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Review article

Isolated Rosai-Dorfman disease of the spine: A systematic literature review

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ABSTRACT

Introduction: Rosai-Dorfman disease (RDD) is a rare non–Langerhans cell histiocytosis involving the central nervous system in 5% of cases. Spinal location occurs in less than 1% of extranodal RDD and can be responsible for neurological manifestations.
We present a systematic review of cases of isolated spinal RDD. We also report a new case of isolated spinal RDD revealed by spinal cord compression.
Materials and methods: The systematic review was performed according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guideline using the MEDLINE and SCOPUS databases and included case reports and case series describing isolated RDD of the spine.
Results: There were 53 patients with isolated spinal RDD (including our case). The mean age was 35.85±16.48 years. Neurological deficit was the most frequent clinical presentation (89%). RDD lesions were mainly located in the thoracic spine (51%), then the cervical spine (32%). The lesion was reported to be extradural (57%), intradural extramedullary (26%), intramedullary (7%), and in the vertebral body (10%). Histological examination showed emperipolesis in 73%. Histocytes were positive for S-100 protein in 83%.
Treatment was based on surgery 96%), radiotherapy, chemotherapy, and adjunctive steroid therapy were indicated in four, one, and eight cases. After a mean follow-up period of 14.84±13.00 months, recurrence of RDD was noted in 15%.
Conclusion: Spinal RDD is a rare condition, requiring meticulous histological examination for accurate diagnosis. Complete surgical resection is the treatment of choice. Adjuvant chemotherapy and radiotherapy can also be indicated in patients demonstrating partial improvement following surgery.

1. Introduction

RDD is a myeloproliferative disorder of histiocytes. It can be responsible for a broad spectrum of clinical manifestations [1]. It is typically revealed by massive and bilateral cervical lymphadenopathy [2]. Extranodal manifestations are present in 43% of patients, with spinal location observed in only 0.6–1% of cases [3]. Isolated spinal involvement is rare and can mimic spinal tuberculosis, meningioma, extradural hematoma, and other tumors [4].

The diagnosis of spinal RDD, which is based on histopathological and immunohistochemical examinations, can be challenging [4]. Histologically, the lesion biopsy shows abnormal histiocytes characterized by CD68+ and CD1a− markers [1].

The pathology of RDD is still not fully understood. It involves viral infections, autoimmune conditions, and gene mutations [2]. The clinical course can range from a self-limited process to disseminated refractory disease with increased mortality. Due to the heterogeneity of the disease, treatment options for RDD vary, including both local and systemic approaches [1].

This systematic review summarized isolated spinal RDD's clinical, radiological, and histopathological findings. We also report a case of an adult patient with an isolated spinal RDD revealed by spinal cord compression.

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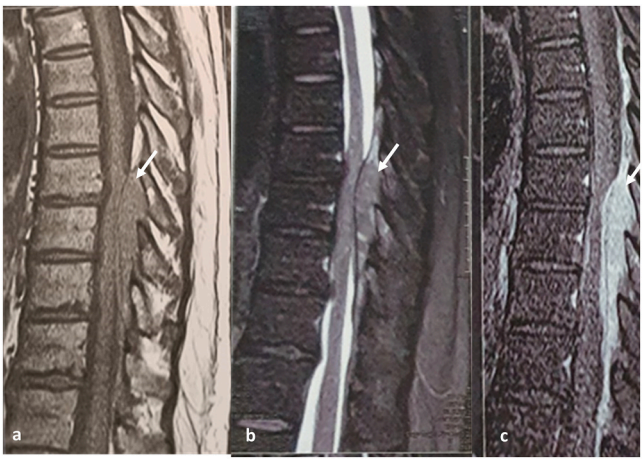

Fig. 1. : (a) Preoperative sagittal T1-weighted imaging (T1-WI) sequences magnetic resonance imaging of the dorsolumbar spine showing isointense extradural lesion extending from T8 to T10. (b) The lesion is isointense to the spinal cord in T2-weighted imaging. (c) T1-WI post gadolinium showing intense homogeneous enhancement of the lesion.

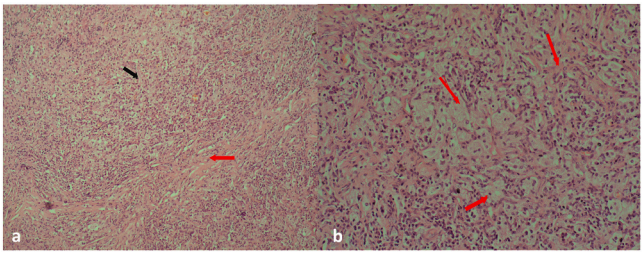

Fig. 2. : (a) X10: pseudo tumoral proliferation with low cell fibrosis (red arrow) and polymorphic inflammatory infiltrate (black arrow), (b) X20: inflammatory infiltrate with foamy histiocytes (red arrows).

2. Materials and methods

2.1. Case-report

We obtained written consent from the patient to publish this case report.

The following data were extracted from the patient's record: patient age and sex, initial symptoms, results of different investigations, and therapeutic management.

2.2. Search strategy for systematic review

2.2.1. Publication search

A systematic review was conducted following Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines [5].

The search included studies published in the SCOPUS and the MEDLINE databases until Mai 2023. We used the following keywords chosen from the Medical Subject Headings (MeSH) of MEDLINE:

(disease, rosai Dorfman$ [MeSH Terms]) AND (compression, spinal cord [MeSH Terms]);

(disease, rosai dorfman [MeSH Terms]) AND (spine [MeSH Terms]); (sinus histiocytosis with massive lymphadenopathy [MeSH Terms]) AND (compression, spinal cord [MeSH Terms]); (sinus histiocytosis with massive lymphadenopathy [MeSH Terms]) AND (spine [MeSH Terms]).

2.2.2. Inclusion criteria

The search included case reports and case series describing isolated spinal localization of RDD. Only English publications were included.

2.2.3. Exclusion criteria

We excluded cases with extraspinal manifestations of RDD. Non-full-text articles and duplicates were also excluded.

2.2.4. Data collection

Data collection was performed by two independent authors. Any discrepancies were resolved through a consensus discussion with a third author.

The following data were collected from each case report: gender, age at diagnosis, symptoms, laboratory investigations, histological findings, imaging features, treatment, and outcome.

Statistical analyses were performed using SPSS.

3. Results

3.1. Case presentation

A 50-year-old male patient with no medical history complained of a one-month history of progressive weakness and numbness in both lower limbs. He also reported gait unsteadiness. He denied bowel or bladder incontinence. There was no history of spinal trauma, fever, weight loss, or night sweats.

Physical examination revealed pyramidal signs in the lower limbs with proximal spastic paraparesis and sensory loss of all the dermatomes

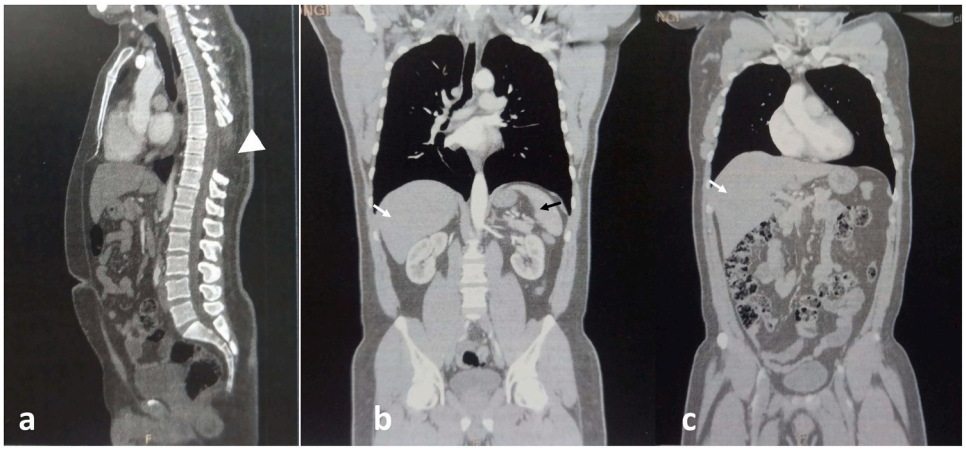

Fig. 3. : Abdominal, pelvic, and thoracic CT-Scan performed after surgical resection (arrowhead) showing no extra-spinal manifestations of RDD: no hepatomegaly (white arrow) and no splenomegaly (black arrow).

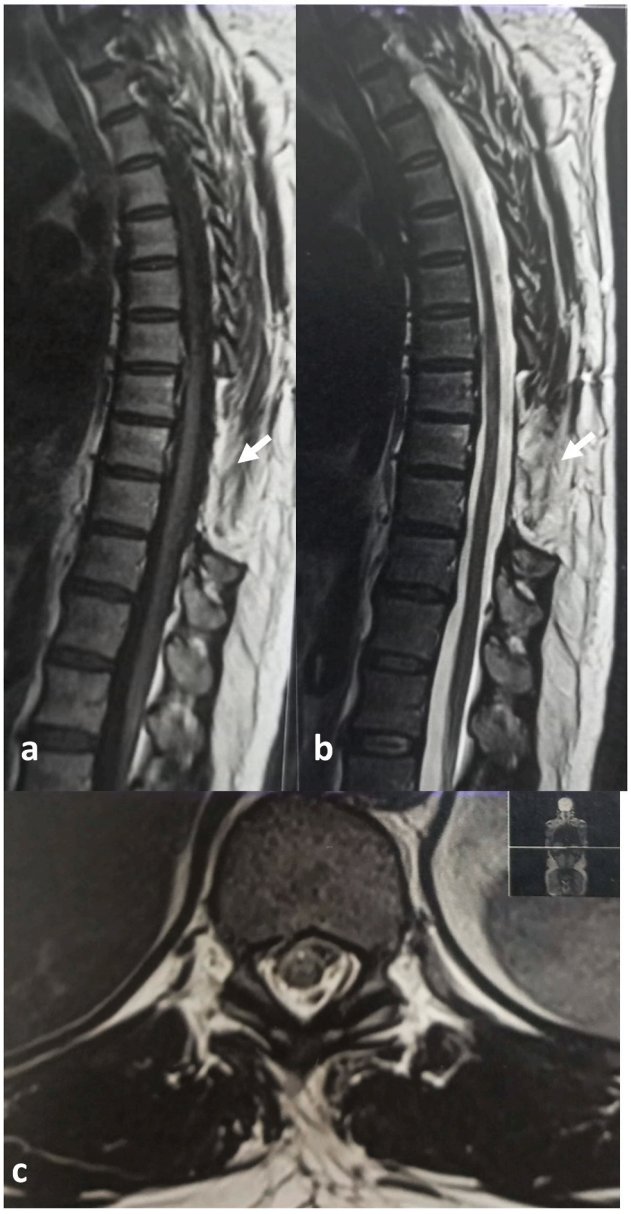

Fig. 4. : (a) Sagittal T1-weighted imaging (T1-WI) sequences magnetic resonance imaging of the dorsolumbar spine (b) Sagittal T2-WI sequences magnetic resonance imaging of the dorsolumbar spine (c) T2-WI axial view at the level of T9: the sequences are showing laminectomy with the absence of recurrence of the RDD lesion 18 months after surgery.

under T8. There were no lymphadenopathy, splenomegaly, or hepatomegaly. The physical examination was otherwise unremarkable. The complete blood count was without abnormalities. Inflammatory biomarkers, liver tests, and renal function were within normal ranges.

The spine magnetic resonance imaging (MRI) showed a thoracic extradural lesion dorsolateral to the spinal cord extending from T8 to T11 with signs of cord compression. The mass was isointense on T1 weighted imaging (WI), ~~and~~ hypo intense on T2WI, and marked gadolinium enhancement (Fig. 1).

The patient underwent urgent surgery consisting of T8-T11 decompressive laminectomy. The mass was strictly epidural and well-encapsulated without any bone infiltration, which helped with complete lumpectomy.

Histopathological findings showed a pseudo tumoral proliferation, which combines in variable proportions extensive low-cell fibrosis with an inflammatory infiltrate composed of plasma cells, lymphocytes, and essentially foamy histiocytes (Fig. 2).

These histiocytes had enlarged, round to oval hyperchromatic nuclei and abundant eosinophilic cytoplasm, often containing engulfed intact inflammatory cells known as emperipolesis. An immunohistochemical study showed a positive stain in histiocytes with PS100 antibodies.

The diagnosis of isolated RDD was established based on histological findings and the absence of visceral involvement. The chest-abdomen-pelvis CT scan did not reveal any other locations of RDD (Fig. 3).

The patient had an uneventful postoperative recovery and continued dexamethasone 12 mg daily starting a day before surgery for 10 days.

At the follow-up one month after the surgery, a physical examination showed a complete resolution of neurologic deficits. The spine MRI found no lesion's recurrence at 18 months after surgery (Fig. 4).

3.2. *Systematic literature review*

The initial search yielded 110 articles. Non-relevant and duplicated articles were removed (Fig. 5). The identified literature cases, including ours, are shown in Table 1 [6–51].

We included 46 articles reporting 52 patients with isolated spinal RDD. There were 53 patients, including our case: 36 males and 17 females. The mean age was 35.85 ± 16.48 years, ranging from 14 to 78 years. The delay between clinical signs onset and the diagnosis of RDD was 4.08 ± 5.03 months [0.2–30]. Clinical manifestations were spinal pain (58%, n=31) [9–13,15,21–23,25,26,29,32,33,35–38,40–42,45,47–49,51], radicular pain (n= 3) [47,49,50], neurological deficit (89%, n=47), and bowel or bladder dysfunction (24%, n=13) [18,21,26,29,32,34,36,39,43–45,50,51]. Laboratory findings were normal in 20 patients (38%). High erythrocyte sedimentation rate (ESR) was noted in 8 patients (15%) [8,18,20,21,24,32,48,50], with a mean ESR of 41.38 ± 20.69 mm [17–67]. Leukocytosis was reported in two patients [6,36]. One patient had a history of Hepatitis B viral infection [22].

Lesions were mostly located in the thoracic spine (n=27, 51%), then the cervical spine (n=17, 32%) and the lumbar spine (n=4, 8%). In three cases, RDD was in the sacrum (6%). In two cases reported, patients had multilevel spine lesions.

MRI was performed in all cases.

On T1-weighted image, RDD lesions were isointense in 18 patients (35%) and hypointense in five patients (10%). On T2-weighted images, RDD lesions were isointense, hypointense, or hyperintense in 9 (17%), 13 (25%), and 6 patients (11%), respectively. After the administration of gadolinium, RDD lesions had homogenous enhancement.

A dura tale sign was noted in three cases (6%) [9,11,33]. The lesion was extradural in 30 patients (57%), intradural extramedullary in 14 cases (26%), and intramedullary in four (7%). It was in the vertebral body in five cases (10%).

Six patients (11%) underwent a CT scan examination, revealing an osteolytic lesion [16,20,27,30,39,40]. PET scan was performed in two cases (4%) showing moderate FDG uptakes [14,16].

The diagnosis of RDD was based on histological findings in all patients. The results of the histological study were not detailed in three cases (6%). The emperipolesis phenomenon was described in 39 patients (73%). Histocytes positiveness for S-100 protein was the most frequent immunohistochemical finding (n=44, 83%), followed by positiveness for CD68 (n=30, 57%) and negativity for CD1a (n=26, 49%).

Surgical treatment was indicated in 51 cases (96%) with a complete resection of lesions in 69% (n=35) and incomplete *resection* in 21% (n=11). It was not specified if resection was complete or not in five cases (10%) [7,8,11,20,40].

Surgical treatment was associated with radiotherapy in four cases (8%) [23,26,35,45]. The doses and regimen of radiotherapy were as follows: 35 Gy in 20 fractions [23], 50.4 Gy in 28 fractions [35], 20 Gy in 10 fractions[45], and 12 Gy in 4 fractions [26]. *Chemotherapy* was associated in one case (2%), Vinblastin therapy was given intravenously (10 mg weekly) for 7 months [25]. Both radiotherapy and

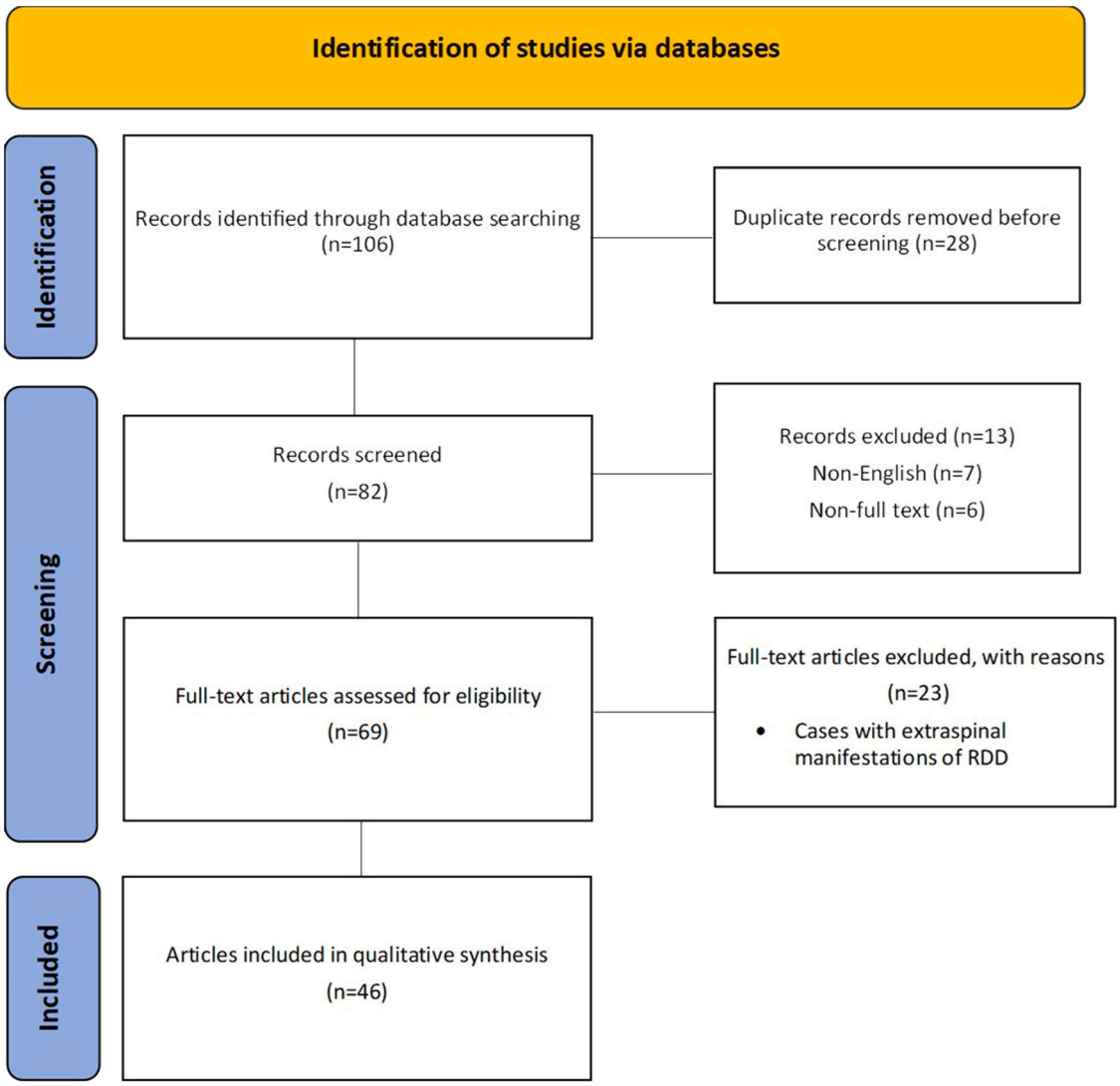

Fig. 5. : Flow chart for the study selection process.

chemotherapy were administrated immediately after surgery in all four cases. Steroids were prescribed in eight cases (15%) [6,14,16,37,45]. The exact dose of steroids was reported in three cases (6%). The mean dose of steroids was 48.8 ± 7.7 mg [40–53.3] of prednisolone per day. Steroids were prescribed for three months in two patients (4%) and for six months in the others (96%), with a gradual tapering. In one cases (2%), spontaneous regression of the lesion with reconstruction of the concerned vertebra was reported [30].

The mean follow-up period was 14.84 ± 13 months [2–60]. After therapeutic management, clinical and imaging improvement were noted in 41 patients (77%). Eight patients (15%) experienced a recurrence of RDD.

Among these patients, seven underwent surgery only and one received an associated radiotherapy [26].

The mean time to recurrence was 19.52 ± 21.64 months [2–60]. [10, 14,19,22,26,37,45,49]. It was not mentioned in one case.

The intradural extramedullary and intramedullary locations were associated with a high risk of recurrence (Odds ratio (OR): 3.91, p:0.048, $_{95\%}$Confidence interval [0.032; 1.08]). However, male gender (OR: 0.73, p:0.43), increased ESR (OR: 2.14, p:0.14), and the age (patients without recurrence: 35.87 years versus patients with recurrence: 38.86 years, p: 0.59) were not associated with recurrence.

The key findings for the entire cohort are summarized in Table 2.

4. Discussion

RDD was initially described by French pathologist Pierre Paul Louis Lucien Destombes in 1965 [52]. Then, Drs. Juan Rosai and Ronald Dorfman individualized it as a clinical entity [53]. RDD is a rare non-Langerhans cell histiocytosis, also called sinus histiocytosis with massive lymphadenopathy [54]. The pathogenesis of RDD remains unclear. Several studies highlighted the role of viral infections, such as herpes viruses, Epstein-Barr virus, cytomegalovirus, and human immunodeficiency virus (HIV) [55,56]. Hepatitis B viral infection was found in one of the reviewed cases [22]. However, data regarding the link between RDD and hepatitis B virus are scarce.

Other studies suggested that autoimmune dysregulation may contribute to the physiopathology of RDD. As for germline mutations in SLC29A3, which has also been reported in patients with familial RDD, suggesting that RDD may belong to a spectrum of disorders with SLC29A3 mutations [58].

RDD usually affects children and young adults, with a mean age at diagnosis of 20.6 years. A male predominance has been reported with a male-to-female ratio of 3:2. These findings are close to those found with isolated spinal RDD [2].

RDD is characterized by atypical expansion of histiocytes, presenting emperipolesis of leukocytes, associated with variable mixed inflammatory cells [2]. Emperipolesis is an uncommon process in which a normal

Table 1
Cases of isolated spinal RDD reported in the literature.

Authors (year)	Gender (age (years))	Symptoms Duration (months)	Laboratory findings	Level	Radiological findings	Diagnostic confirmation	Treatment	Follow-up (months) Outcome
Chen C W R et al. [6] (2012)	Female (16)	Progressive numbness of both legs with urinary incontinenceNS	Leukocytosis 17 × 109/l.	T4-T5	**MRI:** Intradural extramedullary Intermediate T2- and T1-WIsignal intensity	Emperipolesis	SteroidsComplete resection	24No recurrence
Andriko JA et al. [7] (2001)	Male (35)	Weakness and numbness of lower extremities1	NS	T4-T5	**MRI:** Intramedullary	Nodular cellular infiltrates of histiocytes, plasma cells, and lymphocytes Emperipolesis**IHC**: S-100 protein (+), CD68 (+)	Resection	12Improvement
	Male (42)	Weakness and numbness of lower extremities1	NS	T6–T8	**MRI:** extradural	NS	Complete resection	38No recurrence
Osenbach R et al. [8] (1996)	Male (35)	Progressive numbness and weakness of lower extremities 1	Elevated ESR (35 mm/h)	T4–5	**MRI:** Intramedullary	A lymph histiocytic infiltrate with plasma cells **IHC**: S-100 protein (+)	Resection	12ImprovementNo recurrence
Wu L et al. [9] (2014)	Male (43)	Spinal pain Progressive numbness in both hands8	Normal	C5– C6	**MRI:** Intradural extramedullaryHomogeneous enhancement A dura tail sign	Mixed inflammatory infiltrates of lymphocytes, plasma cells, and histiocytes in fibrous tissueEmperipolesis	Complete resection	18 ImprovementNo recurrence
Tian Y et al. [10] (2015)	Male (43)	Spinal painNumbness in both hands **8**	NS	C5-C6	**MRI:** extradural	Many lymphocytes, plasma cells, and foamy histiocytes **IHC**: CD68(+), lysozyme (+), S100 (+), EMA (-)	Complete resection	22Recurrence
Zhu H et al. [11] (2012)	Male (58)	Spinal pain and hypnalgia NS	NS	T8-T10	**MRI:** Intradural extramedullaryHyperintensity on T1-WI, iso intensity on T2-WI, with homogeneous enhancementA dura tail sign	Eosinophilic cytoplasm, in the background of many plasma cells and lymphocytesEmperipolesis**IHC**: S100 (+) CD1a (−)	Resection	NS
Fu X et al. [12] (2015)	Female (25)	Spinal pain Weakness and numbness in both upper limbs Gait unsteadiness **6**	NS	C3-C6	**MRI:** Intradural extramedullaryHypointense on T1-WI, hypointense on T2-WI, homogeneous enhancement	Fibrosis with lymphocytes, plasma cells, and neutrophilsEmperipolesisRussell bodies**IHC**: S-100 protein (+), CD68 (+), CD1a (-), pan-cytokeratin (AE1/AE3) (-), EMA (-), GFAP (-), CD30 (-), CD20 (-), CD3 (-), CD34 (-)	Complete resection	12 No recurrence
Kim DY et al. [13] (2014)	Male (15)	Spinal pain 6	Normal	T6/ T12	**Plain radiographs:** osteolytic lesion of T6 and T12 **MRI:** 2 lesions Hyperintense on T2-WI, homogeneous enhancement	Fibro-osseous tissue with large histiocytes Emperipolesis**IHC**: CD68 (+), S-100 (+), CD1a (-)	Complete resectionReconstruction	12No recurrence
Sciacca S et al. [14] (2015)	Female (75)	Lower limb spastic paraparesis and saddle paresthesiaNS	NS	T9-T11	**MRI:** extradural**PET-CT:** low-grade metabolically active disease confined to T10	Mixed lymphoplasmacytic infiltrate **IHC**: C20 (+), C79a (+), CD3 (+), CD68 (+), CD138 (+), CD56 (-), cyclin D1 (-)	SteroidsIncomplete resection	2Recurrence
Li Y et al. [15] (2018)	Male (28)	Spinal painBilateral lower limb numbness and weakness 12	NS	T6-T9	**MRI:** extraduralIso intense on T1-WI, hypo intense on T2-WI, homogeneous enhanced lesion	Diffuse infiltration of plasmocytes and lymphocytesEmperipolesis**IHC**: CD68 (+), CD21 (+), CD21 (+), CD3 (+), S-100 (+)	Incomplete resection	6Improvement No recurrence
Jayaram A et al. [16] (2020)	Male (24)	Progressive bilateral lower limb weakness 1/2	NS	C7-T4	**CT**: Focal cortical destruction of the T2 vertebra**MRI:** extraduralIsointense on T1-WI and iso to hypointense on T2-WI **PET-CT**: moderate uptake	NS	Steroids Complete resection	2Improvement
James J et al.	Male (35)	Progressive weakness, and numbness in both lower limbs 2	Normal	C3-L1	**MRI:** ExtraduralSpinal cord compression	Histiocytes, lymphocytes, numerous plasma cells, neutrophils, and proliferating blood	Steroids	NSImprovement

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Table 1 (*continued*)

Authors (year)	Gender (age (years))	Symptoms Duration (months)	Laboratory findings	Level	Radiological findings	Diagnostic confirmation	Treatment	Follow-up (months) Outcome
[17] (2017)						vesselsEmperipolesis **IHC**: Histiocytes S100 (+), CD 68 (+) and CD1a (-)		
Rocha-Maguey et al. [18] (2016)	Female (27)	Progressive paraparesis Bladder and bowel symptoms2	Increased ESR (25 mm/h)	C7-T1	**MRI:** Intramedullary Hypointense signal on T2-WI, and homogeneous enhanced lesion	A proliferation of large histiocytesEmperipolesis **IHC**: CD45 (+), CD68 (+), CD60 (+), CD30 (+), CD15 (+), latent membrane protein-1 antibody (-)	Complete resection	6Improvement
Lima et al. [19] (2016)	Female (50)	Progressive spastic paraparesis1/2	NS	T4	**MRI:** Intradural extramedullary A homogeneous enhanced lesionDural tail sign	NS	Complete resection	NSRecurrence
Jiang H et al. [20] (2022)	Female (25)	Spinal pain1	Increased ESR (64 mm/h)	L2-L3	**CT:** bone destruction in the right pedicle of L3**MRI:** Enhanced signal in the vertebral body of L2-L3 and the right pedicle of L3	Numerous foamy histiocytic cells, lymphocytes, and neutrophilsEmperipolesis **IHC:** EMA (+), vimentin (+), LCA (+), CD68 (+), CD79α (+), CD3 (+), S100 (+) CK (-), CD1a (-), Ki-67 (-), P53 (-), CD30 (-)	Resection	36Improvement No recurrence
Xu H et al. [21] (2017)	Male (34)	Spinal painProgressive bilateral lower extremity paresthesia Bladder and bowel dysfunction1	Increased ESR (17 mm/h)	T9–T10	**MRI:** ExtraduralIso-intense on T1WI, heterogeneously hypo-intense on T2WI, homogeneous enhanced lesion Spinal cord compression	Histiocytosis with an infiltrate of lymphocyte and plasma cell**IHC**: CD68 (+), S-100 (+)	Complete resection	5Improvement No recurrence
Lin CK et al. [22] (2019)	Male (32)	Spinal painLower limb weakness3	Hepatitis B virus infection	T11–12	**MRI:** Intradural extramedullaryVertebral body	Emperipolesis **IHC**: S-100 (+)	Incomplete resection	36Recurrence
Baassiri W et al. [23] (2019)	Male (54)	Spinal painBilateral hand numbness Difficulty in swallowing and dysphonia6	NS	Foramen magnum-C1	**MRI:** ExtraduralLocal meningeal enhancement following gadolinium administration	Sheets of foamy histiocytes, multinucleated giant cells, lymphocytes, and plasma cells Emperipolesis **IHC**: CD68 (+), S-100 (+), CD1a (-)	Incomplete resectionRadiotherapy	Improvement
Kozac B et al. [24] (2015)	Male (26)	Progressive bilateral lower extremity numbness, weakness, and gait difficulty1/2	Increased ESR (19 mm/h)	T1-T5	**MRI:** ExtraduralIso-intense to the spinal cord on T1WI, heterogeneously hypo-intense on T2WI with homogenous enhancement Spinal cord compression	Fibroconnective tissue with a lymphoplasmacytic infiltrate and clusters of atypical histiocytes Emperipolesis**IHC**: S-100 (+), CD1a (-)	Complete resection	18ImprovementNo recurrence
Kumar A et al. [25] (2020)	Male (35)	Spinal painParaparesis 2	Normal	C6-T7	**MRI:** Extradural Hypo-intense on T1- and T2-WISpinal cord compression	Inflammatory neoplastic tissue composed of clusters of large macrophages Emperipolesis**IHC**: CD68 (+), S-100 (+), CD1a (-)	Complete resectionFixationChemotherapy	24 ImprovementNo recurrence
Hargett C et al. [26] (2005)	Female (29)	Spinal painLower-extremity numbness, and weakness Bladder and bowel dysfunction1	NS	T5-T9	**MRI:** Extradural Slight enhancementSpinal cord compression	A granulomatous mass with eosinophilic histiocytes, lymphocytes, and plasma cellsEmperipolesis **IHC**: S-100 (+)	Complete resectionFixationRadiotherapy	3Improvement Recurrence
Robert EJ et al. [27] (2006)	Female (23)	Progressive left leg pain and weakness12	NS		**CT:** osteolytic lesion**MRI**: a large sacral lesion infiltrating the soft tissues	CT-guided biopsy procedureEmperipolesis	Complete resectionFixation	12Improvement No recurrence
Ma J et al. [51] (2008)	Male (44)	Spinal pain Progressive weakness of both lower limbsBladder and bowel dysfunction 6	Normal	L3	**Plain radiographs:** a lytic process **MRI:** Intradural, extramedullary 4 lesions Hypo intensity on both T1- and T2-WI,	Large lymphoplasmacytic areas infiltrated with histiocytic cellsEmperipolesis **IHC**: CD68 (+), S100 protein (+), CD1a (-)	Complete resection	12ImprovementNo recurrence

(*continued on next page*)

Table 1 (*continued*)

Authors (year)	Gender (age (years))	Symptoms Duration (months)	Laboratory findings	Level	Radiological findings	Diagnostic confirmation	Treatment	Follow-up (months) Outcome
					with homogeneous enhancementLumbar spine			
Wang Y et al. [28] (2010)	Male (58)	Leg weakness 3	NS	T8-T10	**MRI:** Intradural, extramedullarySlight enhancement after gadolinium infusion	A chronic inflammatory process containing a multinucleated giant cell**IHC**: CD68 (+), S100 protein (+), vimentin (+)	Complete resection	7Improvement No recurrence
Huang Y. C. et al. [29] (2006)	Female (31)	Spinal painLegs weakness, and numbnessBladder dysfunction Relapsing uveitis3	Normal	T6–8	**MRI:** ExtraduralHomogeneous enhancementSpinal cord compressing	Clusters of large eosinophilic histiocytes and interspersed lymphocytes, plasma cellsEmperipolesis **IHC**: S-100 protein (+)	Complete resection	12Improvement No recurrence
Ramon A et al. [30] (2020)	Male (18)	Spinal pain 3	Normal	T9	**CT:** osteolysis of the posterior arch T9 vertebra**MRI:** ExtraduralInflammation without spinal cord compression	Emperipolesis **IHC**: CD163 (+), S100 (+), CD1a (-), ALK1 (-), phosphorylated extracellular signal-regulated kinase (+)	None	3ImprovementSpontaneous reconstruction of the T9 vertebra
Chhabria BA et al. [50] (2018)	Female (19)	Spinal pain Radicular radiation to both lower limbs, and asymmetric weakness Bowel and bladder dysfunction3	Normocytic, normochromic anemia (10 g/dL)Elevated ESR (67 mm/h)		**MRI:** ExtraduralLesion involving the nerve roots of the cauda equina extending up to the sacral level	A histiocyte-rich lesion with lymphoplasmacytic infiltrate Emperipolesis **IHC**: CD68 (+), S-100 (+), CD1a (-)	Incomplete resection	NSImprovement
El Molla M et al. [31] (2014)	Male (76)	Progressive right arm weakness and right footdrop2 and 1/2	NS	C2-C4	**MRI:** Intramedullary Spinal cord compression	Dense fibrosis and intense chronic inflammation with focal neutrophilic infiltrates**IHC**: S-100 (+)	Complete resection	12Improvement No recurrence
Hollowell J. P et al. [32] (2000)	Male (78)	Spinal pain Loss of sensation and strength in the left arm and the legsBladder dysfunction6	Elevated ESR (42 mm)	Cervical, thoracic, and lumbar spine	**MRI:** ExtraduralAn intermediate isointense signal on T1WI and diffuse low signal on T2WI with heterogenous enhancement Spinal cord compression	A dense collagenous matrix containing histiocytes, plasma cells, and lymphocytes Emperipolesis**IHC**: CD68 (+), KP1 (+), vimentin (+), Pan-cytokeratin (+), CAM-5.2 (-), LMW cytokeratin (-), SMA (-), HMB45 (-), mucin (-)	Incomplete resection	18Improvement
Tu J et al. [33] (2017)	Male (41)	Spinal pain 12	NS	C7–T7	**MRI:** ExtraduralIsointense on T1-WI and hypointense on T2-WIA dura tail sign	Fibrous connective tissue with diffuse or nodular tissue cell, lymphocyte, and plasma cell infiltrationEmperipolesis**IHC**: CD68 (+), CD163 (+), S100 (+), CD1a (-), CD34 (-)	Complete resectionFixation	3Improvement
Bhandari A et al. [34] (2006)	Female (23)	Progressive weakness of both lower limbsBowel and bladder dysfunction30	Normal	C3–6, D1–4, D5	**MRI:** Intradural, extramedullary 3 lesionsHypointense on T1-WI and T2-WI with marked homogeneous enhancement	Lymphoplasmacytic infiltrate with intervening histiocytic cells **IHC**: S-100 (+)	Complete resection	5Improvement No recurrence
Roy C et al. [35] (2012)	Male (32)	Spinal pain Asymmetric spastic paraparesis2	Normal	T11-L2	**MRI:** ExtraduralIsointense on T1-WI and hyperintense on T2-WI and STIR	Histiocytes admixed with lymphocytes and plasma cells**IHC**: S-100 (+)	Complete resectionRadiotherapy	3Improvement
Abou-Zeid A. H et al. [36] (2010)	Male (24)	Spinal pain Bilateral lower-limb weaknessBladder dysfunction1 week	Leukocytosis 13.1 × 109 /L	T4-T7	**MRI:** Extradural Isointense on T1- and T2-WI with diffuse enhancement after gadolinium	A histiocytic lesion in a fibrous background admixed with lesser populations of T and B lymphocytes and plasma cellsEmperipolesis**IHC**: CD68 (+), S100 (+), CD1a (-)	Incomplete resectionFixation	18Improvement No recurrence
Al Saad K et al. [37] (2005)	Male (17)	Spinal pain Leg numbnessGait unsteadiness 3	Normal	T8-T10	**MRI**: ExtraduralA T9 vertebral lesion	Neutrophils, lymphocytes, plasma cells, and histiocytesEmperipolesis**IHC**: CD68	Steroids Complete resection Reconstruction	8Recurrence

(*continued on next page*)

Table 1 (*continued*)

Authors (year)	Gender (age (years))	Symptoms Duration (months)	Laboratory findings	Level	Radiological findings	Diagnostic confirmation	Treatment	Follow-up (months) Outcome
						(+), S-100 protein (+), fascin (+), MAC387 (+)		
Huang B Y et al. [38] (2016)	Male (40)	Spinal pain Gait unsteadiness 12	Normal	C3-C5 and C6	**MRI:** ExtraduralMultiple mass lesionsHomogeneous enhancementSpinal cord compression	Large pale histiocytes with numerous macrophages, lymphocytes, and eosinophilic granulocytesEmperipolesis**IHC**: S-100 protein (+), CD1a (-)	Complete resection	18Improvement No recurrence
	Female (14)	Bilateral leg pain4	NS	S1-S2	**MRI:** Extradural Isointense on T1-WI and T2-WI, with homogeneous enhancement	Atypical lymph histiocytic infiltration Emperipolesis**IHC**: S-100 protein (+), CD68 (+)	Complete resection	12Improvement No recurrence
	Male (43)	Spinal pain Numbness in the upper limbs 8	NS	C5-C6	**MRI:** Extradural Isointense on T1-WI and T2-WI, with homogeneous enhancementSpinal cord compression	Emperipolesis**IHC**: S-100 protein (+), CD68 (+), EMA (-), CD1a (-)	Complete resection	12Improvement No recurrence
	Male (55)	Numbness of the lower limbs Gait instability1	NS	T1-T9	**MRI:** Extradural Isointense on T1-WI and slightly hyperintense on T2-WI, with homogeneous enhancement	Fibrous tissue infiltrated by lymphocytes, plasmocytes, and histiocytesEmperipolesis**IHC:** S-100 protein (+), CD68 (+), CD3 (+), CD20 (+), MUL-1 (+), CD1a (-)	Incomplete resection	6Improvement No recurrence
Elsotouhy A et al. [39] (2015)	Male (52)	Lower limb weaknessBowel and bladder dysfunction1/2	NS	T2–T5	**CT:** scoliotic deformity with maximum convexity at D4**MRI:** intradural, extramedullaryHypointense on T1-WI and T2-WI, with homogeneous enhancement	An aggregate of large histiocytes with plasma cells, lymphocytes, and rare neutrophilsEmperipolesis**IHC**: S-100 protein (+)	Complete resection	NSImprovement
Igrutinovic Z et al. [40] (2016)	Female (14)	Spinal pain 1	Normal	C4	**CT:** Osteolytic lesion of C4**MRI:** A spontaneous collapse of the C4 body	Atypical histiocytic proliferationEmperipolesis**IHC**: S-100 protein (+), CD68 (+), CD163 (-), CD1a (-)	Corporectomy Reconstruction	24Improvement No recurrence
Xin Y et al. [41] (2016)	Male (40)	Spinal pain Lower-limb weakness6	Normal	C3-C5C6-C7	**MRI:** Intradural extramedullary2 lesionsIsointense on T1-WI T2-WI with homogeneous enhancementDural tail sign	Lymphoplasmacytic areas with histiocytic cellsEmperipolesis**IHC:** S-100 protein (+), CD68 (+), CD1a (-)	Complete resection	24Improvement No recurrence
	Female (14)	Intermittent pain in the lower extremities4	Normal	S1-S2	**MRI:** Intradural extramedullaryIsointense on T1-WI T2-WI with homogeneous enhancement	Aggregates of pale-staining foamy histiocytesEmperipolesis**IHC:** S-100 protein (+), CD68 (+), CD1a (-)	Incomplete resection	12Improvement No recurrence
Nguyen PX et al. [42] (2021)	Male (19)	Spinal painBilateral leg weakness 2	Normal	T6-T9	**MRI:** Heterogeneous hyperintense on T2WI, isointense on T1WI and homogeneous enhancement	Emperipolesis**IHC:** S100 (+), CD68 (+), CD45 (+), CD1a (-), CD138 (−), GFAP (−)	Complete resection	4Improvement
Atal AA et al. [43] (2019)	Male (45)	Progressive spastic paraparesis Bowel and bladder dysfunction3	NS	T8-T9	**MRI:** extraduralIsointense on T1-WI, T2-WI with homogeneous enhancement	Numerous histiocytes with lymphoplasmacytic infiltratesEmperipolesis**IHC:** S-100 protein (+), CD1a (-)	Complete resection	NSImprovement
Karim R et al. [44] (2022)	Male (33)	Progressive quadriparesisBladder dysfunction1/2	NS	C4-T6	**MRI:** Extradural Isointense on T1-WI, iso-to- hyperintense on T2-WI	**IHC:** S100 (+), CD68 (+)	Complete resection	NSImprovement
Baeesa SS et al. [45] (2020)	Female (50)	Spinal pain Lower-limb weakness Bladder dysfunction2	Normal	T2-T4	**MRI:** ExtraduralHomogeneous enhancement	Acute and chronic inflammatory cell infiltrateEmperipolesis**IHC:** S-100 protein (+), CD1a (-)	SteroidsIncomplete resection	54Improvement No recurrence

(*continued on next page*)

Table 1 (*continued*)

Authors (year)	Gender (age (years))	Symptoms Duration (months)	Laboratory findings	Level	Radiological findings	Diagnostic confirmation	Treatment	Follow-up (months) Outcome
	Male (35)	Spinal pain Lower-limb numbnessNS	Normal	T2-T5	**MRI:** Extradural	Fibrosis with dense infiltration of a population of abnormal histocytesEmperipolesis**IHC:** S100 (+), CD68 (+)	SteroidsComplete resectionRadiotherapy	60Recurrence
Singh S et al. [46] (2019)	Female (15)	Progressive spastic quadriparesis2	NS	C3-C5	**MRI:** Intradural extramedullaryHomogenous enhancement	Histiocytes and multinucleated giant cells mixed with dense inflammatory infiltrates Emperipolesis**IHC:** S-100 protein (+), CD1a (-)	Complete resection	2Improvement
Parmar V et al. [47] (2013)	Male (64)	Spinal pain Radicular pain1	Normal	C5-C6	**MRI:** Intradural extramedullary	Fibrosis with mixed inflammatory infiltrates of lymphocytes, plasma cells, and histiocytes**IHC:** S100 (+), CD68 (+), CD1a (-)	Complete resection	9Improvement No recurrence
Maiti T k et al. [48] (2011)	Female (19)	Spinal painSpastic quadriparesis2	Anemia (10.8 g/dL)Elevated ESR (62 mm/h)	C2-C7	**MRI:** Extradural Homogenous enhancement	Soft tissue with dense aggregates of lymphocytes and histiocytesEmperipolesis**IHC:** S100 (+), CD68 (+), CD1a (-)	Complete resection	18 Improvement Doubtful recurrence
Syed S ALI et al. [49] (2014)	Male (28)	Spinal painRadicular pain3	NS	L3-L5	**MRI:** ExtraduralIsointense on T1-WI, hyperintense on T2-WIHomogenous enhancement	Histiocytes with mixed inflammatory cells composed of lymphocytes and plasma cells**IHC:** S100 (+), CD68 (+),	Incomplete resection	6Recurrence
Our case	Male(50)	Weakness and numbness in both lower limbs Gait unsteadiness1	Normal	T8-T11	**MRI:** ExtraduralIsointense on T1-WI, hypointense on T2-WIIntense enhancement	Fibrous tissue with histiocytes, lymphocytes, and plasma cellsEmperipolesis**IHC:** S100 (+), CK (-)	Steroids Complete resection	6Improvement No recurrence

CT: computed tomography; MRI: magnetic resonance imaging; PET-CT: positron emission tomography-computed tomography; NS: not specified; IHC: immunohistochemistry, T1-WI: T1-weighted image, T2-WI: T2-weighted image.

Table 2
Summary of the characteristics of the reviewed cases.

Patients' characteristics		n=52
Age, mean ± SD, years		35.85 ± 16.48
Male-to-Female ratio		2.11
Laboratory findings	High ESR, n (%)	8 (15)
	ESR, mean ± SD, mm	41.38 ± 20.69
Location	Extradural, n (%)	30 (57)
	Intradural extramedullary, n (%)	14 (26)
	Intramedullary, n (%)	4 (7)
	Osseous lesion, n (%)	6 (10)
Treatment	Surgery, n (%)	51 (96)
	Steroids, n (%)	8 (15)
	Chemotherapy, n (%)	1 (2)
	Radiotherapy, n (%)	4 (8)
Outcome	Improvement, n (%)	41 (77)
	Recurrence rate, n (%)	8 (15)
	Time to recurrence, mean ± SD, months	19.52 ± 21.64

ESR: erythrocyte sedimentation rate, SD: standard deviations.

cell penetrates another normal living cell [59].

The macrophage-dendritic cell lineage's histiocytosis and neoplasms encompass five groups, including RDD. This later is classified into five subgroups: classical (nodal) (1), familial (2), extranodal (3), neoplasia-associated (4), and immune disease-associated RDD (5) [60].

The third subgroup (extranodal RDD) can involve the skin (10%), subcutaneous tissue, deep soft tissue, bone (5–10% of cases), visceral organs, central nervous system (7%), upper respiratory tract, thyroid, and salivary glands [1,61].

Spinal RDD represents only 25% of extranodal RDD affecting the central nervous system. Spinal RDD can be spinal-dural or epidural. Reported cases of secluded spinal RDD are summarized in Table 1. We reported a case of a 50-year-old male patient with isolated RDD of the thoracic spine. Indeed, cervical and thoracic spine are the most common regions of RDD [1].

Clinical manifestations depend on the location and the size of the lesion. As shown in Table 1, all patients complain of signs of spinal cord compression, ranging from spinal pain to neurological deficit and cauda equina syndrome [18,20]. Laboratory findings do not seem to be specific [45]. They can reveal mildly increased ESR and leukocytosis [54].

Spine MRI can show spinal-dural or epidural lesions, which appear isointense or hypointense on T1-weighted images [38,39], and isointense, hypo, or hyperintense on T2- T2-weighted images [38,39]. After the administration of gadolinium, RDD lesions have homogenous enhancement [36]. A dura tail sign can also be seen, mimicking a meningioma [41]. The Dura tail sign was defined by Goldsher et al. as a thickening of the dura adjacent to the tumor and tapering away from it, seen as an enhancement more intense than the tumor, in at least two consecutive sections through the tumor at the same site in more than one imaging plane [62]. A CT scan can reveal osteolytic lesions of the concerned vertebra [27,40].

Histological findings showed fibrous tissue with mixed inflammatory infiltrates of histiocytes, lymphocytes, and plasma cells [7,11,32,38]. Emperipolesis is the hallmark of RDD [45]. It is different from phagocytosis because, during emperipolesis, histiocytes engulf intact cells, which remain viable and can even exit histiocytes [1]. Emperipolesis was present in 73% of the cases reviewed. Histocytes of RDD lesions exhibited positiveness for S-100 and CD68 and negativity for CD1a. The negativity for CD1a would distinguish RDD histiocytes from Langerhans cells [63].

Until this date, no guidelines were established for the management of spinal RDD. In the reviewed cases, surgery was performed in most cases (Table 1). A complete resection of RDD lesion was obtained in 68%. Other teams opted for an association of surgical treatment with radiotherapy, steroids, or chemotherapy. Radiotherapy and chemotherapy were indicated in patients with partial improvement in symptoms after surgery [23,26,35,45]. Since the RDD lesion was extensive (extending from C3 to L1 level), James J et al. reported treating it with steroids alone, which resulted in clinical improvement with radiological resolution of the spinal cord lesion [17]. Initial physical examination revealed spastic lower limbs with grade II power with remarkable improvement after steroids (the power of both lower limbs became Grade IV). In this case, the diagnosis was performed based on a biopsy taken from the lesion through a D1-D2 laminectomy [17]. Ramon A et al. reported spontaneous improvement of a spinal RDD lesion with spontaneous reconstruction of the concerned vertebra in an 18-year-old male patient [30]. Spinal RDD appears benign with a 70% improvement rate and only a 15% recurrence rate.

Our work is among the few systematic reviews emphasizing the clinical presentation, imaging features, and therapeutic management of isolated spinal RDD. However, our review has some limitations. First, we included studies with low class of evidence, such as case reports and small case series. Second, the publication bias was also anther limitation for our study as we only included published studies and we did not search for unpublished work.

5. Conclusions

This review focuses on spine RDD, a rare extranodal manifestation. Diagnosing isolated spinal RDD can be challenging due to the absence of common RDD manifestations such as massive cervical lymphadenopathy. The lack of specific imaging features highlights the importance of surgery, not only for lesion removal but also for ruling out alternative diagnoses. The choice of optimal adjuvant treatments requires further research.

Ethics approval

Our study complies with the Declaration of Helsinki.

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Declaration of Competing Interest

None.

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Informed consent

Written informed consent was obtained from the patient.

Consent for publication

Written informed consent was obtained from the patient to publish this case report.

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