

ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXVII | No. 1

March 2023

Bilateral clinoidal Rosai Dorfman disease
mimicking meningioma – a rare cause of
bilateral blindness

Mayuresh Hinduja,
Hrushikesh Kharosekar,
Vernon Velho,
Laxmikant Bhople

DOI: 10.33962/roneuro-2023-012



Bilateral clinoidal Rosai Dorfman disease mimicking meningioma – a rare cause of bilateral blindness

Mayuresh Hinduja, Hrushikesh Kharosekar,
Vernon Velho, Laxmikant Bhople

Dept. of Neurosurgery, Sir J.J. Group of Hospitals and Grant Medical College, Mumbai, India, INDIA

ABSTRACT

Rosai Dorfman disease is a self-limiting disease usually affecting the lymph nodes. Intracranial lesions are seen in less than 5% of cases. Isolated intracranial RDD without nodal involvement is rare, only 70 cases have been reported to date. Skull base lesions are seen in only 7 cases, petro clival RDD. Clionoidal lesions of RDD have not been reported in the literature, we report a rare case of bilateral clionoidal lesions of RDD presenting with bilateral blindness in a young male.

INTRODUCTION

Rosai Dorfman disease was first described in 1969 by Juan Rosai and Ronald Dorfman as sinus histiocytosis with lymphadenopathy in young male. It commonly presents as painless massive cervical lymphadenopathy and symptoms of fever, malaise and / or anemia. Although all age groups have been effected with the disease, Rosai Dorfman disease commonly effects young males, usually less than 20 years of age group. The disease is extra nodal in up to 43% of patients with or without lymphadenopathy at any point of time in disease. Common extra nodal sites being skin, nose & paranasal sinus, eyelid, orbit & Central nervous system. Intracranial involvement in Rosai Dorfman disease is seen in <5 % of cases of extra nodal disease & in most of these cases leptomeninges are affected.

Isolated cranial involvement without nodal involvement is rare and only 70 such cases are reported in literature till date. To our knowledge bilateral lesions involving the clionoid are not reported in literature, so we report first such case of Rosai Dorfman disease involving bilateral clionoid process.

CASE REPORT

A 21 Year male patient presented to us with complaints of decreased vision in both eyes since 1-month, intermittent headache since 15 days more in morning hours. Patient had one episode of generalized tonic

Keywords
bilateral,
clionoidal,
Rosai Dorfman disease,
blindness



Corresponding author:
Hrushikesh Kharosekar

Dept of Neurosurgery, Sir J.J. group
of Hospitals and Grant Medical
College, Mumbai, India

hkharosekar@gmail.com

Copyright and usage. This is an Open Access article, distributed under the terms of the Creative Commons Attribution Non-Commercial No Derivatives License (<https://creativecommons.org/licenses/by-nc-nd/4.0/>) which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is unaltered and is properly cited.

The written permission of the Romanian Society of Neurosurgery must be obtained for commercial re-use or in order to create a derivative work.

ISSN online 2344-4959
© Romanian Society of
Neurosurgery



First published
March 2023 by
London Academic Publishing
www.lapub.co.uk

clonic seizure 2 days back and then he was referred to our hospital. On neurological examination patient only had visual deficits, his vision was 6/12 (snellen chart) on right side and only perception of hand movements on left side. Patient was investigated further with radiological examination. MRI brain with contrast (Fig 2-4) was done which showed two large well circumscribed lobulated extra axial mass lesion seen along both planum sphenoidale region extending upto both anterior clinoid processes of size 5.2 X 3.2 X 1.2 cm on left side and 1.4 x 2.7 X 3 cm on right side with extension to bilateral anterior temporal lobe and inferior frontal lobe. The lesion was isointense on T1 image and hypointense on T2 image with intense post contrast enhancement. The mass was abutting the cavernous sinus bilaterally without any obvious infiltration. And the lesions were compressing optic chiasma from both sides. Patient was planned for staged surgery for both sides considering the lesions as meningioma. Left sided lesion was approached in first stage as he was developing optic atrophy on that side.

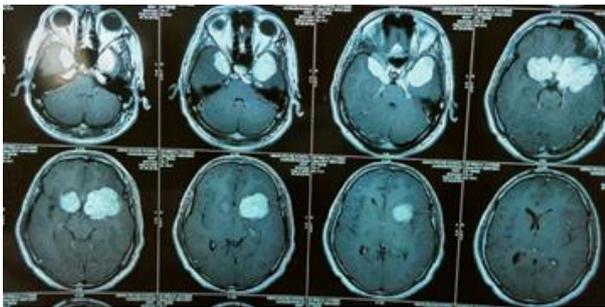


Figure 1. Preoperative MRI Brain with contrast showing bilateral clinoidal lesion (Axial images).

Intraoperatively lesion was reddish grey in color, firm and moderately vascular. Complete excision was achieved. To our surprise Histopathological examination was showing fibro-collagenous tissue with dense chronic inflammatory infiltrate comprising plasma cells, lymphocytes at places showed large histiocytes with phagocytosis of intact lymphocytes and plasma cells (emperipolesis). Russell bodies also noted. CD1a marker was absent suggestive of Rosai Dorfman disease. (Fig 5)

As the disease is self limiting with good prognosis after complete excision, patient underwent second stage surgery immediately after

15 days on right side. Complete excision was achieved, patient was discharged with minimal improvement in vision on right side. Histopathology of the lesion on right side was also suggestive of Rosai Dorfman disease. Post operative MRI brain with contrast after 1 month was suggestive of complete removal of both lesions, no residual lesion. patient was planned for radiotherapy.

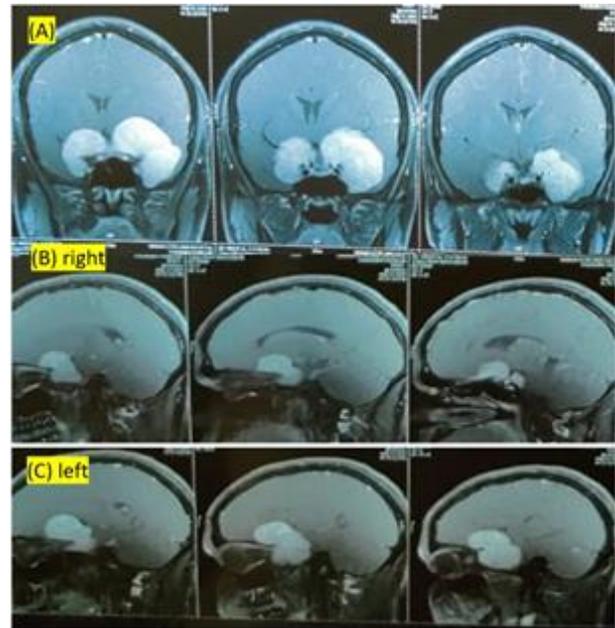


Figure 2. Preoperative MRI Brain with contrast showing bilateral clinoidal lesion (Saggital & Coronal images).

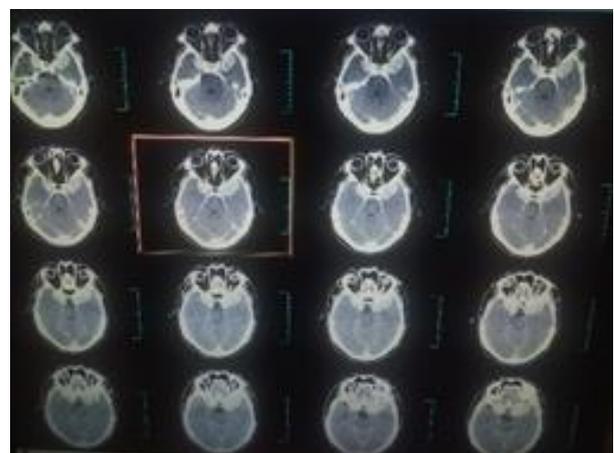


Figure 3. Preoperative CT Brain with contrast showing bilateral clinoidal lesion.

DISCUSSION

RDD is a benign, self-limiting histiocytic disease with

poorly understood etiopathogenesis. Intracranial manifestations are seen in only 5% of cases. There is no age preponderance, though male predominance is seen. The clinical symptoms depend on the location of the lesion, and include headache, nausea and vomiting, seizures, cranial nerve deficits, and weakness.

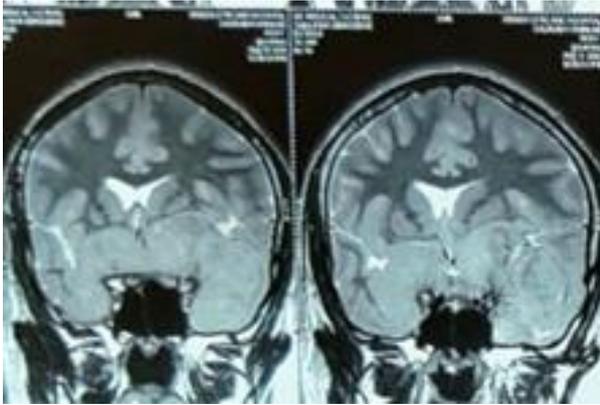


Figure 4. Preoperative MRI Brain with contrast showing bilateral clinoidal lesion (Coronal images, T2 weighted images).

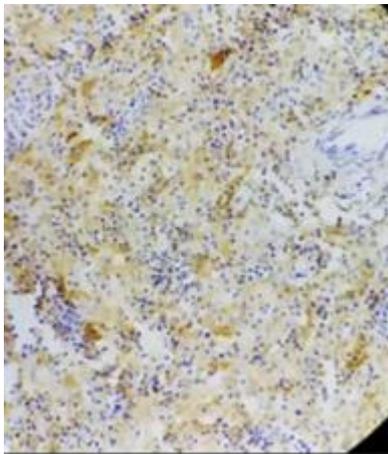


Figure 5. Histopathological slide.



Figure 6. Preoperative images CT brain with contrast before second stage surgery.

In literature only 70 cases have been reported of isolated cranial manifestations in RDD. No case has been reported of clinoidal RDD. So this first case so

far presenting with bilateral lesions of RDD involving the clinoid process on both sides. 7 cases of petroclival lesions have been reported in literature.

A previous study found intracranial lesions re-growth or recurrence of symptoms in 14% of 29 patients with a mean follow-up period of 10.1 years.

Radiologically lesions are typically dural based, extra-axial, well-circumscribed mimicking meningioma.[16–19] On CT, they are homogeneous hyperdense or isodense masses. MRI is gold standard investigation for evaluating lesions of intracranial RDD. On T1-weighted images, the lesions appear as iso to hyperintense masses with clear margins. On T2-weighted images, the lesions usually appear as isointense masses. On contrast images, the lesions demonstrate homogeneous enhancement, and the dural tail can commonly be seen.

In our case, CT demonstrated bilateral well-defined, homogenous, hyperdense masses based on clinoid process; MRI showed dural based well-defined, homogenous lesions based on clinoid process left more than right with cavernous sinus extension on both sides. The lesions were isointense on T1 weighted images, hypointense on T2-weighted images, and homogeneously enhanced. So our working diagnosis was meningioma preoperatively.[12,20] Preoperative radiological findings using the current MRI sequences are difficult to distinguish RDD from meningiomas, but the absence of hyperostosis, bony erosion, or calcification should suggest RDD as a differential diagnosis of meningiomas.[17] Radiologically the lesions in RDD are similar to meningioma on T1 post contrast enhancement, but they have low signal intensity in T2 weighted images, which is very unlikely with meningioma. Other intracranial lesions with T2 image low signal are melanoma, malignant lymphoma, markedly fibrous or calcified meningioma. The reason of low signal in T2 weighted image in this disease is presence of free radicals released by macrophages during phagocytosis. N-isopropyl P-[123I] iodoamphetamine ([123I]IMP) single-photon emission computerized tomography (SPECT) reveals a hot lesion on early and delayed images which is unlikely in cases of meningioma. Final diagnosis of RDD can only be confirmed by histopathological and immunohistochemical examinations.

On histopathology Rosai Dorfman disease shows

fibrosis, histiocytes and lymphoplasmacytic cells. Russell bodies also seen. Histopathological examination is definitive for diagnosis of RDD, especially visualization of emperipolesis and s100, and absence of CD1a (a marker of Langhens cell histiocytosis) . Histopathological differential diagnosis includes lymphoplasmacytic meningioma, Langerhans histiocytosis, lymphoma, plasmacytoma and sarcoidosis.

Lymphoplasmacytic meningioma can be differentiated by presence of typical meningioma picture with epithelial membrane antigen positivity. Langerhans histiocytosis can be differentiated by presence of eosinophils, both s100 and CD1a. Lymphoma can be differentiated by presence of CD15 and CD30 along with Reed Stenberg cells. Plasmacytoma also gives similar picture but the lineage of plasma cells is not polyclonal as in Rosai Dorfman disease. Sarcoidosis can be ruled out by presence of granulomatous inflammation.

In cases of chronic inflammatory background in histology with prominent histiocytes and an absence of infectious agent should alert to proceed with immunohistochemistry staining for s-100. S-100 positivity suggests either Rosai Dorfman disease or Langerhans cell histiocytosis. LCH the histiocytes typically show reniform or indented nucleus, contain birbeck granules and CD1a positivity which differentiated it from Rosai Dorfman disease.

Surgery with complete resection of the lesion is the goal. Use of steroids has shown some promising results in some cases. The lesion is radiosensitive and radiotherapy is a good option for residual or recurrent lesions.

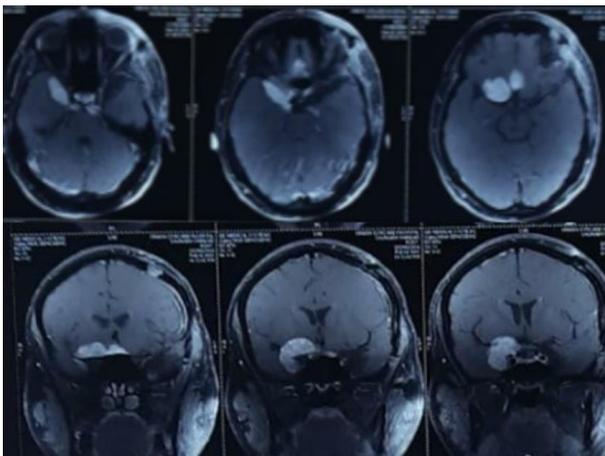


Figure 7. Preoperative images MRI brain with contrast before second stage surgery.

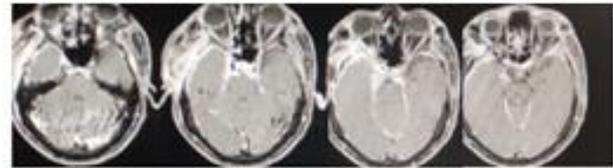
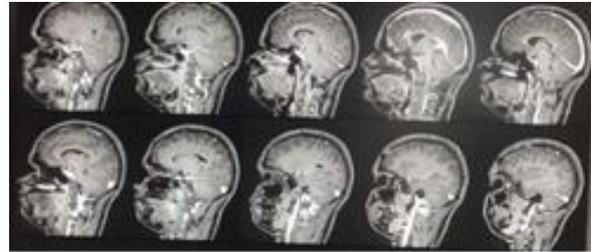


Figure 8. Post operative MRI Brain with contrast after second surgery at 1 month follow up.

CONCLUSION

Rosai Dorfman disease mimics intracranial dural based lesions causing mass effect. It may or may not be associated with extranodal disease. It should be included in differential diagnosis in cases with low intensity T2 image lesion and histologically fibrotic chronic inflammatory lesions of CNS with predominant histiocytic component. Resection of the intracranial lesion is the most effective treatment. In case of subtotal resection, the application of adjunctive radiotherapy and/or steroid agents should be advised.

REFERENCES

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a newly recognized benign clinicopathological entity. *Arch Pathol* 1969;87:63-70.
2. Foucar E, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Review of the entity. *Semin Diagn Pathol* 1990;7:19-73.
3. Vemuganti GK, Naik MN, Honavar SG. Rosai dorfman disease of the orbit. *J Hematol Oncol* 2008;1:7.
4. Walid MS, Grigorian AA. Ethmo-spheno-intracranial RosaiDorfman disease. *Indian J Cancer* 2010;47:80-1.
5. Kattner KA, Stroink AR, Roth TC, Lee JM. Rosai-Dorfman disease mimicking parasagittal meningioma: case presentation and review of the literature. *SurgNeurol* 2000;53:452-7; discussion 457.
6. Udono H, Fukuyama K, Okamoto H, Tabuchi K. Rosai-Dorfman disease presenting multiple intracranial lesion with unique findings on magnetic resonance imaging. *J Neurosurg* 1999;91:335-339.
7. Sze G, Zimmerman RD: The magnetic resonance imaging of infections and inflammatory diseases. *Radiol Clin North Am* 26:839-859, 1988.

8. Prayson RA, Rowe JJ. Dural-based Rosai-Dorfman disease: differential diagnostic considerations. *J Clin Neurosci*. 2014 Nov;21(11):1872-3. doi: 10.1016/j.jocn.2014.07.011. Epub 2014 Sep 30. PMID: 25281034.
9. Krishnamoorthy V, Parmar CF, Panikar D. Isolated intracranial Rosai Dorfman disease. *Neurol India*. 2011;59:443-6.