Case Report

Rosai-Dorfman disease of cranial and spinal origin - A case series

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ABSTRACT

Background: Rosai-Dorfman disease (RDD) is an idiopathic nonneoplastic lymphadenopathy disorder which is characterized by lymph node enlargement, but it may also present**s** primarily involving a variety of extranodal sites, including central nerves system and craniospinal axis. This study reports five cases of craniospinal RDD, with review of epidemiology, clinical presentation, imaging, and histopathological features with current management strategies.

Case Description: Five cases of RDD are diagnosed at Hamad General Hospital, Qatar, during 2013–2018. Two cases had dural-based cranial lesions with overlying cranial involvement while three cases were having extradural thoracic spine lesions. All cases underwent surgical intervention and confirmed by histopathology.

Conclusion: Craniospinal RDD is a rare clinical presentation and poses significant diagnostic challenges preoperatively due to its similarity with other neoplastic or inflammatory diseases. Surgical option to remove compressive neural pathology provides a good clinical outcome with no recurrence in long-term follow-up.

Keywords: Central nerves system, Disease, Dural based, Lymphadenopathy, Meningioma, Resection. Rosai-Dorfman, Sinus histiocytosis, Spinal

INTRODUCTION

Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive lymphadenopathy, is a benign, idiopathic histiocytic proliferative disease with pathognomonic histological and immunohistochemical characteristics.[6] RDD was initially described in French literature as a lipid storage disorder possibly developing after inflammation (ade´nites avec surcharge lipidique) by Destombes in 1965[9] and was recognized as a unique histiolymph**o**proliferative disease of the lymph nodes by Rosai and Dorfman in 1969.[24] RDD is histologically characterized by infiltration of lymph nodes or extranodal tissues by nonmalignant histiocytes exhibiting emperipolesis, a nondestructive phagocytosis of lymphocytes or erythrocytes, which represents the hallmark of this disease and is required for definitive diagnosis.[23] Extranodal involvement was subsequently recognized and an analysis of the RDD registry published in 1990 showed that 43% of cases were extranodal.[11] RDD usually presents with massive painless cervical lymphadenopathy along with malaise, fever, weight loss, anemia, leukocytosis, elevated erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia.[11]

Extranodal lesions have been reported at various sites including the central nerves system (CNS) and the bone; however, bone involvement was reported to be <10% with only two cases reported

involving the skull bone to date.[11] CNS involvement accounts for only 5% with 71–82% of patients having no associated systemic disease.[3,14,18,29] Spinal cases are described in 20–25% of all CNS cases, but symptomatic intramedullary spinal cord involvement is extremely rare.[14,15] We present five cases of RDD with isolated CNS involvement managed at Hamad General Hospital in Qatar. Two cases have dural-based cranial presentation and other three cases have extradural location involving thoracic spine.

CASES DESCRIPTION

Case No.1

A 28-year-old female patient presented with 10 days history of increasing headache, associated with weakness of the right upper and lower limbs. On clinical examination, she had normal vital signs, fully conscious and oriented, right hemiparesis Grade 4 (Medical Research Council Scale). There were no signs of lymphadenopathy. Computerized tomography (CT) scan and magnetic resonance imaging (MRI) of the brain revealed a calvarial epicentered mass lesion with sharp beveled-edged bony defect, extracranial and intracranial extradural and sulcal extension, and a surrounding brain edema. The lesion displayed iso- to low signal intensity on T2W, intermediate signal on T1W MRI, postcontrast enhancement, reduced relative cerebral blood volume (rCBV) on perfusion MRI, and no diffusion restriction [Figure 1]. The primary diagnostic consideration was Langerhans cell histiocytosis (LCH) because of the sharp beveled-edged lytic bony lesion along with soft-tissue component, with a differential diagnosis of other non-LCH like RDD. Lymphoma was considered unlikely due to the absence of diffusion restriction, and the possibility of meningioma was also low due to the absence of elevated rCBV in the lesion. The patient underwent left parietal craniotomy for removal of dural-based lesion that appeared yellowish and was thickened, rubbery in consistency. Biopsy of cortical lesion and drilling of bony infiltrating lesion were also done. Histopathology revealed a histiocytic lesion with infiltration into bony trabeculae of the skull. Many histiocytes are large with abundant pale cytoplasm engulfing intact lymphocytes (emperipolesis). The diagnosis of cranial RDD was established [Figure 2]. She had uneventful postoperative hospital stay, and subsequent work-up with CT chest, abdomen, and pelvis remained normal. The patient was discharged without any deficits. Serial MRI postoperative follow in the past 14 months did not show any recurrence.

Case No.2

A 25-year-old male patient presented to emergency department (ED) after found unconscious, with an unwitnessed seizure activity. On arrival in ED, he regained his
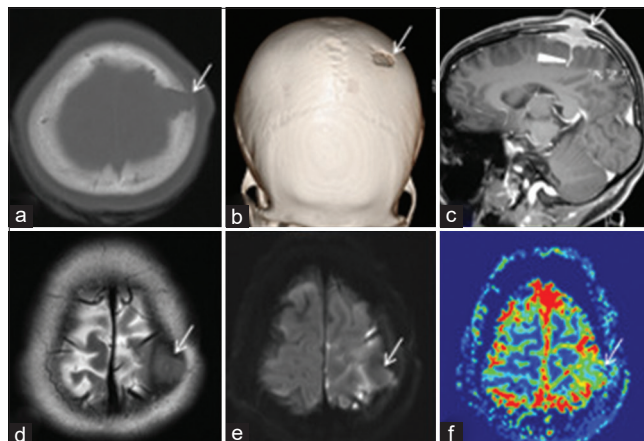

Figure 1: Axial (a) and 3D volume rendered (b) computerized tomography scan bone window images show a sharp defect in the anterior left parietal bone with beveled edges (arrow). Sagittal postcontrast 3D T1 MPRAGE magnetic resonance imaging (MRI) (c) shows enhancing soft tissue in the scalp and intracranial extra-axial location extending across and filling the calvarial defect (arrow); with extension of enhancing soft tissue into the underlying sulcus (arrowhead). Axial T2W MRI (d) shows that the lesion (arrow) is iso to slightly hypointense to cortex. Axial DWI MRI (e) shows no diffusion restriction within the lesion (arrow). Axial perfusion MRI relative cerebral blood volume (rCBV) image (f) shows reduced rCBV within the lesion (arrow) compared with the adjacent cortex.
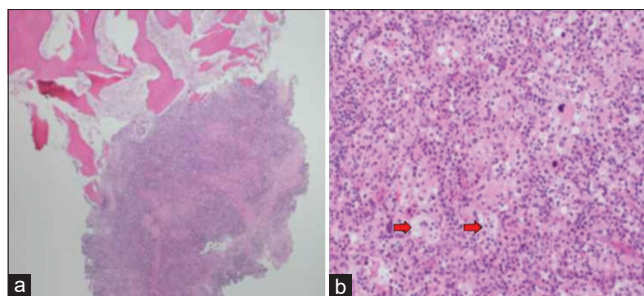

Figure 2: (a) Photomicrograph depicting bony trabeculae infiltrated by Rosai-Dorfman disease (hematoxylin and eosin stain ×40. (b) High-power view shows histiocytic proliferation along with diffuse lymphoplasmacytic infiltrate. Some histiocytes are large with emperipolesis (red arrows) (hematoxylin and eosin stain ×200).

consciousness but had retrograde amnesia. On examination, he had normal vital signs and his Glasgow Coma Scale (GCS) was 15/15 with no neurological deficits. CT scan showed left posterior frontal hypodense lesion. MRI revealed a left inferior parietal extra-axial mass lesion displaying predominant low signal intensity on T2W, intermediate signal on T1W MRI images, postcontrast enhancement, reduced rCBV on perfusion MRI, and no diffusion restriction. There were contiguous enhancing dural tails and a T2 hypointense enhancing tiny nodular focus in the adjacent brain. There was perilesional brain edema. No bony

involvement was seen [Figure 3]. A preoperative radiological diagnosis of granulomatous infection or sarcoidosis with the differential diagnosis of other rare disorders like histiocytosis was considered. The patient underwent left frontoparietal craniotomy with resection of the extra-axial mass lesion. Histopathology showed a histiocytic lesion arising from thick fibrous dura. The histiocytes were large and many of them showing emperipolesis. By immunoperoxidase stains, these were diffusely and strongly positive for S100 antibody, consistent with RDD [Figure 4]. This patient had an uneventful postoperative hospital course and was discharged without any neurological deficits.
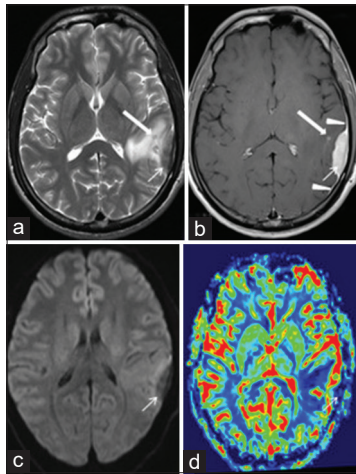

Figure 3: Axial T2W magnetic resonance imaging (MRI) (a) shows a left inferior parietal extra-axial soft-tissue lesion (arrow) with surrounding hyperintense edema. The lesion is hypointense to cortex and there is also a tiny hypointense nodule in the adjacent brain (thick arrow) with intact calvarium. Axial postcontrast TW MRI (b) shows uniform enhancement of the lesion (arrow). Note the contiguous enhancing dural tails (arrowheads) and enhancement of the tiny nodular focus in the adjacent brain (thick arrow). Axial DWI MRI (c) shows no diffusion restriction within the lesion (arrow). Axial perfusion MRI relative cerebral blood volume (rCBV) image (d) shows reduced rCBV within the lesion (arrow) compared with the adjacent brain.
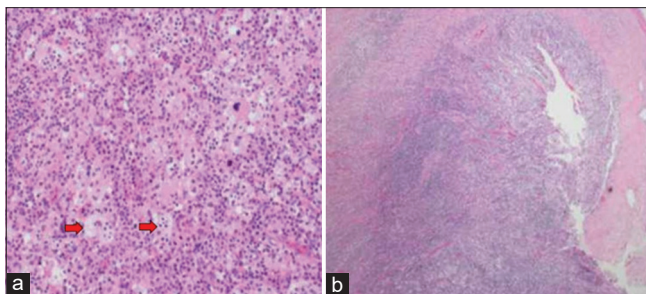

Figure 4: (a) Large histiocytes with abundant pale cytoplasm engulfing intact lymphocytes (red arrows) (hematoxylin and eosin stain ×200). (b) Photomicrograph showing inflammatory histiocytic proliferation arising from thick fibrous dura (right upper corner) (hematoxylin and eosin stain ×40).

Case No.3

A 25-year-old male patient presented with complaints of progressive lower limbs weakness and difficulty in walking for the past 10 days. On examination, his GCS was 15/15 and he had bilateral lower limb weakness Grade 3/5, hyperreflexia, and muscle spasticity with sensory level of thoracic (T) vertebra no. 6. MRI of spine showed enhancing right paravertebral soft-tissue lesion extending from cervical (C) vertebra No. 7 through T4 vertebral levels with extradural extension encasing the spinal cord with cord edema and patchy areas of myelitis. There was no significant vertebral bony involvement [Figure 5]. The radiological differential diagnosis was granulomatous infection or lymphoma/leukemia, with the differential diagnosis of other rare disorders like histiocytosis. The patient underwent laminoplasties from T1 to T2 and excision of epidural mass lesion. Histopathology revealed a dura-based histiocytic lesion with some histiocytes containing intact lymphocytes. The histiocytes were diffusely positive for S100 immunostain, consistent with spinal RDD [Figure 6]. Bone marrow biopsy showed 5–20% of histiocytic cells with trilineal hematopoiesis. Histiocytes stuffed with intact hematopoietic cells. Postoperatively, the patient showed mild improvement of his power and was continued to have active rehabilitation management.

Case No.4

A 52-year-old male patient presented progressive difficulty in walking and urinary retention for the past 3 days. On examination, he had bilateral lower limb weakness Grade 2/5, with bilateral Babinski' sign extensor bilaterally and muscle spasticity with sensory level of T6 and lax anal tone. MRI of the spine showed a large epidural enhancing soft-tissue lesion nearly filling the whole spinal canal at T4 level with dural tails superiorly and inferiorly. The lesion produced significant compression and focal displacement of the thoracic spinal cord to the left anterolateral aspect. In addition, there was a slightly T1 and T2 hyperintense enhancing lesion within the T4 vertebral body, which could bring in the differential diagnosis of vertebral bony and spinal canal metastatic deposits. However, a correlative CT scan image clearly showed that the bony lesion was an incidental vertebral body hemangioma [Figure 7]. A perioperative radiological diagnosis of meningioma versus granulomatous infection or lymphoma/leukemia, with the differential diagnosis of other rare disorders like histiocytosis was entertained. The patient underwent laminectomies from T3 to T5 and excision of extradural mass. Histopathology showed a dura-based lesion composed of histiocytic proliferation with lymphoplasmacytic infiltrate along with lymphophagocytosis, compatible with RDD [Figure 8]. Postoperative MRI showed no residual tumoral tissue. The patient showed improvement in his power postoperatively
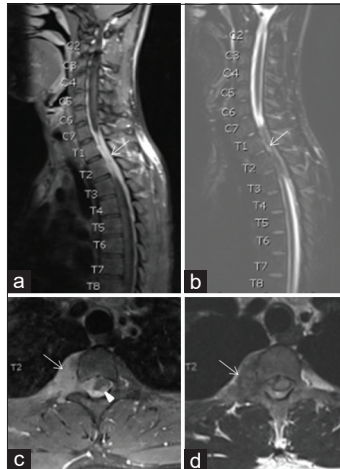
Figure 5: Sagittal postcontrast T1W magnetic resonance imaging (MRI) of the cervicothoracic spine (a) shows a right-sided enhancing epidural soft-tissue lesion (arrow) which appears hypointense on T2W-STIR MRI (b) at C7-T4 spinal canal levels. Axial postcontrast T1W MRI (c) shows extension through the right neural foramen with a sizeable paravertebral soft-tissue component (arrow). Note a tiny enhancing component within the spinal cord parenchyma (arrowhead). Axial T2WI (d) shows hypointensity of the paraspinal lesion and diffuse spinal cord hyperintensity. No vertebral bony involvement is seen.
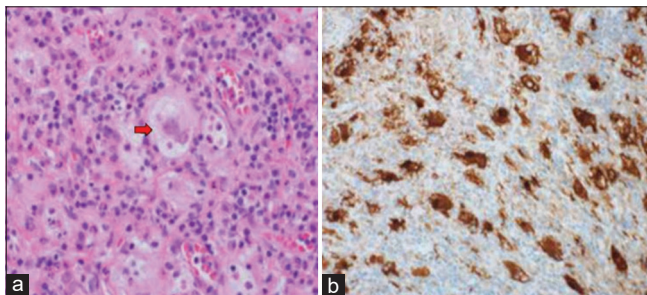

Figure 6: (a) Photomicrograph showing large histiocytes with emperipolesis (red arrow) (hematoxylin and eosin stain ×400). (b) The histiocytes show strong diffuse nuclear and cytoplasmic staining for S100 antibody (immunoperoxidase ×200).

and was followed with rehabilitation medicine. The patient was discharged from their side without deficits.

Case No.5

A 40-year-old Qatari male presented with 1-day history of bilateral lower limbs weakness associated with urinary incontinence. On examination, he had bilateral lower limb weakness, more pronounced distally along with bilateral spasticity and pinprick sensory level at T8. MRI of the spine showed a large epidural enhancing soft-tissue lesion nearly filling the whole spinal canal extending from T6 to T11, with dural tails superiorly and inferiorly. The lesion produced significant compression and focal displacement of the
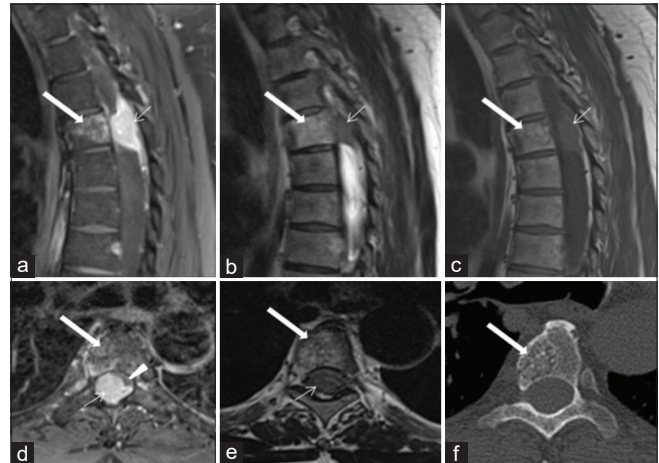

Figure 7: Sagittal postcontrast T1W magnetic resonance imaging (MRI) of the thoracic spine (a) shows an enhancing epidural soft-tissue lesion nearly filling the spinal canal (arrow) with dural tails superiorly and inferiorly, which appears hypointense on T2W (b) and T1W (c) MRI at T4 vertebral level. Axial postcontrast MRI (d) shows severe compression and displacement of the spinal cord to the left anterolateral aspect (arrowhead) by the enhancing lesion (arrow). Axial T2W MRI (e) shows hypointensity of the lesion filling the canal (arrow) and no hyperintense fluid signal of surrounding CSF is seen as it is obliterated. All of the above MR sequences show a slightly T1 and T2 hyperintense, enhancing lesion within the T4 vertebral body (thick arrow), which could introduce the differential diagnosis of vertebral bony and spinal canal metastatic components. Axial bone window computerized tomography scan (f) image, however, clearly shows that the bony lesion is just an incidental vertebral body hemangioma (thick arrow) with characteristic "polka-dot" appearance of its vertical striations.
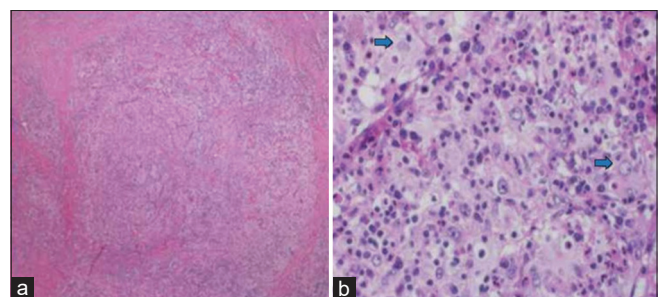

Figure 8: (a) Photomicrograph showing diffuse lymphohistiocytic proliferation in collagenous stroma (hematoxylin and eosin stain ×40). (b) High-power view shows histiocytic proliferation along with diffuse lymphoplasmacytic infiltrate. Some histiocytes with engulfed intact lymphocytes are present (blue arrows) (hematoxylin and eosin stain ×200).

thoracic spinal cord to the left anterolateral aspect. The patient underwent T7-11 laminotomy and excision of extradural mass. Histopathology showed a dura-based lesion composed of histiocytic proliferation with lymphoplasmacytic infiltrate along with lymphophagocytosis, compatible with RDD. Postoperative MRI showed no residual tumoral tissue. The

patient showed improvement in his power postoperatively, subjected to with rehabilitation and was discharged with mild spasticity of both lower limbs with power 5/5 on the latest outpatient follow-up visit.

DISCUSSION

RDD is a benign, nonneoplastic histiocytic proliferative disease characterized by prominent cervical lymph node enlargement; however, cases also illustrated that RDD might involve multiple organ systems and be indeed fatal.[8] Extranodal involvement is subsequently recognized in 43%. The most common sites of extranodal involvement are skin, upper respiratory, orbits, testes, and bones.[11] Bone involvement is rare, occurring in fewer than 10% of patients, while primary bone involvement has been reported in only a handful of patients.[1,3,5] In less than 5% of the extranodal RDD cases, CNS involvement has been previously reported and disease is usually suspected according to a typical clinical presentation which is painless lymphadenopathy and the diagnosis is confirmed by biopsy.[25] However, 65.5% of cases of RDD with CNS involvement are isolated, with no lymphadenopathy or other extranodal sites involvement.[25] During literature review, we have found 62 cases with spinal RDD, including two cases in our series. The mean age of spinal RDD is 34.7 years (range 2.5–78 years) with a male predominance (M/F = 1.48) with 71.4% of cases of spinal RDD being isolated, 21.4% of cases have associated lymphadenopathy, and 19.7% of patients have intracranial lesions simultaneously.[28] Before our case (case-no.1), only one case of skull involvement reported by Allegranza *et al*. in 1991,[3] making it be an extremely rare occurrence.

The etiology of RDD remains unclear since its recognition in 1969 and infectious etiology is the aspect that attracts most of the attention.[11] Human herpesvirus-6 genome has been demonstrated to present in lymph nodes affected by RDD.[9] Some authors also reported that abnormal immunologic response was another etiology, such as Becroft *et al*.,[7] demonstrated a defect in cellular immunity which promotes monocyte recruitment from the circulation into the nodal or extranodal sites and expressed that it is important in the histiocytic reaction. The rarity of osseous RDD and its dense inflammatory component often causes diagnostic difficulties and it is often confused with other lesions, such as lymphoma, LCH, osteomyelitis, storage diseases, and sarcomas.[3] Despite all these assumptions, the exact pathogenesis of RDD still remains controversial.

The patient with cranial RDD is often free from the systemic symptoms and may present with only neurological symptoms.[2] Our case (no.1) presented with headache and right side hemiparesis while the second case presented with seizure attack, possibly due to compressive parenchymal phenomenon. Both of our spinal cases presented with progressive lower limb weakness which was proceeded by back pain. Laboratory findings are generally nonspecific with elevated inflammatory markers such as ESR and less consistently rheumatoid factor. Many patients demonstrated a polyclonal gammopathy on serum immunoelectrophoretic of IgG, IgA, and IgM4. With the exception of elevated ESR, no other abnormal laboratory data could be found, which are similar to the cases we presented.[19,20]

Intracranial or spinal RDD usually presents as extra-axial dural based lesion which is iso- to hypointense to gray matter on T1 and T2 W MRI, usually solitary and rarely multiple.[5,6] These findings could also be seen in other more common conditions like highly cellular tumors including meningioma, metastasis, lymphoma, leukemic deposits, and granulomatous lesions such as sarcoidosis or tuberculosis. RDD may rarely show enhancing dural tails mimicking meningioma.[13,20,21,30] RDD lesions may show mild blooming on susceptibility weighted imaging, diffusion restriction on diffusion-weighted imaging, and low rCBV in perfusion MRI.[16] However, these findings are variable, both of our intracranial RDD cases showed no diffusion restriction. Diffusion restriction is also commonly seen in lymphoma and meningioma. Lymphoma usually shows low rCBV, whereas rCBV is elevated in meningioma. Rare case of RDD with increased rCBV mimicking meningioma has been described in the literature, presumably due to the high positivity of CD34 and CD31 antibodies which are surrogate markers of intrinsic vascularization of lesions.[17] FDG PET uptake can be variable, with nodal and lacrimal disease usually showing avid FDG uptake, and other sites not much FDG avid.[22]

Histologically, RDD is characterized by proliferation of histiocytes that are large with round nuclei and abundant pale cytoplasm, along with diffuse lymphoplasmacytic infiltrate.[6,11,24] One of the characteristic features is the presence of lymphophagocytosis without cell destruction (emperipolesis). No caseating necrosis or epithelioid granulomas should be present. By immunoperoxidase stains, the histiocytes in RDD are typically positive for S100 and negative for CD1a. The histological differential diagnosis includes LCH, lymphoplasmacyte-rich meningioma, and hypertrophic pachymeningitis.[6] In LCH, the histiocytes are of Langerhans cell type which is characterized by having elongated nuclei with nuclear grooves. They are typically positive for CD1a and Langerin immunostains. Lymphoplasmacytic-rich meningioma is composed of meningothelial cells with intense lymphoplasmacytic infiltrate and the meningothelial cells are positive for EMA immunostain. Hypertrophic pachymeningitis is characterized by fibroblastic proliferation in collagenous stroma along with lymphoplasmacytic infiltrate. Some of these cases are IgG4-related sclerosing disease, in which the inflammatory infiltrate is rich in plasma cells with increase

in IgG4/IgG ratio.[19] BRAF mutations have been suggested to help differentiate RDD and LCH that remains unequivocal cases based on immunophenotype and morphological features. BRAF^{v600} mutation reported to be associated with RDD that may have future therapeutic implications in management of RDD. None of our cases have had genetic study as it is not yet available in our institution.[10]

Spontaneous resolution and stable asymptomatic disease are observed in about of 90% of cases. In cases without CSN involvement, surgical resection is justified both to remove compressive lesion and establish a histopathological diagnosis.[11] However, complete resection may not always achievable due to multiple factors such as multiple foci, adherence to underlying neural parenchyma, or invasion of surrounding critical structures.[2] Corticosteroid administration has shown some therapeutic benefits that is used in patients with systemic RDD, but the recurrence is seen once steroids are tapered off.[23,26] It is noted that extent of surgical resection is not associated with prognosis.[28] The use of adjuvant therapy does not improve prognosis but considered a palliative option in situation of subtotal excision or complicated clinical conditions.[28] For inoperable masses, radiotherapy can be used for local control of the disease. Newly reported therapies, including tyrosine kinase inhibitor imatinib and the anti-CD20 monoclonal antibody rituximab, have also been used in systemic RDD.[4,27]

Long-term prognosis is believed to be associated with a number of nodal groups and extranodal systems involved, indicating that patients with extranodal lesions will probably have a poorer prognosis than those with nodal diseases.[11,12] According to Xu *et al.*,[28] there is no difference in the prognosis between cases with isolated CNS RDD or those with intracranial lesions and cases with isolated spinal RDD.

CONCLUSION

Craniospinal RDD is a rare clinical condition and its clinical presentation mimics other compressive pathology. In isolated craniospinal involvement, it poses a significant diagnostic challenge due to its similarity with other neoplastic and inflammatory pathologies involving craniospinal axis. Some peculiar neuroradiological features may help to suspect a preoperative diagnosis; however, histopathological distinction remains the standard of diagnosis. Surgical option is deemed necessary to treat compressive parenchymal clinical presentation and establish a definitive diagnosis. Craniospinal RDD has shown a good clinical outcome with complete neuroradiological resolution and no recurrence in long-term follow-up.

Declaration of patient consent

Institutional Review Board permission obtained for the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Abdelwahab IF, Klein MJ, Springfield DS, Hermann G. A solitary lesion of talus with mixed sclerotic and lytic changes: Rosai-Dorfman disease of 25 years' duration. Skeletal Radiol 2004;33:230-3.
2. Adeleye AO. Diagnosis and management of Rosai-Dorfman disease involving the central nervous system. Neurol Res 2010;32:572-8.
3. Allegranza A, Barbareschi M, Solero CL, Fornari M, Lasio G. Primary lymphohistiocytic tumour of bone: A primary osseous localization of Rosai-Dorfman disease. Histopathology 1991;18:83-6.
4. Alqanatish JT, Houghton K, Bond M, Senger C, Tucker LB. Rituximab treatment in a child with Rosai-Dorfman disease and systemic lupus erythematosus. J Rheumatol 2010;37:1783-4.
5. Al-Saad K, Thorner P, Ngan BY, Gerstle JT, Kulkarni AV, Babyn P, *et al*. Extranodal Rosai-Dorfman disease with multifocal bone and epidural involvement causing recurrent spinal cord compression. Pediatr Dev Pathol 2005;8:593-8.
6. Andriko JA, 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;14:172-8.
7. Becroft DM, Dix MR, Gillman JC, MacGregor BJ, Shaw RL. Benign sinus histiocytosis with massive lymphadenopathy: Transient immunological defects in a child with mediastinal involvement. J Clin Pathol 1973;26:463-9.
8. Buchino JJ, Byrd RP, Kmetz DR. Disseminated sinus histiocytosis with massive lymphadenopathy: Its pathologic aspects. Arch Pathol Lab Med 1982;106:13-6.
9. Destombes P. Adenitis with lipid excess, in children or young adults, seen in the Antilles and in Mali. (4 cases). Bull Soc Pathol Exot Filiales 1965;58:1169-75.
10. Fatobene G, Haroche J, Hélias-Rodzwicz Z, Charlotte F, Taly V, Ferreira AM, *et al*. BRAF V600E mutation detected in a case of Rosai-Dorfman disease. Haematologica 2018;103:e377-9.
11. 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.
12. Geara AR, Ayoubi MA, Achram MC, Chamseddine NM. Rosai-Dorfman disease mimicking neurofibromatosis: Case presentation and review of the literature. Clin Radiol 2004;59:625-30.
13. Gupta K, Bagdi N, Sunitha P, Ghosal N. Isolated intracranial Rosai-Dorfman disease mimicking meningioma in a child: A case report and review of the literature. Br J Radiol 2011;84:e138-41.
14. 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;98:165-8.

15. Hargett C, Bassett T. Atypical presentation of sinus histiocytosis with massive lymphadenopathy as an epidural spinal cord tumor: A case presentation and literature review. J Spinal Disord Tech 2005;18:193-6.

16. Hingwala D, Neelima R, Kesavadas C, Thomas B, Kapilamoorthy TR, Radhakrishnan VV. Advanced MRI in Rosai-Dorfman disease: Correlation with histopathology. J Neuroradiol 2011;38:113-7.

17. Idir I, Cuvinciuc V, Uro-Coste E, Penna M, Boetto S, Cognard C, *et al*. MR perfusion of intracranial Rosai-Dorfman disease mimicking meningioma. J Neuroradiol 2011;38:133-4.

18. Kattner KA, Stroink AR, Roth TC, Lee JM. Rosai-Dorfman disease mimicking parasagittal meningioma: Case presentation and review of literature. Surg Neurol 2000;53:452-7.

19. Lu LX, Della-Torre E, Stone JH, Clark SW. IgG4-related hypertrophic pachymeningitis: Clinical features, diagnostic criteria, and treatment. JAMA Neurol 2014;71:785-93.

20. Ludemann W, Banan R, Samii A, Koutzoglou M, Di Rocco C. Cerebral Rosai-Dorfman disease. Child Nerv Syst 2015;31:529-32.

21. Lungren MP, Petrella JR, Cummings TJ, Grant GA. Isolated intracranial Rosai-Dorfman disease in a child. AJNR Am J Neuroradiol 2009;30:E148-9.

22. Raslan O, Ketonen LM, Fuller GN, Schellingerhout D. Intracranial Rosai-Dorfman disease with relapsing spinal lesions. J Clin Oncol 2008;26:3087-9.

23. Raveenthiran V, Dhanalakshmi M, Hayavadana Rao PV, Viswanathan P. Rosai-Dorfman disease: Report of a 3-year-old girl with critical review of treatment options. Eur J Pediatr Surg 2003;13:350-4.

24. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Arch Pathol 1969;87:63-70.

25. Sandoval-Sus JD, Sandoval-Leon AC, Chapman JR, Velazquez-Vega J, Borja MJ, Rosenberg S, *et al*. Rosai-Dorfman disease of the central nervous system: Report of 6 cases and review of the literature. Medicine (Baltimore) 2014;93:165-75.

26. Sita G, Guffanti A, Colombi M, Ferrari A, Neri A, Baldini L. Rosai-Dorfman syndrome with extranodal localizations and response to glucocorticoids: A case report. Haematologica 1996;81:165-7.

27. Utikal J, Ugurel S, Kurzen H, Erben P, Reiter A, Hochhaus A, *et al*. Imatinib as a treatment option for systemic non-Langerhans cell histiocytoses. Arch Dermatol 2007;143:736-40.

28. Xu H, Zhang F, Lu F, Jiang J. Spinal Rosai-Dorfman disease: Case report and literature review. Eur Spine J 2017;26:117-27.

29. Yang X, Liu J, Ren Y, Richard SA, Zhang Y. Isolated intracranial Rosai-Dorfman disease mimicking petroclival meningioma in a child. Case report and review of the literature. Medicine (Baltimore) 2017;96:e8754.

30. Zhu H, Qiu LH, Dou YF, Wu JS, Zhong P, Jiang CC, *et al*. Imaging characteristics of Rosai-Dorfman disease in the central nervous system. Eur J Radiol 2012;81:1265-72.