Received: 14 November 2023 | Revised: 17 May 2024 | Accepted: 25 May 2024

DOI: 10.1002/ccr3.9053

CASE REPORT

Clinical Case Reports Open Access WILEY

Radiotherapeutic outcomes of Rosai–Dorfman disease with falx cerebri and superior sagittal sinus involvement: A rare case report with long-term follow-up

Hamid Nasrollahi[1] | **Susan Andalibi**[1] | **Mansour Ansari**[1] | **Maral Mokhtari**[2] | **Ehsan Mohammad Hosseini**[3] | **Mina Foroughi**[4] | **Arman Sourani**[5]

[1]Department of Radiation Oncology, Shiraz University of Medical Sciences, Shiraz, Iran

[2]Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran

[3]Department of Neurosurgery, School of Medicine, Shiraz University of Medical Sciences, Shiraz, Iran

[4]Isfahan Medical Students' Research Committee (IMSRC), Isfahan University of Medical Sciences, Isfahan, Iran

[5]Department of Neurosurgery, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran

Correspondence
Susan Andalibi, Department of Radiation Oncology, Shiraz University of Medical Sciences, Shiraz, Iran.
Email: susan.andalibi@yahoo.com

Key Clinical Message

Intracranial RDD is rare medical event mimicking different diagnoses. Although the surgical resection is the best treatment option, but radiation therapy can also achieves long-term suboptimal outcomes.

Abstract

An 83-year-old male with a history of tension-type headaches was evaluated. He was conscious with no focal neurological deficits. His brain MRI revealed an enhancable bifrontal tumor originating from falx cerebri and superior sagittal sinus dura. Due to the patient's preference and decline for gross total resection, she underwent a stereotactic biopsy. The pathology was positive for Rosai–Dorfman diseases. He received definitive targeted radiation with a total dose of 4500 cGy administered in 200 cGy daily fractions. His 4-year follow-up showed regional tumor control with excellent neurological outcome.

KEYWORDS

brain tumors, lymphoproliferative diseases, Rosai–Dorfman disease, sinus histiocytosis with massive lymphadenopathy, stereotactic biopsy, targeted radiotherapy

1 | INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy (SHML), often known as Rosai–Dorfman Disease (RDD), is a rare benign lymphoproliferative condition. RDD can be categorized as nodal or extranodal, it has been recorded in all age groups but in the majority of cases it manifests in teenagers and young adults as large cervical lymphadenopathy.[1] The skin, orbit, respiratory system, and bone are typical extranodal locations; however rare, central nervous system (CNS) involvement has been documented.[2] A well-circumscribed, dural-based lesion is the most typical appearance of intracranial RDD; intraparenchymal involvement is less frequent.[3,4] However, due to close radiological similarities, these lesions cannot be effectively differentiated from other intracranial lesions. There are no standardized treatment guidelines due to the rarity of RDD-related CNS involvement. Surgery is typically the treatment of choice for localized presentations; however as resection is not always achievable, alternative therapeutic modalities can be needed for disease management.[4,5]

In this report, we have illustrated a falcine-sinus-based RDD lesion that was treated via radiotherapy with long-term follow-up.

2 | CASE PRESENTATION

2.1 | Medical history

An 83-year-old man history with a history of headaches was appointed. The headache had persisted for 1 month with no obvious alleviation and a tension-like pattern. There were no other associated concurrent complaints. Systemic and neurological examinations were negligible. He was a smoker with no considerable past medical or familial records. Lab tests were unremarkable.

2.2 | Work ups

In Brain MRI there was a dural-based, extra-axial mass-like lesion over the anterior part of the falx cerebri overwhelming the superior sagittal sinus, bilaterally. The lesion had a dural base with avid contrast enhancement and was associated with mild peripheral edema (Figure 1). Considering multiple clinical diagnoses, such primary brain tumors or metastatic lesions, a diagnostic stereotactic biopsy was performed.

An immunohistochemistry pathologic investigation confirmed sinus histiocytosis, Rosai–Dorfman disease (Figure 2).

2.3 | Treatment plans

The patient was consulted in a neuro-oncology session including both neurosurgeons and radiation-oncologists, emphasizing that neurosurgical resection of the lesion is the gold-standard treatment option. However, the patient rejected the neurosurgery and preferred "no surgery alternatives." The patient was readvised about the superiority of the surgery over other alternative methods but there was no surgical consent in result. Finally, considering the patient's decline for neurosurgical resection of the lesion, radiation therapy was initiated as an alternative treatment. He received definitive targeted radiation with a total dose of 4500 cGy administered in 180 cGy daily fractions following 3DCRT protocol.

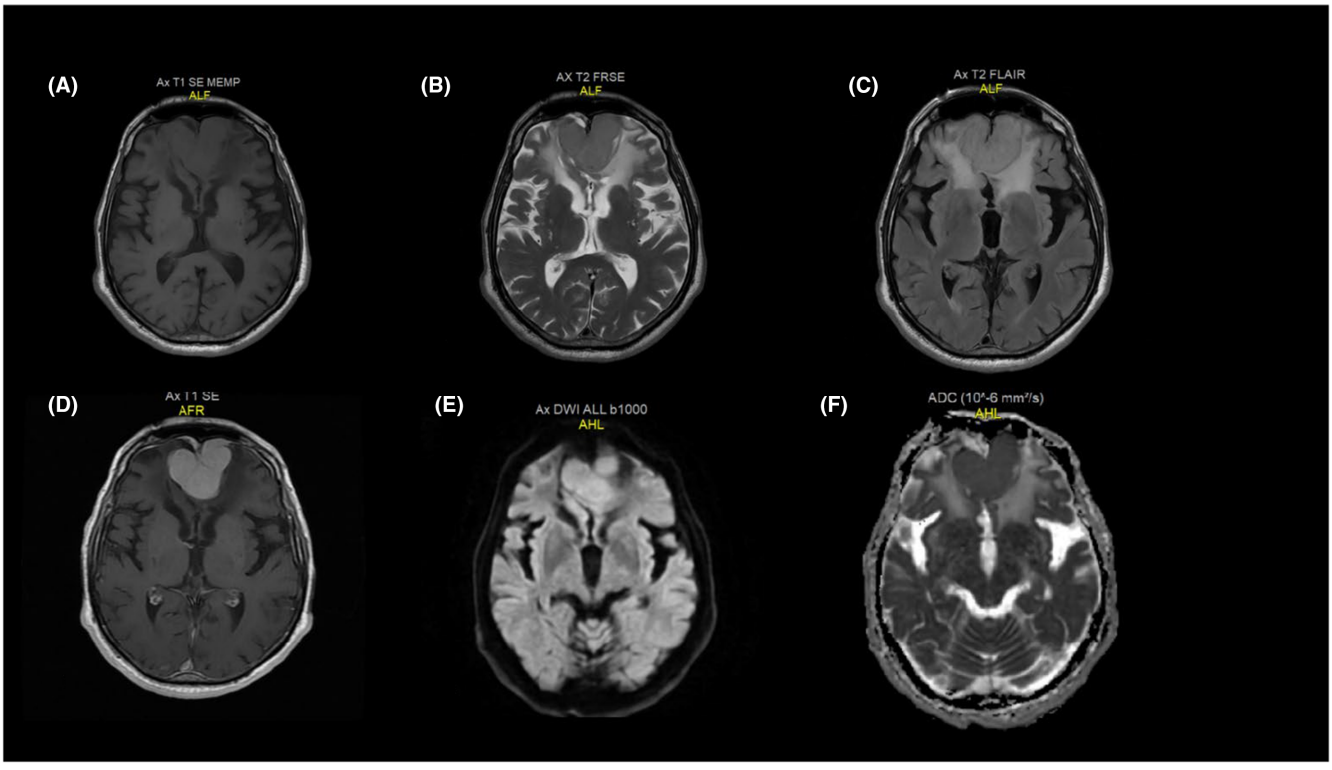

FIGURE 1 MRI sequences of the tumor. T1 (A), T2 (B), FLAIR (C), DWI and ADC images show superior sagittal sinus and falcine involvement of the tumor with mass effect on frontal horns of the lateral ventricles. No hydrocephalus is detectable. T1 with contrast (D) shows avid tumor enhancement. Please notice the tumor edema in a-c images. DWI (E) and ADC (F) sequences also were obtained for regional ischemia.

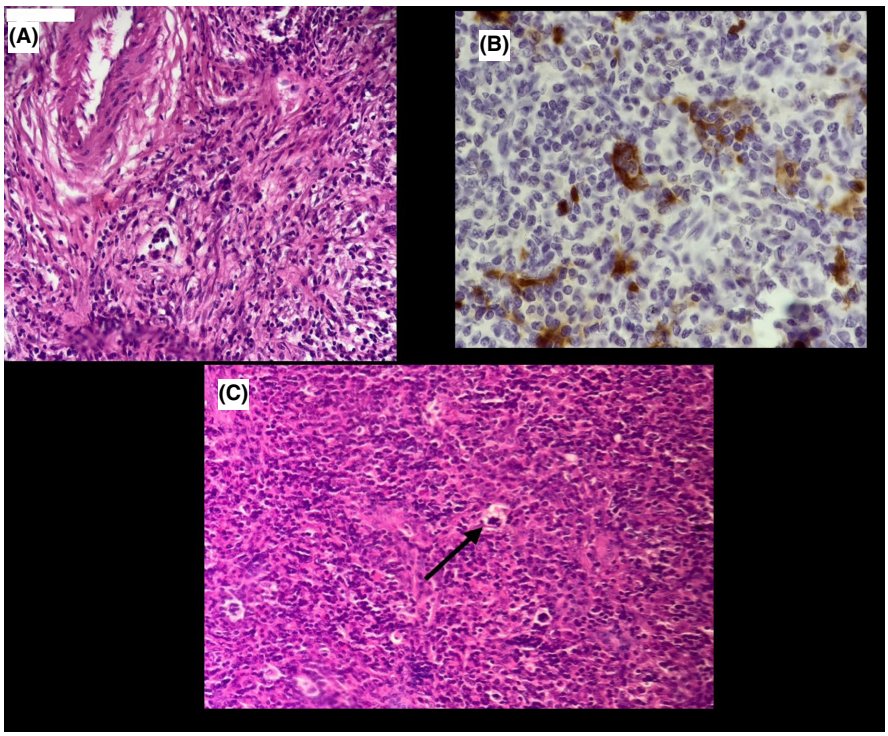

FIGURE 2 (A, B) Sections show paracortical expansion with emperipolesis and abundant lymphoplasma cell infiltration, H&E, ×200, and ×400, respectively. (C) S100 immunostaining which highlights the emperipolesis of lymphocytes, ×400.

2.4 | Follow-up and outcomes

There was no postoperative complications, thus no steroid therapy was advised. After treatment completion, the patient was followed. The follow-up protocol contained a routine physical examination and brain MRI every 6 months. After 4 years of follow-up, the lesion had no obvious growth and remained stable with no new neurological deficits. There was no significant short-term and long-terms post radiation side effects during the follow-up period.

3 | DISCUSSION

RDD is a benign histoproliferative condition with a typical presentation of extensive cervical lymphadenopathy.[6] It frequently affects patients with generalized immunological failure, and many instances following viral infections.[7,8] It has more prevalence in the young and males.[9]

About 40% of RDD cases are extranodal involving the musculoskeletal, skin, and nervous systems.[6] RDD nervous system is an uncommon event. Considering the existing literature, intracranial affection is more prevalent than spinal lesions, yet both can be involved simultaneously.[9] Headache, seizure, focal neurological deficits, limb sensory-motor dysfunctions, and every neurological presentation can be a symptom of nervous system RDD.[10]

An enhanceable, dural-based, mass-like lesion is the typical presentation of intracranial RDD mimicking meningioma in many aspects.[10,11] It may also be accompanied by perilesional vasogenic edema, which can have a mass effect on nearby structures or be associated with regional bone erosion.[11,12] Fukushima etal. reported and reviewed multiple cases of intracranial RDD lesions. Considering the anatomical perspectives, the majority of them were hemispheric lesions.[3] In an interesting paper, Symss et al. reviewed three cases of intracranial RDD lesions with dural involvement. Dural deflections such as tentorium cerebelli, sellar dural structures, and falx cerebri are the main dural invasions of the RDD.[13] Our case was a bifrontal, falcine lesion based on superior sagittal sinus dura with mass effect on the nearby structures. These features are a rarer presentation of the RDD, signifying its vast radiologic presentations.

In the review of the literature, global trend is toward more definitive treatment modalities such as surgical resections. However, there are emerging reports and evidences on alternative treatment options, as well. Radiotherapy and chemotherapy are both available options for those who do not undergo surgical resection due to any cause. The role or radiotherapy in regional tumor control has been elucidated.[1] Multiple studies showed radiation therapy can provides favorable regional tumor control with minimal posttreatment disabilities as compared with systemic chemotherapy or surgical resection. In addition, lower invasiveness, faster recovery, lower posttreatment morbidities of radiation therapy as compared to surgical resection are not negligible, as well. These advantages can of particular interest in selected patients who defer surgery or gross total resection was not met.

However there are radiation induced long-term complications.[13] Recurrence, radio necrosis, radiation-induced malignancies, mass effect, dementia and regrowth are the most important complications of radiation harpy in RDD. Considering the low-quality evidence in such clinical scenarios, a case-by-case discretion should be regarded as the alternative treatments are followed.[1,13,14]

Surgical resection of the mass is considered the gold standard treatment with acceptable outcomes.[12,15] However, subtotal resections or radiosurgery can be an alternative protocol in those with a lower resectability chance.[5] Hadjipanayis et al. reported a clinical experience on subtotal resection combined with stereotactic radiosurgery with acceptable clinical outcomes.[15]

Considering the radioablative protocols, both stereotactic radiation and fractionated radiotherapy applying a total dose of 2000–4500 cGy with 200 cGy/dose have both been employed.[15–17]

Systemic therapy is considered a salvage treatment paradigm for those who failed radiosurgery or neurosurgical resection[12,16] Steroids, rituximab, 6-mercaptopurine (6-MP), and methotrexate (MTX) have been used in the systemic treatment protocols.[1,18] Still, there is no consensus on the best treatment strategies in refractory cases, requiring further reports in this regard.[5]

4 | CONCLUSIONS

Intracranial RDD is a challenging neuro-oncological medical situation requiring multiple treatment paradigms available. Considering the patient's safety and the anatomical considerations, radiotherapy and neurosurgical resection are both viable options. While neurosurgical resection remains the gold standard treatment strategy, those who receive radiotherapy can experience sustained local oncologic control with minimal side effects.

AUTHOR CONTRIBUTIONS

Hamid Nasrollahi: Conceptualization; data curation; methodology; supervision; validation; writing – review and editing. **Susan Andalibi:** Conceptualization; data curation; investigation; methodology; project administration; supervision; validation; writing – original draft. **Mansour Ansari:** Conceptualization; formal analysis; software; validation; writing – original draft. **Maral Mokhtari:** Formal analysis; project administration; resources; validation; visualization; writing – review and editing. **Ehsan Mohammad Hosseini:** Conceptualization; formal analysis; project administration; resources; supervision; validation; visualization; writing – review and editing. **Mina Foroughi:** Formal analysis; validation; writing – original draft. **Arman Sourani:** Conceptualization; investigation; methodology; supervision; validation; writing – original draft; writing – review and editing.

ACKNOWLEDGMENTS

The authors would like to thank Dr. Christian Bowers for his tremendous help during the manuscript preparation.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

Data and original images in the current study are available from the corresponding author upon reasonable request. Authors can confirm that all relevant data are included in the article and/or its supplementary information files.

ETHICS STATEMENT

All procedures performed were under the institutional and/or national research committee's ethical standards and the 1964 Helsinki Declaration and later amendments or comparable ethical standards. Shiraz University neurosurgery department board members supervised and approved this research on behalf of the Ethical Committee of Shiraz University of Medical Sciences.

CONSENT

An informed consent was obtained from the patient for publication. All authors have agreed on the submission and possible publication in the corresponding journal.

ORCID

Hamid Nasrollahi https://orcid.org/0000-0002-7423-3312
Susan Andalibi https://orcid.org/0009-0006-3800-1035
Mansour Ansari https://orcid.org/0000-0002-8249-1776
Maral Mokhtari https://orcid.org/0000-0002-4815-2631
Ehsan Mohammad Hosseini https://orcid.org/0000-0002-9786-9954
Mina Foroughi https://orcid.org/0000-0001-6640-7493
Arman Sourani https://orcid.org/0000-0002-8998-9446

REFERENCES

1. Abla O, Jacobsen E, Picarsic J, et al. Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood*. 2018;131(26):2877-2890.
2. Zhang JT, Tian HJ, Lang SY, Wang XQ. Primary intracerebral Rosai-Dorfman disease. *J Clin Neurosci*. 2010;17(10):1286-1288.

3. Fukushima T, Yachi K, Ogino A, et al. Isolated intracranial Rosai-Dorfman disease without dural attachment–case report. *Neurol Med Chir (Tokyo)*. 2011;51(2):136-140.
4. Unadkat BS, Kashikar SV, Bhansali PJ, et al. Intracranial Rosai-Dorfman disease: a case to remember. *Cureus*. 2022;14(12):e32605.
5. Hollon T, Camelo-Piragua SI, McKean EL, Sullivan SE, Garton HJ. Surgical management of skull base Rosai-Dorfman disease. *World Neurosurg*. 2016;87(661):e5-e12.
6. Bruce-Brand C, Schneider JW, Schubert P. Rosai-Dorfman disease: an overview. *J Clin Pathol*. 2020;73(11):697-705.
7. Harley EH. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) in a patient with elevated Epstein-Barr virus titers. *J Natl Med Assoc*. 1991;83(10):922-924.
8. Magableh HM, Jaber HD, Magableh AM, et al. Rosai-Dorfman disease: case series and literature review. *Cureus*. 2023;15(2):e35193.
9. Sandoval-Sus JD, Sandoval-Leon AC, Chapman JR, et al. Rosai-Dorfman disease of the central nervous system: report of 6 cases and review of the literature. *Medicine (Baltimore)*. 2014;93(3):165-175.
10. Zhang X, Yin W, Guo Y, et al. Rosai-Dorfman disease of the central nervous system: a clinical, radiological, and prognostic study of 12 cases. *Front Oncol*. 2022;12:1013419.
11. Zhu H, Qiu LH, Dou YF, et al. Imaging characteristics of Rosai-Dorfman disease in the central nervous system. *Eur J Radiol*. 2012;81(6):1265-1272.
12. Adeleye AO, Amir G, Fraifeld S, Shoshan Y, Umansky F, Spektor S. Diagnosis and management of Rosai-Dorfman disease involving the central nervous system. *Neurol Res*. 2010;32(6):572-578.
13. Symss NP, Cugati G, Vasudevan MC, Ramamurthi R, Pande A. Intracranial Rosai Dorfman disease: report of three cases and literature review. *Asian J Neurosurg*. 2010;5(2):19-30.
14. Das S, Biswas A, Roy S, et al. Recurrent intracranial Rosai-Dorfman disease: management of a challenging case. *Asian J Neurosurg*. 2017;12(3):537-540.
15. Hadjipanayis CG, Bejjani G, Wiley C, Hasegawa T, Maddock M, Kondziolka D. Intracranial Rosai-Dorfman disease treated with microsurgical resection and stereotactic radiosurgery. Case report. *J Neurosurg*. 2003;98(1):165-168.
16. Cooper SL, Jenrette JM. Rosai-Dorfman disease: management of CNS and systemic involvement. *Clin Adv Hematol Oncol*. 2012;10(3):199-202.
17. Desmond PK, Tamas R, Neil RM. Rosai-Dorfman disease presenting with widespread intracranial and spinal cord involvement. *Neurology*. 2006;67(9):1551-1555.
18. Goyal G, Ravindran A, Young JR, et al. Clinicopathological features, treatment approaches, and outcomes in Rosai-Dorfman disease. *Haematologica*. 2020;105(2):348-357.

How to cite this article: Nasrollahi H, Andalibi S, Ansari M, et al. Radiotherapeutic outcomes of Rosai–Dorfman disease with falx cerebri and superior sagittal sinus involvement: A rare case report with long-term follow-up. *Clin Case Rep*. 2024;12:e9053. doi:10.1002/ccr3.9053