Received: 7 February 2022 | Accepted: 25 March 2022

DOI: 10.1002/ccr3.5737

Clinical Case Reports Open Access WILEY

CASE REPORT

Differentiation of intracranial Rosai-Dorfman histiocytosis from meningioma using MR perfusion

Hugo F. Bueno[1] | **Pankaj K. Agarwalla**[2] | **Anupama Chundury**[3] | **Ada Baisre de Leon**[4] | **Neena Mirani**[4] | **Esther A. Nimchinsky**[1]

[1]Department of Radiology, Rutgers University-New Jersey Medical School, Newark, New Jersey, USA

[2]Department of Neurosurgery, Rutgers University-New Jersey Medical School, Newark, New Jersey, USA

[3]Department of Radiation Oncology, Cancer Institute of New Jersey, New Brunswick, New Jersey, USA

[4]Department of Pathology, Rutgers University-New Jersey Medical School, Newark, New Jersey, USA

Correspondence
Esther A. Nimchinsky, Department of Radiology, Rutgers University-New Jersey Medical School, 180 South Orange Ave, MSB F506, Newark, NJ 07103, USA.
Email: nimchins@njms.rutgers.edu

Funding information
None

Abstract

Intracranial Rosai-Dorfman disease may be indistinguishable from meningioma. This distinction is essential, as they are treated very differently. We present two cases where perfusion imaging helped make this distinction, allowing one to be treated successfully without craniotomy. Perfusion imaging may be a powerful adjunct in cases where RDD mimics meningioma.

KEYWORDS

histiocytosis, meningioma, MR perfusion, Rosai-Dorfman disease

1 | INTRODUCTION

Rosai-Dorfman disease (RDD) or sinus histiocytosis with massive lymphadenopathy (SHML) is an uncommon histiocytic disorder, presenting most commonly as bilateral painless cervical lymphadenopathy.[1] Extranodal disease may occur in the absence of nodal disease and may involve virtually all other organ systems, with nasal cavity involvement in 11% of cases and CNS involvement in 4%–5%.[1] Intracranial RDD also has a variable presentation, the most common being a well-circumscribed, dural-based lesion.[2,3] This poses a diagnostic dilemma as it may be indistinguishable from meningioma on conventional imaging, and as a result, RDD is often managed surgically, sometimes unnecessarily. We describe two cases of intracranial RDD where MR perfusion characteristics differentiated the lesion from meningioma, avoiding craniotomy in one case.

2 | CASE 1

A 50-year-old man with a history of RDD presented with a left eyelid lesion. MRI showed bilateral nasal cavity lesions and an avidly enhancing dural-based mass over the left frontal lobe, which, based on its MR appearance, was

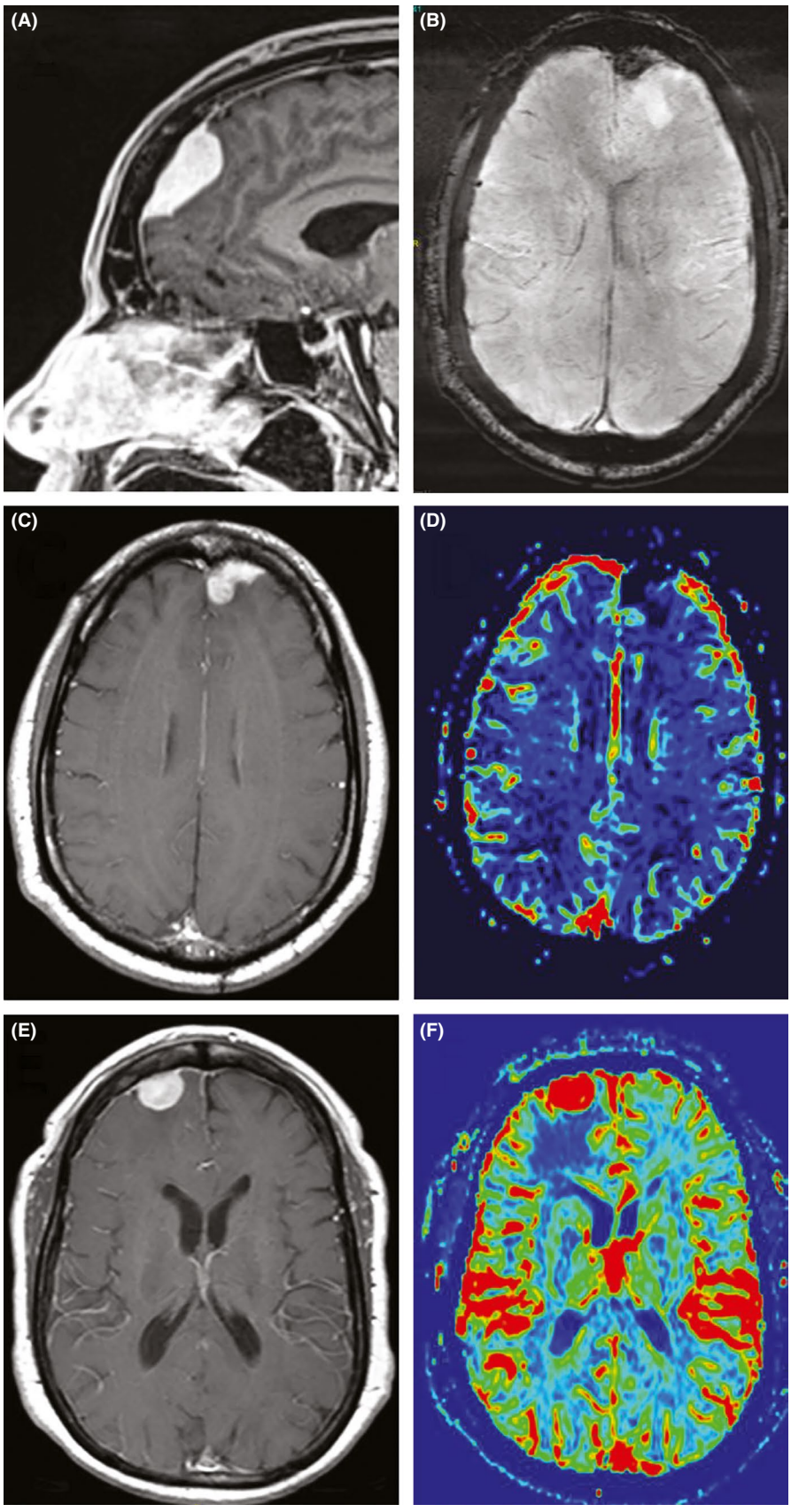

FIGURE 1 Case 1. 50-year-old man with history of RDD. (A) Sagittal postcontrast image showing a dural-based enhancing extra-axial mass with a dural tail. (B) Axial SWI demonstrates hypointense signal at the site of the lesion, suggesting mineralization, typically seen with meningiomas. (C) Axial postcontrast image again shows the enhancing mass which demonstrates (D) decreased relative cerebral blood volume (rCBV) compared to surrounding tissue. In contrast, (E) a typical meningioma demonstrates (F) markedly increased rCBV

interpreted as a meningioma (Figure 1A–C). MR perfusion, however, showed decreased perfusion (Figure 1D), rather than the increased perfusion typically seen with meningioma (Figure 1E–F). The mass was resected, and pathology showed a histiocytic lesion, composed of S-100 and CD68-expressing histiocytes, abundant plasma cells, and lymphocytes. Focal emperipolesis was identified. B-cell-rich lymphoid clusters were seen and the percentage of IgG plasma cells positive for IgG-4 was <10%. These findings confirmed Rosai-Dorfman disease. The patient has since been lost to follow-up.

3 | CASE 2

A 35-year-old-man without past medical history presented to the emergency department complaining of vision loss and chronic nasal congestion. MRI revealed a mass filling the nasal cavity, eroding the middle turbinates, and extending through the paranasal sinuses (Figure 2A). Initial biopsy demonstrated nonspecific inflammation. To obtain a more definitive diagnosis and address his nasal symptoms, the intranasal component was resected. Pathology showed densely fibrotic nodules infiltrated by abundant inflammatory cells and focal emperipolesis (Figure 2B). The inflammatory infiltrate was composed of CD68-expressing histiocytes against a background of abundant CD20-positive B cells and CD3-positive T cells. Several large cells expressed S-100 protein. IgG-4 cells were also present. KRAS- and BRAF-activating mutation were negative. These findings confirmed a diagnosis of RDD. In addition, he had a dural-based mass at the right planum sphenoidale, encasing the distal optic nerve, impinging on the optic chiasm, and encasing and narrowing the supraclinoid segment of the right internal carotid artery (Figure 2C, E). Conventional angiogram did not demonstrate the typical contrast blush seen with meningiomas. Because of his age, gender, and coexistent nasal mass, it was suspected that the intracranial mass might also represent RDD, and MR perfusion was performed. The mass demonstrated decreased perfusion (Figure 2D) supporting the suspicion that this represented RDD.

Given the lesion location, the decision was made to start treatment with steroids instead of surgical resection. Following five weeks of treatment with dexamethasone, the mass decreased in volume by approximately 50%, and the mass effect on the optic chiasm as well as vasogenic edema in the right frontal lobe resolved. Dexamethasone was continued for approximately 15 weeks and then switched to prednisone taper. The patient experienced side effects from the steroid treatment, but further taper was delayed due to a small increase in tumor volume. Therefore, a decision was made to change the modality of treatment, and radiation therapy was initiated. Tumor volume decreased by half at the first follow-up (9 weeks after completing radiation therapy) with further halving at the second follow-up (32 weeks after completion of radiation). Eighteen months after presentation, tumor volume was 16% of its original value, and the patient's headaches, as well as vision loss, had improved significantly without surgical resection (Figure 3).

4 | DISCUSSION

RDD was originally described by Rosai and Dorfman as sinus histiocytosis with massive lymphadenopathy.[4] It usually presents in young or middle-aged men with painless adenopathy, but extranodal disease may involve virtually all other organ systems.[1] Isolated intracranial RDD is seen in fewer than 5% of cases, often mimicking a meningioma. A recent review of the literature (285 cases) noted patients with CNS involvement present with exclusive intracranial involvement in 77% of cases, exclusive spinal involvement in 14% of cases and a 1.9:1 male to female ratio.[5] In one of the largest intracranial RDD series, consisting of 10 cases, marked enhancement, perilesional edema, and a dural tail sign, all features that may be associated with meningioma, were seen in all cases.[3] This distinction may be particularly important in cases where a surgical approach may result in significant morbidity. Both cases we encountered demonstrated decreased perfusion, unlike meningiomas, which typically demonstrate increased perfusion.

Intracranial RDD has been described as a hypovascular tumor with minimal to no neoangiogenesis.[6] Findings of digital subtraction angiography (DSA) have been conflicting, with lesions described variably as hypovascular[7] or hypervascular,[8] although this modality precludes quantification.

As noted previously, both our cases demonstrated hypoperfusion on MR perfusion, leading to the correct diagnosis. Although one previous report suggested hypervascularity of intracranial RDD on MR perfusion,[9] this may be accounted for by differences in technique. In that instance, perfusion was assessed in comparison with contralateral white matter, which normally shows decreased perfusion relative to gray matter. In fact, in that case, relative cerebral blood volume (rCBV) in the lesion appeared to be comparable to adjacent gray matter. This also differs somewhat from our case, where rCBV in RDD was lower than adjacent gray matter. Despite this reported case of at least mild hypervascularity, perfusion imaging still appears useful in differentiation, given that even in that case, rCBV did not reach the very high levels seen in

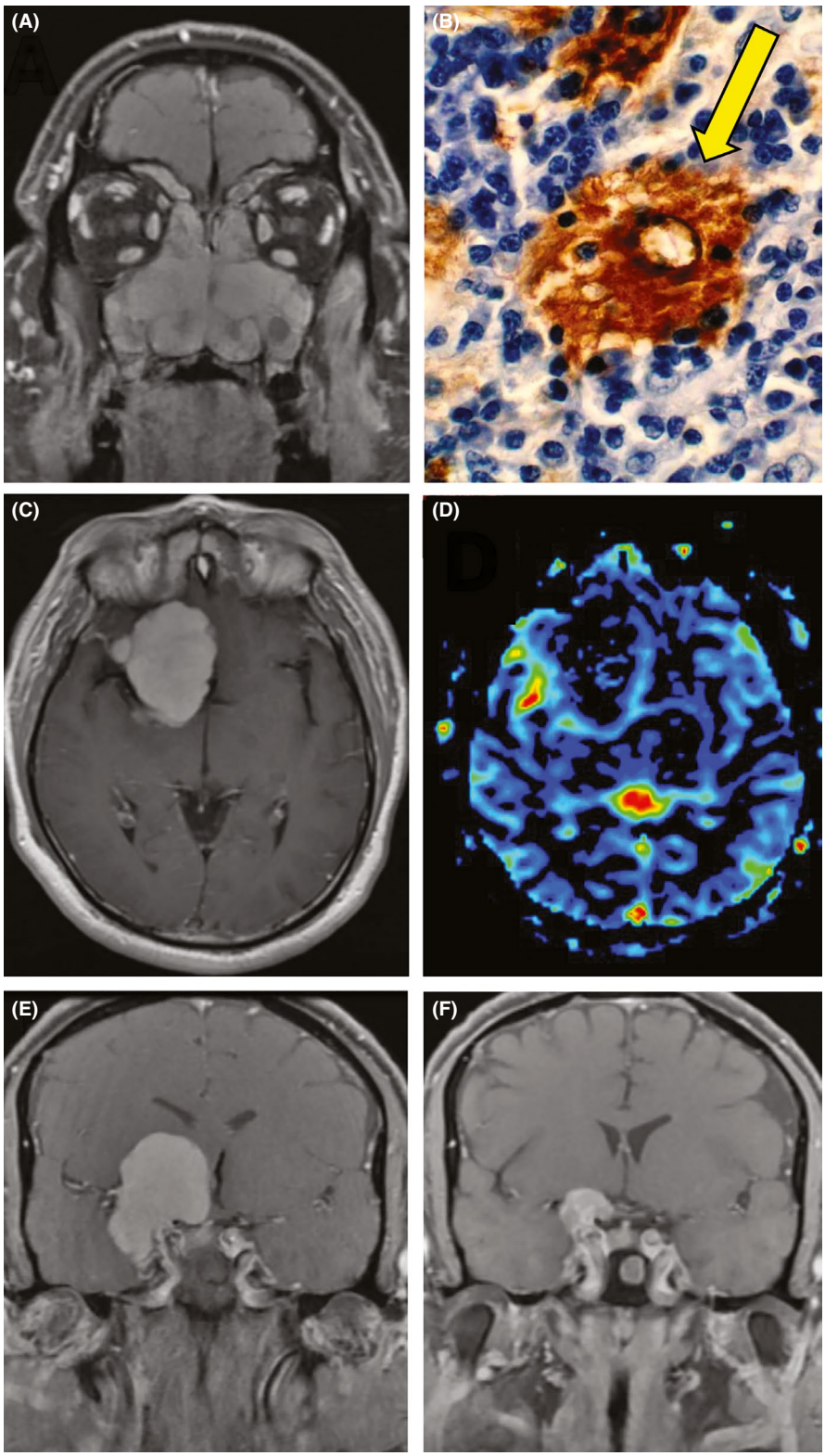

FIGURE 2 Case 2. (A) Coronal image through the paranasal sinuses demonstrates a mass filling the sinonasal cavity. (B) Pathologic specimen of the sinonasal mass with S-100 immunostain demonstrates emperipolesis with intracytoplasmic lymphocytes, compatible with RDD. (C) Axial postcontrast image through the mass demonstrates avid enhancement, and (D) MR perfusion shows decreased rCBV. (E) Coronal postcontrast image through the frontal lobes demonstrates an avidly enhancing, dural-based mass encasing the supraclinoid segment of the internal carotid artery. (F) Repeat imaging at 18 months demonstrates marked decrease in volume of the mass after treatment

meningiomas. Our findings are also strengthened by a recent report of two cases where MRI rCBV was decreased.[10]

According to consensus guidelines published in 2018,[11] therapeutic strategies are tailored on an individual basis and may involve surgery, steroids, and radiotherapy. In our case, management for patient 2 demonstrated a favorable outcome at 18-month follow-up and has thus far obviated the need for craniotomy.

MR perfusion may be a useful, noninvasive adjunct to traditional MRI for the workup of dural-based masses where, because of history, other findings, or demographics, the diagnosis of meningioma is questionable, and

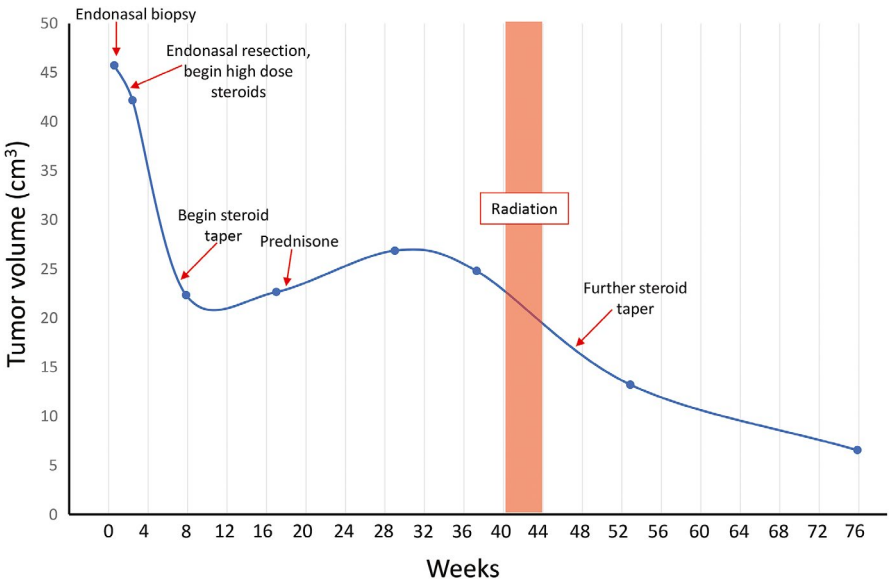

FIGURE 3 Case 2 tumor volume and response to therapy over time. This graph demonstrates tumor volume as measured in cm^3 and changes in time with red arrows noting points where therapeutic changes were initiated or made. The red block between weeks 40 and 44 denotes the time where radiation therapy was administered

may ultimately prove a useful tool to guide therapy in the absence of tissue sampling. To our knowledge, this is the first reported case where the perfusion characteristics of a mass in RDD were exploited to avoid surgical resection with a positive outcome, and propose that further use of MR perfusion may strengthen our ability to distinguish between these entities and spare potentially morbid surgery.

ACKNOWLEDGMENTS

We would like to acknowledge Dr Natalya Soloveychik for her assistance in providing clinical details of one case.

CONFLICT OF INTEREST

The authors have no personal, financial, or institutional disclosures in relation to this article.

AUTHOR CONTRIBUTIONS

HB analyzed data, wrote manuscript, and provide intellectual contribution. PKA provided intellectual contribution, clinical assessment, critically reviewed the manuscript. AC involved in intellectual contribution, clinical assessment, and critical review of manuscript. ABdL involved in pathologic analysis, intellectual contribution, and critical review of manuscript. NM involved in pathologic analysis, figure contribution, and critical review of manuscript. EN conceived project, wrote manuscript, analyzed data, and generated figures.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ORCID

Esther A. Nimchinsky https://orcid.org/0000-0002-3440-5220

REFERENCES

1. Foucar E, Rosai J, Dorfman R. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): review of the entity. *Semin Diagn Pathol*. 1990;7:19-73.
2. Andriko J-AW, Morrison A, Colegial CH, Davis BJ, Jones RV. Rosai-Dorfman disease isolated to the central nervous system: a report of 11 cases. *Mod Pathol*. 2001;14:172-178.
3. Zhu H, Qiu L-H, Dou Y-F, et al. Imaging characteristics of Rosai-Dorfman disease in the central nervous system. *Eur J Radiol*. 2012;81:1265-1272.
4. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. *Arch Pathol*. 1969;87:63-70.
5. Tatit RT, Raffa P, de Almeida Motta GC, et al. Rosai-Dorfman disease mimicking images of meningiomas: two case reports and literature review. *Surg Neurol Int*. 2021;12:292.
6. Hingwala D, Neelima R, Kesavadas C, Thomas B, Kapilamoorthy TR, Radhakrishnan VV. Advanced MRI in Rosai-Dorfman disease: correlation with histopathology. *J Neuroradiol*. 2011;38:113-117.
7. Huang H-Y, Huang C-C, Lui C-C, Chen H-J, Chen W-J. Isolated intracranial Rosai-Dorfman disease: case report and literature review. *Pathol Int*. 1998;48:396-402.
8. Konishi E, Ibayashi N, Yamamoto S, Scheithauer BW. Isolated intracranial Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy). *Am J Neuroradiol*. 2003;24:515-518.

9. Idir I, Cuvinciuc V, Uro-Coste E, et al. MR perfusion of intracranial Rosai-Dorfman disease mimicking meningioma. *J Neuroradiol*. 2011;38:133-134.
10. Safi SS, Murshed K, Ali A, Vattoth S, Haider A, Al-Bozom I. Rosai-Dorfman disease of cranial and spinal origin - A case series. *Surg Neurol Int*. 2020;11:298.
11. Abla O, Jacobsen E, Picarsic J, et al. Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood*. 2018;131:2877-2890.

How to cite this article: Bueno HF, Agarwalla PK, Chundury A, Baisre de Leon A, Mirani N, Nimchinsky EA. Differentiation of intracranial Rosai-Dorfman histiocytosis from meningioma using MR perfusion. *Clin Case Rep*. 2022;10:e05737. doi:10.1002/ccr3.5737