diffusion coefficient (ADC), mild blooming on SWI and decreased perfusion can help to make the diagnosis of Rosai–Dorfman disease. These newer tools can also be used to clarify the pathology of Rosai–Dorfman disease.

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Introduction

Rosai–Dorfman disease (sinus histiocytosis with massive lymphadenopathy) is an idiopathic benign disorder characterized by an abnormal proliferation of histiocytes.

The characteristic clinical features of this rare but well-established disorder were first described by Rosai and Dorfman in 1969 as lymphadenopathy, fever and leucocytosis [1]. The most common presentation is bilateral painless cervical lymphadenopathy. Extranodal involvement may occur in 43% of cases, most commonly affecting the upper respiratory tract, paranasal sinuses, soft tissues of the head and neck, and skin [2].

Intracranial or intraspinal involvement is seen in only 4% of cases [3,4]. Due to the rarity of the disease, and the
multiple differential considerations and non-specific imaging findings from cross-sectional imaging techniques, such as computed tomography (CT) and conventional magnetic resonance imaging (MRI), the diagnosis is principally based on its characteristic pathological findings. However, more sophisticated MRI techniques such as diffusion tensor imaging (DTI), susceptibility-weighted imaging (SWI) and perfusion-weighted imaging (PWI) have a role to play in pointing towards the specific diagnosis of Rosai–Dorfman disease. In the present case report, the imaging findings of the disease using advanced imaging techniques are described, and an attempt is made to correlate these findings with the pathological features.

Case report

A 36-year-old male patient complained of right-sided facial pain over the past 18 months, with painless diplopia on looking to the right for the past 6 months, and progressively decreasing vision in the right eye, with complete vision loss and drooping of right eyelid, for the past 3 months.

On examination, there was pupillary asymmetry (the right larger than the left), with right lateral and up-gaze paresis (cranial nerve III and VI palsy). There was no perception of light in the right eye, and visual acuity in the left eye was 6/18. Corneal reflex on the right side was absent, with paresthesia in the right trigeminal nerve V1 and V2 dermatomes. No lymphadenopathy was present.

Routine hematological investigations revealed an increased erythrocyte sedimentation rate (62 mm/h), although the rest of the blood investigations were within normal limits.

CT revealed a hyperdense lesion in the right paracavernous region with moderate homogeneous contrast enhancement. MRI was performed with a 1.5-T clinical scanner (Avanto SQ-Engine; Siemens, Erlangen, Germany), using a 12-channel phased-array head coil. Routine MRI sequences showed a lobulated mass in the region of the right cavernous sinus. The mass was profoundly hypointense on T2-weighted (5580/110/9; TR/TE/echo train length) and FLAIR (8000/108/2500; TR/TE/TI) images, and isointense on T1-weighted (666/11/1; TR/TE/averages) images. There was also intense homogeneous post-contrast enhancement (Fig. 1). Although the right internal carotid artery (ICA) was encased by the mass, its diameter and flow void were unaffected. There was vasogenic edema of the adjacent right temporal lobe. The mass extended into the orbit via the superior orbital fissure and also posteriorly into the posterior cranial fossa.

On diffusion-weighted imaging (DWI; 3500/105/30; TR/TE/number of directions), the mass appeared hypointense. The apparent diffusion coefficient (ADC) values were $635.6 \times 10^{-6}$ mm$^2$/s, with a standard deviation (SD) of 222.6 within the mass, whereas the normal

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**Figure 1** A. Axial contrast-enhanced CT shows a homogeneously enhancing mass in the right cavernous sinus. B. Axial T1-weighted image shows that the lesion is isointense, and the flow void of the right internal carotid artery (ICA) is well maintained. C. On the axial T2-weighted image, the lesion is profoundly hypointense. D–F. Post-contrast T1-weighted images (axial, coronal, sagittal) show intense homogeneous enhancement.
Table 1 MRI parameters within the lesion compared with those in the normal contralateral side.

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Means</th>
<th>SD</th>
<th>Asymmetry index</th>
</tr>
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<tbody>
<tr>
<td>ADC</td>
<td>635.5</td>
<td>222.6</td>
<td>0.816</td>
</tr>
<tr>
<td>b0</td>
<td>126.5</td>
<td>41.1</td>
<td>3.078</td>
</tr>
<tr>
<td>FA</td>
<td>541.1</td>
<td>202.2</td>
<td>1.048</td>
</tr>
<tr>
<td>RA</td>
<td>530.6</td>
<td>259.3</td>
<td>1.695</td>
</tr>
<tr>
<td>rCBV</td>
<td>48.6</td>
<td>50.9</td>
<td>0.354</td>
</tr>
<tr>
<td>rCBF</td>
<td>105.5</td>
<td>60.8</td>
<td>0.878</td>
</tr>
</tbody>
</table>

ADC: Apparent diffusion coefficient × 10⁻⁶ mm²/s; FA: Fractional anisotropy; RA: Relative anisotropy; rCBV: relative cerebral blood volume; rCBF: relative cerebral blood flow

Contralateral brain parenchyma had an ADC value of 778.7 × 10⁻⁶ mm²/s (SD: 137.4; Table 1). The fractional anisotropy (FA) was significantly increased (541.1; SD: 202.2) compared with the normal contralateral parenchyma (516.2; SD: 145.3). The mass showed hypointensity on SWI (48/40/20; TR/TE/flip angle), and focal areas of blooming were noted at the periphery of the mass. Dynamic susceptibility T2*-weighted contrast perfusion MRI (1770/43/50; TR/TE/measurements) showed that the mass was hypoperfused, with a decrease in relative cerebral blood volume (rCBV) and relative cerebral blood flow (rCBF) compared with the normal contralateral parenchyma (Fig. 2).

The patient underwent a right frontotemporal craniotomy and decompression of the mass lesion. In the immediate postoperative period, he developed a motor deficit of the trigeminal nerve, which was slow to recover. Vision in the right eye improved, but the right extraocular muscle paresis and V1—V2 paresthesia persisted.

Histopathology of the mass revealed diffusely scattered lymphonuclear and plasma cells with sheets of large, pale, foamy-looking histiocytes and foci of emperiplois. These cells were supported by a fibrovascular connective tissue (Fig. 3). Round, refractile, eosinophilic Russell bodies were also seen, and there was strong immunopositivity for S-100

Figure 2 A. DWI reveals the hypointense lesion. B. The ADC map shows a reduction in the diffusion coefficient within the lesion (red circles indicate the areas where various diffusion parameters were measured). C. The FA map shows elevated values. D. SWI shows diffuse, mild, hypointense blooming in the lesion. E. Dynamic contrast-enhanced PWI shows no significant increase in rCBV.
Figure 3  A. Hematoxylin and eosin staining of a histological section (× 400) reveals an inflammatory infiltrate composed of lymphocytes, plasma cells and histiocytes. The characteristic emperipolesis (lymphophagocytosis) is also seen in histiocytes (arrow). B. Hematoxylin and eosin staining of a histological section (× 100) shows extensive areas of fibrosis between the inflammatory cells. C. The histiocytes demonstrate strong immunopositivity for S-100 protein (× 400).

protein. These findings were suggestive of Rosai–Dorfman disease.

Discussion

Isolated intracranial Rosai–Dorfman disease without lymphadenopathy is unusual [4]. The typical imaging features described in the literature include a dura-based mass that is hyperdense on plain CT, isointense or mildly hypointense on T1-weighted MRI and profoundly hypointense on T2-weighted images [2], with significant post-contrast enhancement. These findings overlap with other conditions such as meningiomas [5], other meningeal tumors such as solitary fibrous tumor of the meninges, lymphoproliferative disorders [6], and infectious and non-infectious granulomas [7]. Nevertheless, the advanced neuroimaging techniques of DTI, PWI and SWI all proved helpful in the differentiation of Rosai–Dorfman disease from the above-mentioned pathologies.

The definitive diagnosis of Rosai–Dorfman disease is based on its distinctive histopathological appearances [8] and immunohistochemistry [9]. The disease is characterized by abundant sheets of large and medium-sized vacuolated histiocytes in a fibrous stroma, interspersed with foci of chronic inflammatory cells. The high FA value found in the disease is probably due to the structured arrangement of the dense fibrous tissue. A high FA has been reported in tumors with a high fibrous content such as fibroblastic meningioma [9]. The hypointensity seen on DWI may be the result of a dominant T2 effect ("T2 blackout" phenomenon). Interestingly, the mass had low ADC values but, again, this may be explained by the presence of the dense fibrous tissue within the mass, as it may be assumed that the parallel layers of fibrous stroma within the mass limit the diffusion of water. Similar findings are also seen in other pathologies, such as granulomas [7] and fibroblastic meningiomas, which involve organized fibrous structures.

Hypointensities on SWI in granulomatous lesions of the brain have already been described [7]. The diffuse mild blooming noted on SWI within the lesion are most likely due to the free-radical or mineral deposition (manganese and non-heme iron) seen with chronic inflammation [10]; it is usually not caused by calcification, which is never seen in dural cases of Rosai–Dorfman disease. The disease has been shown to decrease perfusion, and asymmetry indices for rCBV and rCBF (abnormal side/normal side) show that these parameters are decreased on the side of the lesion.

Rosai–Dorfman disease is essentially a lymphoproliferative disorder with no neoangiogenesis, as evidenced by histopathological findings, and that feature is important in differentiating the disease from meningiomas, hemangiopericytomas and solitary fibrous tumors of the meninges, which typically have markedly increased perfusion parameters [2,11]. However, a recent report has shown that Rosai–Dorfman disease may also show increased perfusion due to its high positivity for CD34 and CD31 antibodies, which are surrogate markers for the intrinsic vascularization of lesions [11].

Thus, combining the findings of conventional cross-sectional imaging with high FA, low ADC, mild blooming on SWI and decreased perfusion can help to make the diagnosis of Rosai–Dorfman disease.
Conclusion

This case report briefly outlines the findings for Rosai–Dorfman disease using the advanced imaging techniques of DTI, SWI and perfusion MRI, all of which can substantiate the findings of routine MRI sequences. To the best of our knowledge, such findings have never been previously described in the literature. Thus, these newer imaging tools can be used to further elucidate the pathology of Rosai–Dorfman disease.

Conflict of Interest statement

Nothing declared.

References