Scholastic Medical Sciences (ISSN: 3139-7352) Case Report

Volume 3 Issue 12 December 2026

Cervical Myelopathy caused by Rosai–Dorfman Disease: A Rare Extranodal Spinal Presentation

Dr. Leander Lazarus Gomes[1], Dr. Kamlesh Rangari[2], Dr. Harish Naik[3], Dr. Hrushikesh Kharosekar[4], Prof. Vernon Velho[5]

[1]Senior Resident -3rd year, Department of Neurosurgery, Grant Government Medical College and Sir J.J. Group of Hospitals, Mumbai.

[2]Assistant Professor, Department of Neurosurgery, Grant Government Medical College and Sir J.J. Group of Hospitals, Mumbai.

[3]Associate Professor, Department of Neurosurgery, Grant Government Medical College and Sir J.J. Group of Hospitals, Mumbai.

[4]Associate Professor, Department of Neurosurgery, Grant Government Medical College and Sir J.J. Group of Hospitals, Mumbai.

[5]Professor & H.O.D, Department of Neurosurgery, Grant Government Medical College and Sir J.J. Group of Hospitals, Mumbai.

Corresponding Author: Dr. Hrushikesh Kharosekar, Department of Neurosurgery, Grant Government Medical College and Sir J.J. Group of Hospitals, Mumbai.

Received: November 17, 2026; **Published:** November 26, 2026

Abstract

Background: Rosai–Dorfman disease (RDD) is a rare, benign histiocytic disorder classically presenting with massive cervical lymphadenopathy. Extranodal involvement occurs in a significant subset of patients; however, primary spinal involvement causing cervical myelopathy is exceptionally rare and often mimics neoplastic or inflammatory conditions.

Case Presentation: We report a case of a 24-year-old male who presented with progressive cervical myelopathy. Magnetic resonance imaging (MRI) revealed an intradural extramedullary lesion at the C2-C7 level with cord compression at the level. The patient underwent surgical excision with laminotomy and laminoplasty. Histopathological examination demonstrated sheets of large histiocytes with emperipolesis. Immunohistochemistry was positive for S100 and CD68 and few cells showing positivity for CD 163, confirming Rosai–Dorfman disease. Postoperatively, the patient showed significant neurological improvement. No evidence of systemic disease was identified.

Conclusion: Spinal Rosai–Dorfman disease should be considered in the differential diagnosis of cervical compressive myelopathy. Surgical decompression provides both diagnostic confirmation and symptomatic relief. Awareness of this rare entity is crucial to avoid misdiagnosis and overtreatment.

Keywords: Rosai–Dorfman disease; cervical myelopathy; spinal cord compression; extranodal histiocytosis; emperipolesis

Introduction

Rosai–Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare non-Langerhans cell histiocytosis first described in 1969(1), etiology of which was rare and unknown. While nodal disease predominates, extranodal involvement occurs in approximately 40% of cases, commonly affecting the skin, upper respiratory tract, and central nervous system(2). Spinal involvement is extremely uncommon and poses a diagnostic challenge due to its radiological resemblance to meningioma, lymphoma, or metastatic disease. Also spinal

©@2026. Dr. Hrushikesh Kharosekar., et al. All rights reserved by the Author. Published by Scholastic Open Access. This Article is Distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY 4.0).

involvement is rarer as an isolated entity, occurring in 20-25% cases of RDD with CNS disease [3].

We present a rare case of cervical spinal Rosai–Dorfman disease presenting as progressive cervical myelopathy with o/h/o intracranial Rosai Dorfman lesions.

Case Presentation

A 24-year-old male had presented with weakness and numbness of both lower limbs and right arm pain and neck pain since 2-3 months.

Patient had unsteadiness in holding the objects in the right hand along with difficulty in standing up. Also he had difficulty in performing fine movements. There was no history of fever, weight loss, or lymphadenopathy.

Patient had past history of a dural based convexity lesion whose HPE was s/o Rosai Dorfman disease 2 years back where he had undergone a staged surgery for both sides considering that the lesion was a meningioma. The left side was approached first as optic atrophy was developing on that side with the right side being operated 15 days later where complete excision was achieved [1].

Neurological examination-

Power is grade 3 of both the upper limbs

Lower limb reflexes-tone was increased and reflexes were brisk

Investigations-

Pre-op MRI of the cervical spine demonstrated irregular homogenous enhancing dural thickening seen in the cervical spinal canal from C2-C7 levels, left CP angle and along the tentorial leaflets.

Also there was cervical cord compression from C2-C7 with cord edema from C2-C5 levels (T2W1 hyperintense signal).

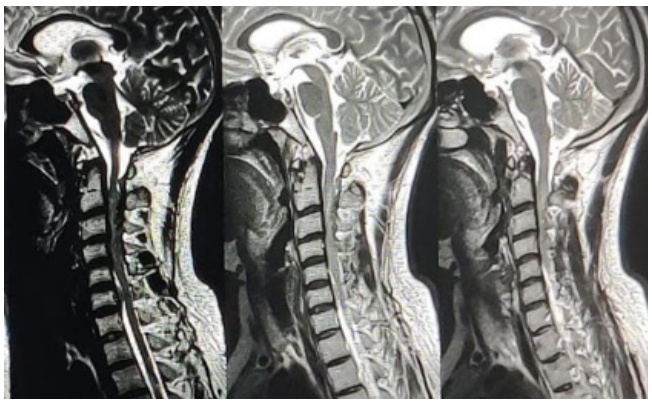


Figure 1: Cervical cord compression from C2-C7 with cord edema from C2-C5 levels (T2W1 hyperintense signal).

Routine laboratory investigations were unremarkable. Whole-body imaging showed no lymphadenopathy or extranodal systemic involvement.

Management

The patient underwent Surgical Excision with Laminotomy and Laminoplasty.

Operative and Specimen images

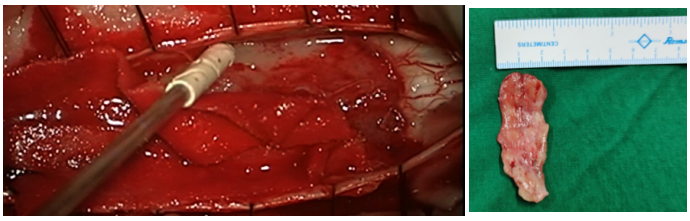


Figure 2: (Excised specimen of C2-C5 intradural extramedullary tumor measuring 4x2x0.2cm, greyish white in color and soft to firm in consistency).

Post op MRI-Mild dural enhancement noted at C4-C5 level(posterior) & C6 level(anterior) of maximum thickness 3.3x3.2x11.2cm seen in the posterior vertebral region at the operative site from C2-D1 vertebral level. Vertebral posterior element bilateral lamina not visualized from C2-C5 s/o post laminectomy status.



Figure 3: Post op MRI s/o Mild dural enhancement noted at C4-C5 level(posterior) & C6 level(anterior) of maximum thickness 3.3x3.2x11.2cm seen in the posterior vertebral region at the operative site from C2-D1 vertebral level.

Histopathological Findings-

Microscopic examination revealed sheets of large histiocytes with abundant pale eosinophilic cytoplasm and vesicular nuclei, exhibiting prominent emperipolesis. Areas of vascular proliferation and variable fibrosis was also seen.

Immunohistochemical analysis showed[2]:

- S100: positive &
- CD68: positive (with few of cells showing positivity for CD163)
- CD1a: negative
- IgG was the predominant population of these cells (small subset of IgG4)

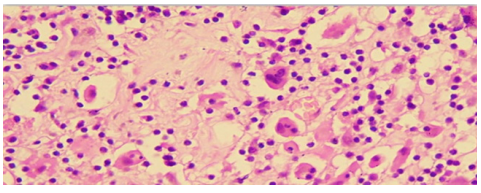


Figure 4: H&E Stain showing large pale histiocytes with pale eosinophilic cytoplasm, round nucleus with vesicular chromatin and distinct nucleoli.

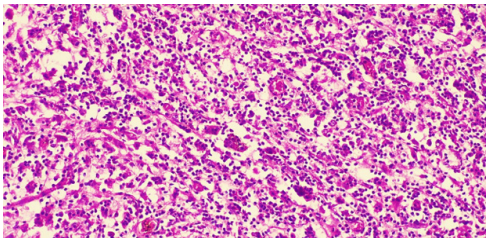


Figure 5: H&E Stain (high magnification) showing histiocytes having small lymphocytes in their cytoplasm known as Emperipolesis.

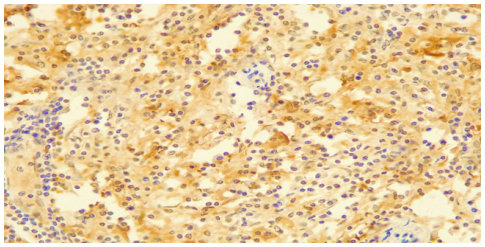


Figure 6: Immunihistochemistry staining showing CD68 & S100 positivity.

These findings were diagnostic of Rosai–Dorfman disease.

Outcome and Follow-Up

Postoperatively, the patient experienced significant neurological improvement, with recovery of gait and limb strength. At follow up after 6 months and 1 year the patient remained neurologically stable with no radiological evidence of recurrence. Adjuvant therapy was not initiated due to complete surgical decompression and absence of systemic disease.

Discussion

Rosai–Dorfman disease (RDD) is an uncommon histiocytic disorder with a highly variable clinical spectrum. Initially described by Destombes in 1965 and subsequently characterized as sinus histiocytosis with massive lymphadenopathy, RDD was traditionally considered primarily a disorder of the lymphatic system [(3)]. Subsequent recognition of extranodal disease has substantially broadened the clinical spectrum of this entity. RDD may involve the skin, upper respiratory tract, orbit, bone, central nervous system (CNS), and several visceral organs [(4)]. The biological behavior of the disease is heterogeneous, ranging from spontaneous regression to persistent, recurrent, or multifocal disease.

CNS involvement is distinctly uncommon, occurring in fewer than 5% of patients with RDD, while isolated spinal disease constitutes only a small fraction of these cases [(5,6)]. Consequently, spinal RDD remains an unusual diagnosis for the neurosurgeon and is frequently encountered only after histopathological examination of a presumed neoplastic lesion. The rarity of this entity becomes particularly important when the disease occurs within the cervical spinal canal, where even a relatively small space-occupying lesion can result in clinically significant spinal cord dysfunction.

Cervical RDD as a cause of myelopathy…..

The clinical presentation of spinal RDD is largely determined by the anatomical location and degree of neural compression rather than by the intrinsic biological aggressiveness of the disease. Patients may present with axial pain, radiculopathy, sensory abnormalities, limb weakness, gait disturbance, or features of long-tract dysfunction. In the cervical region, progressive cord compression may manifest as spasticity, hyperreflexia, pathological reflexes, impaired hand function, gait imbalance, and, in advanced cases, sphincter dysfunction.

This distinction is clinically important. RDD is generally regarded as a benign histiocytic disorder, yet a lesion located in the cervical spinal canal can produce substantial neurological morbidity. Thus, the term "benign" should not imply a benign clinical course when the disease occupies a functionally critical anatomical compartment. In symptomatic patients, neurological deterioration results primarily from mechanical compression of the spinal cord rather than malignant transformation of the histiocytic lesion.

The present case exemplifies this clinical paradox, in which a rare histiocytic disorder produced clinically significant cervical myelopathy. The case therefore highlights the need to consider unusual inflammatory and histiocytic disorders when evaluating a cervical compressive lesion, particularly when its radiological characteristics are atypical for the more common spinal tumors.

Distribution of spinal RDD….

Spinal involvement in RDD is uncommon, and the reported anatomical distribution is variable. The cervical and thoracic regions appear to be most frequently affected, with lesions generally presenting as extradural or intradural extramedullary masses with prominent dural involvement [(7)]. Lin et al.[(8)] reported that approximately 72.5% of isolated spinal RDD lesions in their series were located in the thoracic spine, emphasizing the relative rarity of cervical involvement.

RDD predominantly affects young to middle-aged adults, with most cases occurring between the second and fourth decades of life and a reported male predominance. The precise etiology remains uncertain. Various infectious, viral, immune-mediated, and autoimmune mechanisms have been proposed, although no single causative factor has been conclusively established [(9)]. RDD which is not inclusive of other extranodal sites, but only of the central nervous system is seen in fewer than 5% of patients[(10)].

The present case assumes additional importance because the patient had previously been diagnosed with an intracranial dural-based RDD lesion involving the bilateral clinoidal region[(1)]. The subsequent development of a cervical spinal lesion represents an unusual pattern of CNS disease involving two anatomically distinct compartments. Although multifocal extranodal RDD is recognized, the occurrence of sequential intracranial and spinal lesions in the same patient is distinctly uncommon.

This clinical history has practical implications. In a patient with established RDD, the appearance of a new dural-based lesion at another site should prompt consideration of RDD recurrence or multifocal disease rather than automatically being classified as an unrelated common neoplasm. Conversely, when spinal RDD is diagnosed, clinicians should remain alert to the possibility of disease elsewhere in the CNS or other extranodal sites.

Why spinal RDD is difficult to diagnose….

The diagnosis of spinal RDD before surgery can be challenging because there are no pathognomonic clinical or radiological findings. The lesions frequently resemble other dural-based masses, particularly meningiomas. This creates a diagnostic dilemma because meningioma is considerably more common and is often the initial radiological impression in a patient with a well-circumscribed, avidly enhancing dural-based spinal mass.

MRI typically demonstrates a well-defined lesion with iso- to hypointense signal on T1-weighted sequences and variable, often

relatively low, signal intensity on T2-weighted images [11]. Following administration of contrast, marked enhancement is commonly observed. Dural attachment and a dural tail may also be present, further reinforcing the radiological impression of meningioma [12].

However, these findings are not specific. Lymphoma, metastasis, inflammatory lesions, granulomatous disorders, and other histiocytic proliferations may demonstrate overlapping imaging characteristics. Therefore, radiological diagnosis should be considered provisional when the clinical setting or imaging pattern is unusual.

In the present case, the previous history of intracranial RDD provides an important retrospective diagnostic clue. A new cervical dural-based lesion in such a patient should raise the possibility of spinal RDD, particularly when the lesion demonstrates imaging characteristics similar to the previous intracranial disease.

Role of functional imaging….

Assessment of the extent of disease may be particularly useful in patients with extranodal or CNS RDD. Dhull et al.[13] described the use of 18F-FDG PET/CT in RDD and demonstrated its potential value in identifying metabolically active disease and evaluating the distribution of involvement. Although PET/CT is not required for every patient with localized spinal disease, it may be useful when multifocal or systemic involvement is suspected.

In a patient with a history of CNS RDD who develops a new spinal lesion, whole-body assessment may therefore provide useful information regarding disease burden and help determine whether the lesion represents isolated spinal involvement or part of a more extensive disease process.

Histopathology: the decisive diagnostic step….

Although imaging may suggest RDD, histopathological examination remains the definitive method of establishing the diagnosis. The characteristic microscopic finding is the presence of large histiocytes containing intact lymphocytes, plasma cells, or other inflammatory cells within their cytoplasm. This phenomenon, termed emperipolesis, is a defining pathological feature of RDD which has also been cited by Mirra et. al[14].

Emperipolesis is particularly valuable in distinguishing RDD from other inflammatory and neoplastic lesions. Importantly, the cells present within the histiocyte remain morphologically recognizable rather than undergoing the destructive process characteristic of conventional phagocytosis. Recognition of this feature in an otherwise nonspecific dural-based lesion should prompt consideration of RDD.

Immunohistochemistry provides additional diagnostic support. RDD histiocytes typically demonstrate positivity for S-100 and CD68, while CD1a is characteristically negative [15]. This immunophenotypic pattern helps distinguish RDD from Langerhans cell histiocytosis and other histiocytic disorders.

The pathological findings in our patient were consistent with this characteristic profile, including emperipolesis and the appropriate immunohistochemical expression. The pathological diagnosis was therefore essential in establishing the nature of the cervical lesion and in correlating it with the patient's previous intracranial RDD.

RDD and its variable natural history…..

An additional characteristic of RDD is its unpredictable clinical course. Not all lesions require immediate intervention, and spontaneous regression has been reported in both nodal and extranodal disease. Pulsoni et al.[16], in their review of 40 patients, demonstrated that spontaneous remission may occur in a substantial proportion of cases. Similarly, Ritter et al.[17] reported spontaneous resolution of multiple spinal RDD lesions following treatment with non-steroidal anti-inflammatory medication.

These observations suggest that RDD should not be regarded as a uniformly progressive disease. Nevertheless, the possibility of spontaneous regression must be interpreted in relation to the anatomical location of the lesion. Observation may be reasonable for an asymptomatic and stable lesion, but it is less appropriate when a cervical lesion is producing progressive cord compression.

The cervical spinal cord has limited tolerance for prolonged compression, and neurological deficits may become irreversible if decompression is delayed. Therefore, the potential for spontaneous regression should not discourage timely intervention in patients with clinically significant myelopathy.

Surgical management…..

For symptomatic spinal RDD, surgery remains an important component of management [18,19]. The rationale for surgery extends beyond obtaining tissue for diagnosis. In patients with neurological compromise, decompression directly addresses the primary cause of morbidity by relieving pressure on the spinal cord.

Whenever anatomically feasible, complete removal is desirable; however, the surgical objective should be balanced against the risk of neurological injury. Dural attachment, adherence to the spinal cord, nerve roots, or adjacent vascular structures may make complete excision difficult. In such circumstances, adequate decompression with maximal safe resection may be more appropriate than pursuing radical excision at the expense of neurological function.

The favorable postoperative course in our patient reinforces the value of early surgical intervention in symptomatic cervical spinal RDD. The neurological outcome also illustrates an important principle in spinal RDD: although the underlying disorder is benign, the consequences of untreated mechanical compression can be serious and potentially irreversible.

Role of nonsurgical and adjuvant therapy…..

Because RDD demonstrates variable biological behavior, treatment must be individualized. Observation may be appropriate in patients with asymptomatic lesions, minimal disease burden, or evidence of spontaneous regression. In contrast, symptomatic lesions producing spinal cord compression generally require surgical treatment.

For recurrent, residual, multifocal, unresectable, or systemically disseminated disease, corticosteroids, radiotherapy, and systemic medical therapies have been used with variable success [20,21]. Radiotherapy may have a role when residual disease cannot be safely removed surgically or when recurrence occurs in a surgically inaccessible location. Systemic treatment becomes more relevant when RDD involves multiple organs or demonstrates progressive disease despite local therapy.

The absence of a universally accepted treatment protocol reflects the rarity and biological heterogeneity of RDD. Management is therefore best individualized according to neurological status, lesion location, extent of disease, resectability, recurrence, and systemic involvement.

Follow-up and risk of multifocal disease…..

Long-term surveillance is important in patients with CNS RDD because the disease may recur or manifest at a different anatomical site.

This consideration becomes particularly relevant in the present case because the patient had already demonstrated intracranial disease before developing cervical spinal involvement.

The temporal separation between the cranial and spinal manifestations suggests that CNS RDD may occasionally remain clinically silent at one site while subsequently becoming symptomatic at another. Consequently, successful surgical treatment of a spinal lesion should not necessarily be regarded as the end point of management. Continued clinical assessment and appropriate interval imaging may be warranted, particularly in patients with a history of multifocal or recurrent disease.

Clinical significance of the present case….

The present case illustrates the diagnostic and therapeutic challenges posed by an uncommon extranodal manifestation of RDD. The cervical location is clinically important because of the potential for spinal cord compression and progressive myelopathy. The previous intracranial RDD provides an additional dimension to the case, suggesting multifocal CNS involvement rather than an isolated spinal event.

From a neurosurgical perspective, three lessons are particularly relevant. First, RDD should be included in the differential diagnosis of a cervical dural-based lesion, even when the radiological appearance strongly suggests meningioma. Second, the diagnosis cannot reliably be established on imaging alone and requires histopathological and immunohistochemical confirmation. Third, once neurological compression develops, timely decompression is crucial because the clinical consequences of cervical cord compression may be more significant than the inherently benign biological behavior of RDD.

The rarity of spinal RDD can itself contribute to diagnostic delay. However, recognition of characteristic pathological findings, awareness of its radiological overlap with meningioma, and attention to a previous history of RDD can substantially improve diagnostic accuracy.

Conclusion

Cervical spinal Rosai–Dorfman disease represents an exceptionally uncommon extranodal manifestation of a rare histiocytic disorder and should be considered in the differential diagnosis of a cervical dural-based lesion causing myelopathy. Its radiological resemblance to meningioma can create a significant diagnostic challenge, making histopathological confirmation indispensable. The demonstration of emperipolesis together with S-100 and CD68 positivity and CD1a negativity provides strong diagnostic support.

The present case is particularly distinctive because of the occurrence of both intracranial and cervical spinal RDD in the same patient, with the spinal lesion producing clinically significant cervical myelopathy. This observation highlights the potential for RDD to involve multiple CNS compartments and reinforces the importance of long-term surveillance.

Ultimately, the clinical behavior of spinal RDD is determined not merely by its benign histological nature but by where the disease occurs and what structures it compresses. In the cervical spine, timely recognition and surgical decompression can prevent progressive neurological deterioration and result in meaningful functional recovery. Thus, RDD should remain an important, albeit rare, diagnostic consideration when evaluating a dural-based cervical spinal lesion, particularly in patients with a previous history of RDD.

Take home message

Rosai-Dorfman is considered a benign entity, where surgical resection is considered justified for decompression and establishment of the diagnosis. In the case where resection is not possible then chemotherapy for systemic RDD and radiotherapy for inoperable cases can be used. However in our case which was previously operated for a dural based convexity lesion whose HPE was also s/o RDD, surgical excision of the cervical lesions was done along with follow up and the patient also shows favorable prognosis.

In our case it was seen that surgical treatment was alone effective for the cervical extranodal RDD metastasis, probably from the previous intracranial RDD without the need for post operative radiotherapy and chemotherapy.

This can be taken as a lesson to emphasize on whole spine screening in patients with intracranial RDD to pick up and spinal lesions if any, due to the favorable prognosis of such lesions if picked up early.

Declarations

Ethics approval and consent to participate: Not applicable. Formal ethical approval was not required for this case report as per our institutional guidelines.

Consent for Publication: Written informed consent was obtained from the patient for publication of this case report along with the accompanying clinical details and images.

Availability of data and material: All the relevant data supporting the findings of this case report have been included in the manuscript. Any further information may be obtained from the corresponding author upon request for the same.

Competing interests: The authors declare that they have no competing interests.

Funding: No funding was received for the preparation of this case report.

References

1. Mayuresh Hinduja, Hrushikesh Kharosekar, Vernon Velho, Laxmikant Bhople (2023). Bilateral clinoidal Rosai Dorfman disease mimicking meningioma-a rare cause of bilateral blindness. *Romanian Neurosurgery* XXXVII(1): pp.75-79

2. Burgdorf W, Zelger B (2009). The histiocytosis. In:Elder DE, editor. Lever's Histopathology of the skin. 10th ed. Philadelphia: Lippincott Williams and Wilkins. pp. 678-9.

3. Destombes P (1965). Adenitis with lipid excess, in children or young adults, seen in the Antilles & in Mali. (4 cases). *Bull Soc Pathol Exot Fil.* 58:1169-75

4. Rosai J, Dorfman RF (1969). Sinus histiocytosis with massive lymphadenopathy: a newly recognized benign clinicopathological entity. *Arch Pathol*. 87:63-70

5. Ma J, Xiao J. Wang L (2008). Extranodal Rosai-Dorfman disease with multilevel lumbar spinal lesions. *J Neurosurg Spine*. 9:55-7

6. Sandoval-Sus JD, Sandoval-Leon AC, Chapman JR, Williams L, Garcia R, Rodriguez F, et al (2014). Rosai-Dorfman disease of the central nervous system: report of 6 cases and review of the literature. *Medicine (Baltimore)*. 93:165-75

7. Hu,Pp., Wei,F., Liu,Xg. Et al. Diagnosis and treatment of Rosai-Dorfman disease of the spine: a systematic literature review. *Syst Rev* 10, 31(2021)

8. Lin CK, Tsai YD (2019). Nonresectable thoracic Rosai-Dorfman Disease: a case report and review of literature. *World Neurosurg*. 132:309-13

9. Osenbach RK (1996). Isolated extranodal sinus histiocytosis presenting as an intramedullary spinal cord tumor with paraplegia. *Case report. J Neurosurg*. 85:692-696

10. Cooper SL, Jenrette JM (2012). Rosai Dorfman disease: Management of CNS and systemic involvement. *Clin Adv Hematol Oncol*. 10:199-202.

11. Johnston JM, Limbrick DD, Ray WZ, Brown S, Shimony J, Park TS (2009). Isolated cerebellar Rosai-Dorfman granuloma mimicking Lhermitte-Duclos disease. Case report. *J Neurosurg Pediatrr*. 4:118-120.

12. Wu L, Xu Y (2014). Rosai-Dorfman disease: a rare lesion with dural tail sign mimicking spinal meningioma. *Spine J*. 14:3058-9.

13. Dhull VS, Passah A, Rana N, Kaur K, Tripati M, Kumar R (2016). 18F-FDG PET/CT of widespread Rosai-Dorfman disease. *Clin Nucl Med.* 41:57-59

14. Mirra SS, Tindal SC, Check IJ, Brynes RK, MooreWW (1983). Inflammatory meningeal masses of unexplained origin. An ultrastructural and immunological study. *J Neuropathol Exp Neurol*. 42:453-468.

15. Huang BY, Zhang H, Zong WJ, Sun YH (2016). Rosai-Dorfman disease of rare isolated spinal involvement: Report of 4 cases and literature review. *World Neurosurg*. 85: 367.e11-367.e16.

16. Pulsoni A, Anghel G, Falcucci P, Matera R, Pescarmona E, Ribersani M, et al (2002). Treatment of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): report of a case and literature review. *Am J Hematol*. 69:67-71

17. Rittner RE, Baumann U, Laenger F, Hartung D, Rosenthal H, Hueper K (2012). Whole-body diffusion-weighted MRI in a case of Rosai-Dorfman disease with exclusive multifocal skeletal involvement. *Skelet Radio*. 41:709-13.

18. Kayali H, Onguru O, Erdogan E, Sirin S, Timurkaynak E (2004). Isolated intracranial Rosai-Dorfman disease mimicking meningioma. *Clin Neuropathol*. 23:204-208.

19. Griffiths SJ, Tang W, Parameswaran R, Kelsey A, West CG (2004). Isolated intracranial Rosai-Dorfman disease mimicking meningioma in a child. *Br J Neurosurg.* 18:293-297.

20. Lohr HF, Godderz W, Wolfe T, Heike M, Knuth A, Meyer Zum Buschenfelde KH, Dippold W (1995). Long-term survival in a patient with Rosai-Dorfman disease treated with Interferon-alpha. *Eur J Cancer*. 31A;2427-2428.

21. Abla O, Jacobson E, Picarsic J, et. Al (2018). Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood*. 131:2877-2890.