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Case Report

Intradural extramedullary eosinophilic granuloma of the spine with emergency presentation: A case report

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ABSTRACT

Background: Intradural extramedullary spinal eosinophilic granuloma is a very unusual manifestation of Langerhans Cell Histiocytosis (LCH) that is typically misdiagnosed due to its nonspecific clinical and radiological features.

Case Description: We report a case of a 22-year-old female patient who presented with acute onset paraplegia. The magnetic resonance imaging was initially suggestive of tuberculoma, which is prevalent in tuberculosis-endemic regions. Intraoperative findings and histopathology, however, established the diagnosis of LCH. The lesion was intradural, and CD1a and S-100 protein staining demonstrated classic Langerhans cells.

Conclusion: The paper stresses the need for a multidisciplinary team in the proper diagnosis and handling of spinal LCH. Further research is needed to develop optimal management protocols for this rare condition.

Keywords: Histopathological diagnosis, Intradural extramedullary eosinophilic granuloma, Langerhans cell histiocytosis, Spinal lesion, Tuberculoma

INTRODUCTION

Intradural extramedullary spinal eosinophilic granuloma is a rare condition with difficult diagnostic strategies. The lesions may be mistaken for other pathology, for example, tuberculomas, in regions of the world where tuberculosis is endemic. Langerhans cell histiocytosis (LCH) is an abnormal proliferation of Langerhans cells, and these cells are specialized dendritic cells that are normally found in the epidermis. LCH can have a broad spectrum of presentations, from single lesions to multisystem disease, and although predominantly a disease of children, it does affect adults. Spinal involvement and an intradural extramedullary location are exceedingly unusual and may simulate other pathologies both clinically and radiologically.[9,13,16] The uncommon nature and nonspecific presentation of spinal LCH often lead to delayed diagnosis and treatment. Patients may present with a broad range of symptoms, including back pain, neurological deficits, and features suggestive of spinal cord compression. Imaging modalities like magnetic resonance

imaging (MRI) are significant in recognizing these lesions, but definitive diagnosis typically requires histopathological examination. The differential diagnosis for spinal lesions would include infection (e.g., tuberculosis), inflammatory processes, and neoplastic disease.[18,24,25] The recent literature has emphasized the importance of early diagnosis and treatment in improving patient outcomes in LCH. Newer imaging techniques, including diffusion-weighted MRI and positron emission tomography-computed tomography, have been useful in differentiating LCH from other spinal pathologies. Furthermore, multidisciplinary input from radiologists, pathologists, and oncologists are important in the management of such difficult cases. Early intervention can prevent crippling neurological disability and improve the quality of life for such patients.[29]

In this report, we have a case of a 22-year-old female with intradural extramedullary eosinophilic granuloma of the spine. This case highlights the importance of LCH in the differential diagnoses for spinal lesions and re-emphasizes histopathology in arriving at a proper diagnosis.

CASE DESCRIPTION

A 22-year-old female presented with 1 week of constipation, followed by paresthesia in both lower extremities that progressed to paraparesis and then paraplegia over 5 days. The patient also reported a disturbed sensory level to the umbilicus. Severe neurological deficits were present on initial physical examination, and imaging studies were promptly ordered.

MRI of the spine was performed, which revealed a lesion at the T9 level. The lesion extended from the extradural space to the posterior bony elements of the lamina and spinous process, with further extension into the paraspinal muscles. On T2-weighted MRI, the lesion appeared hypointense, raising suspicion for a granulomatous process [Figures 1a-d and 2]. The pattern of enhancement was ring-like, which is often suggestive of an infectious etiology, such as tuberculosis, which is endemic in Iraq. However, the differential diagnosis also included other possibilities, such as neoplastic and inflammatory conditions.

The patient underwent emergency surgery due to the rapid progression of neurological symptoms. Intraoperatively, the lesion was found to be intradural rather than extradural, as initially suspected. The mass was noted to be firm and fibrous with clear demarcation from the surrounding spinal cord. Multiple biopsies were taken, and the lesion was totally resected to decompress the spinal cord [Figures 3a-e]. Postsurgery, she was referred to hematology for continued treatment.

Histopathological examination of the resected tissue revealed a proliferation of Langerhans cells with characteristic grooved, coffee bean-shaped nuclei [Figure 4a]. Immunohistochemical staining was positive for S-100 protein [Figure 4b] and CD1a [Figure 4c], confirming the diagnosis of LCH. The lesional cells were negative for glial fibrillary acidic protein [Figure 4d], excluding neural tumors, and negative for epithelial membrane antigen [Figure 4e], excluding metastatic tumors in the differential diagnosis. The patient experienced complete resolution of paresthesia on the 1st postoperative day and regained sphincter control by the end of the 1st week. By 1 month, she could stand and walk, and by 3 months, she was walking normally.

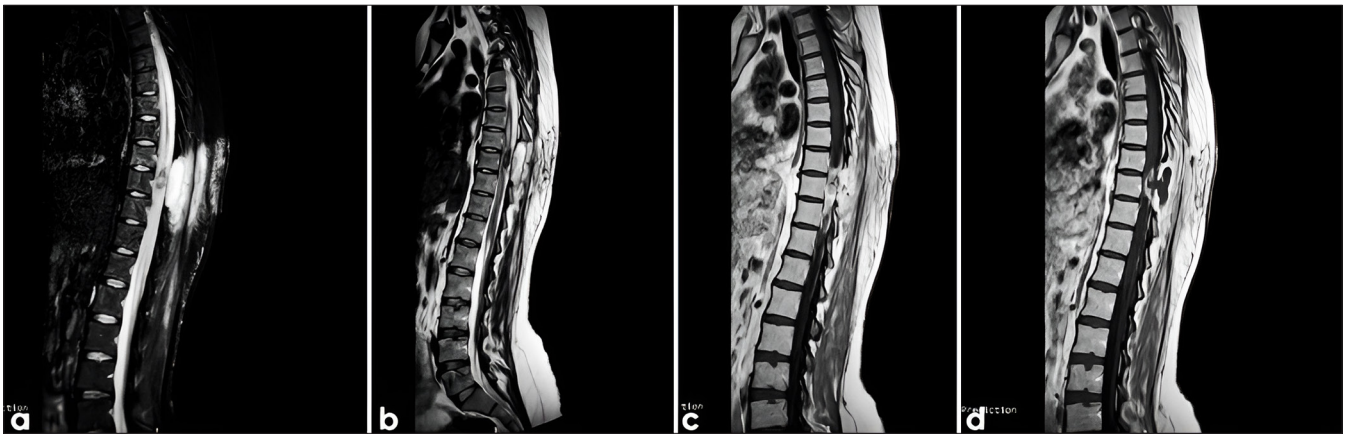

Figure 1: (a) Preoperative (Preop) Sag-short tau inversion recovery. Shows the mass at the T9 level, with suspicion of extradural as there is an involvement of the spinous process and paraspinal soft tissues. (b) Preop Sag-T2 shows a mass in the T9, which is an intradural extramedullary lesion. (c) Preop Sag-T1-postcontrast shows the enhancement of the mass in a heterogeneous pattern with the enhancement of the spinous process and surrounding muscles up to the fascia. (d) Preop. The pattern of enhancement is ring enhancing, raising suspicion of infection and, most likely, TB, as it is endemic in our country.

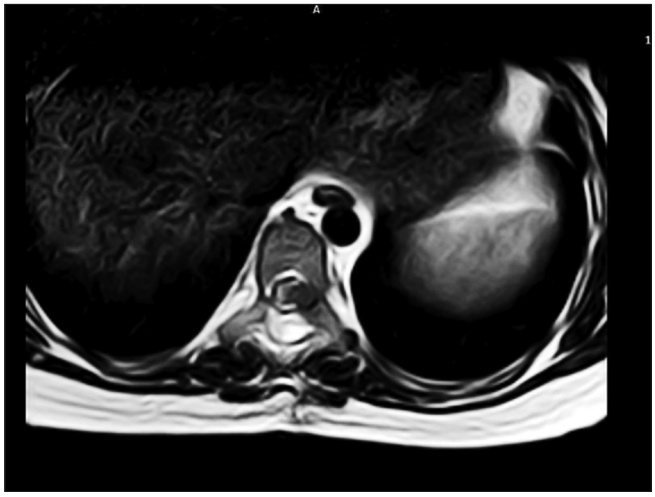

Figure 2: Preop axial-T2 showing the mass is hypointense and left-sided, displacing the spinal cord to the right.

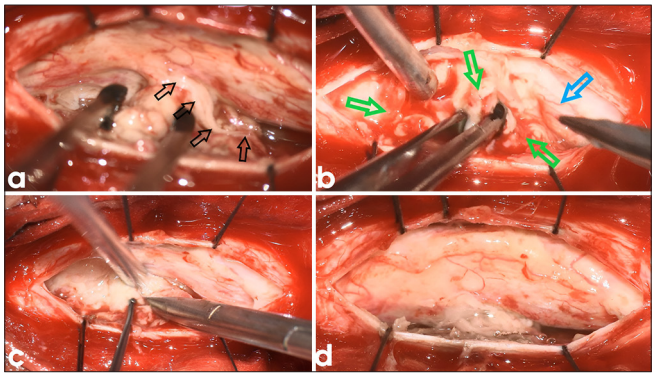

Figure 3: (a) Intraoperative image demonstrating the tumor (black arrows) compressing and displacing the spinal roots. (b) Intraoperative image showing meticulous dissection and preservation of an intact spinal root (green and blue arrows). (c) Intraoperative image revealing a fibrous granuloma that was initially biopsied for pathological evaluation. (d) Intraoperative image depicting the gross total resection of the lesion, ensuring decompression of the spinal cord.

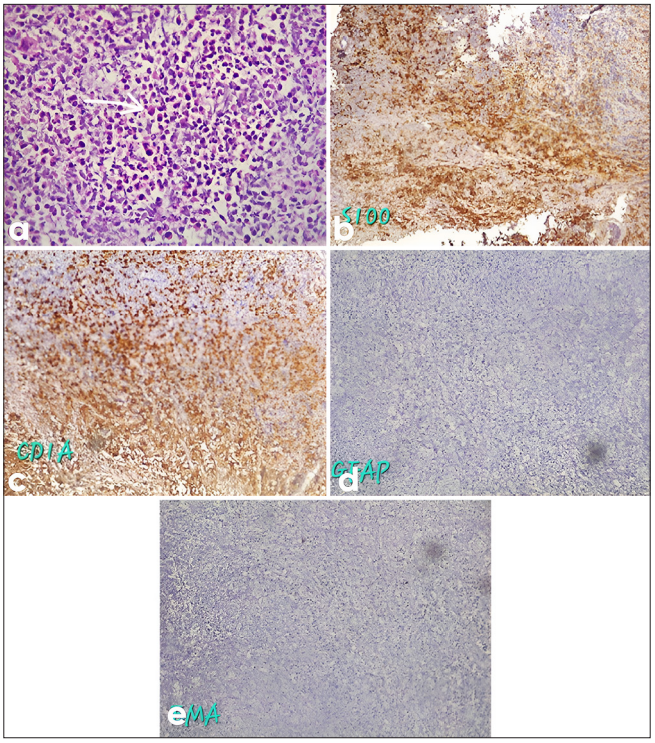

Figure 4: (a) Hematoxylin and eosin (H&E) staining showing an infiltrate of eosinophils mixed with histiocytic cells. A Langerhans cell with a characteristic grooved, coffee-bean-shaped nucleus is indicated by the arrow, supporting the diagnosis of Langerhans cell histiocytosis (LCH), ×400 (Arrow). (b) Immunohistochemistry (IHC) demonstrating strong diffuse positivity for S-100 protein in lesional cells, confirming the presence of Langerhans cells, ×200. (c) CD1a IHC stain showing strong membrane positivity in lesional cells, a hallmark feature of LCH, ×200. (d) Glial fibrillary acidic protein (GFAP) IHC stain demonstrating negative staining, excluding glial-derived tumors, ×200. (e) Epithelial membrane antigen (EMA) IHC stain showing negative staining, helping to exclude metastatic carcinoma from the differential diagnosis, ×200.

Systematic review

Methodology

A systematic review was conducted to evaluate the management and outcomes of spinal (LCH), following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines.[36] A comprehensive literature search was performed using PubMed and Scopus, employing the search terms ("*Langerhans Cell Histiocytosis*" *OR* "*LCH*") *AND* ("*spinal*" *OR* "*vertebral*" *OR* "*extramedullary*"). The initial search yielded 197 articles from PubMed and 315 articles from Scopus, totaling 512 studies. After removing duplicates and resolving discrepancies, 340 studies remained for screening. Eligible studies were clinical studies (case reports, case series, retrospective or prospective studies) that presented full details of patient presentation, diagnosis, treatment, and follow-up. Only English language articles were included, and there were no date restrictions. Nonclinical studies, abstracts, reviews, animal studies, and studies with no detailed management information were excluded. Following an initial title and abstract screening, 81 studies were selected for full-text review. The final selection was made after applying quality assessment tools, including the CARE guidelines for case reports and the ROBINS-I tool for observational studies [Tables 1 and 2].[17,49] Key variables, including patient demographics, lesion location, clinical presentation, treatment approach, neurological outcomes, recurrence, and follow-up duration, were systematically extracted and analyzed.

A qualitative synthesis was performed to compare different treatment modalities and their effectiveness in spinal LCH management.

RESULTS

A total of 46 studies were included [Table 3].[1-8,10-12,15,16,19-23,26-28,30-35,37-43,45-48,50-53,55-57] The patient population ranged from

Table 1: Quality assessment of case reports using Case Report (CARE) guidelines.

Author, year	Patient information	Clinical findings	Diagnostic assessment	Therapeutic interventions	Follow-up outcomes	Discussion/ conclusions	Overall quality
Tortori-Donati *et al*., 1996[51]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Relevant	High
Geusens *et al*., 1998 [22]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Relevant	High
Garg *et al*., 2003[19]	Comprehensive	Well-described	Detailed	Well-documented	Documented	Well-analyzed	High
Karagoz Guzey *et al*., 2003[28]	Comprehensive	Well-described	Extensive	Well-documented	Documented	Relevant	High
Metellus *et al*., 2007[35]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Relevant	High
Vadivelu *et al*., 2007[53]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Relevant	High
Aw *et al*., 2008[3]	Comprehensive	Moderate	Thorough	Well-documented	Documented	Well-analyzed	High
Sayhan *et al*., 2019[46]	Moderate	Moderate	Moderate	Not Well-documented	Documented	Well-analyzed	Moderate
Zhong *et al*., 2010[57]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Relevant	High
Ariff *et al*., 2011[2]	Comprehensive	Detailed	Extensive	Well-documented	Documented	Extensive	High
Ha *et al*., 2012[23]	Detailed	Moderate	Moderate	Not Well-documented	Documented	Moderate	Moderate
Paxinos *et al*., 2011[42]	Detailed	Moderate	Moderate	Not Well-documented	Documented	Moderate	Moderate
Tyagi *et al*., 2011[52]	Comprehensive	Well-described	Extensive	Well-documented	Documented	Extensive	High
Feng *et al*., 2013[15]	Detailed	Moderate	Moderate	Not Well-documented	Documented	Moderate	Moderate
Montemurro *et al*., 2013[37]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Well-analyzed	High
Wang *et al*., 2015[55]	Comprehensive	Well-described	Thorough	Well-documented	Documented	Relevant	High
Özdemir *et al*., 2016[41]	Comprehensive	Well-described	Thorough	Well-documented	Documented	Relevant	High
Sadashiva *et al*., 2016[45]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Well-analyzed	High
Balachandran *et al*., 2017[4]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Relevant	High
Chua *et al*., 2017[11]	Comprehensive	Well-described	Moderate	Well-documented	Documented	Well-analyzed	High
Lan *et al*., 2018[30]	Comprehensive	Well-described	Extensive	Well-documented	Documented	Extensive	High

(*Contd*...)

Table 1: (*Continued*).

Author, year	Patient information	Clinical findings	Diagnostic assessment	Therapeutic interventions	Follow-up outcomes	Discussion/ conclusions	Overall quality
Nakashima *et al.*, 2018[39]	Comprehensive	Well-described	Extensive	Well-documented	Documented	Extensive	High
Schär *et al.*, 2019 [47]	Comprehensive	Well-described	Extensive	Well-documented	Documented	Extensive	High
Burkes and Anderson, 2019[6]	Comprehensive	Well-described	Extensive	Well-documented	Documented	Extensive	High
Chan *et al.*, 2019[8]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Well-analyzed	High
Lim and Cho, 2020[33]	Detailed	Moderate	Moderate	Not Well-documented	Documented	Moderate	Moderate
Singh *et al.*, 2019[48]	Detailed	Moderate	Moderate	Not Well-documented	Documented	Moderate	Moderate
Champaneri and Banerjee, 2020[7]	Comprehensive	Well-described	Moderate	Well-documented	Documented	Well-analyzed	High
Erdogan *et al.*, 2021[14]	Comprehensive	Detailed	Thorough	Well-documented	Documented	Well-analyzed	High
Foti *et al.*, 2023[16]	Detailed	Moderate	Moderate	Not Well-documented	Documented	Moderate	Moderate
Al-Salihi *et al.*, 2023[1]	Comprehensive	Detailed	Extensive	Well-documented	Documented	Extensive	High
Chaulagain *et al.*, 2023[10]	Detailed	Moderate	Moderate	Well-documented	Documented	Moderate	Moderate
Dayyani *et al.*, 2023[12]	Comprehensive	Moderate	Limited	Not Well-documented	Not Documented	Limited	Limited

Table 2: ROBINS-I assessment of the included studies.

Author, Year	Confounding	Selection of Patients	Classification of Interventions	Deviations from Intended Interventions	Missing Data	Measurement of Outcomes	Selection of Reported Results
Garg *et al.* (2004)[20]	Moderate	High	High	Moderate	Low	High	High
Tan *et al.* (2004)[50]	Moderate	Moderate	Moderate	Moderate	Moderate	High	Moderate
Brown *et al.*, 2005[5]	Moderate	Moderate	Low	Moderate	Low	Moderate	Moderate
Puigdevall *et al.* (2008)[43]	Moderate	Moderate	Low	Moderate	Low	Moderate	Moderate
Jiang *et al.* (2010)[27]	Moderate	Moderate	Low	Moderate	Low	Moderate	Moderate
Jiang *et al.* (2011)[26]	Moderate	Moderate	Low	Moderate	Low	Moderate	Moderate
Lee *et al.* (2014)[31]	Moderate	Moderate	Low	Moderate	Low	Moderate	Moderate
Lü *et al.*, 2014[34]	Moderate	Moderate	Low	Moderate	Low	Moderate	Moderate
Lee *et al.* (2017)[32]	Moderate	Moderate	Low	Moderate	Low	Moderate	Moderate
Xu *et al.*, 2018 [56]	Moderate	Moderate	Moderate	High	Moderate	Moderate	Moderate
Nakamura *et al.*, 2019[38]	Moderate	Low	Moderate	Moderate	Low	Low	Low
Gatineau-Sailliant *et al.*, 2020[21]	Low	Moderate	Low	Moderate	Low	Moderate	Low
Otsuki *et al.*, 2024[40]	Low	Low	Moderate	Moderate	Low	Moderate	Low

infancy to adulthood, with the youngest reported case being 11 months old and the oldest 71 years old. The majority of the cases were pediatric. Males were more frequently affected. The geographical distribution of cases included reports from China, the USA, Canada, South Korea, and several European nations. The majority of studies were case reports, with few retrospective series offering more comprehensive data on treatment outcomes. Patients presented with a broad range of symptoms, predominantly local pain, and restricted spinal mobility. Neurological deficits were also present, ranging from mild radiculopathy to quadriparesis. The other prominent symptoms included torticollis, abnormal gait, and polyuria-polydipsia syndrome in one patient with systemic involvement. The duration of symptoms before diagnosis was also extremely variable, ranging from acute over days to chronic over several years. Osteolytic vertebral lesions with variable degrees of vertebral plana and soft tissue extension characterized spinal involvement. Xu *et al*. (2018)[56] reported that multifocal disease was present in 11.8% of cases, with the cervical spine being the most commonly affected region (63.7%). Other affected regions included the thoracic (21.9%) and lumbar spine (13%), while sacral involvement was rare (1.4%). The majority of the patients presented with single vertebral involvement. Vertebral body collapse was a typical feature, as seen in the Xu *et al*.[56] study and Nakamura *et al*.[38] study, with extensive cases resulting in kyphotic deformities. MRI findings consistently showed hyperintense lesions on T2-weighted imaging with soft tissue extension in a small number of cases.

Management strategies varied depending on disease severity, neurological involvement, and lesion stability [Table 4].[1-8,10-12,15,16,19-23,26-28,30-35,37-43,45-48,50-53,55-57] Many cases were successfully managed conservatively with immobilization, corticosteroids, or chemotherapy, particularly vinblastine-based regimens, which led to complete vertebral reconstitution in pediatric cases. Long-term follow-up showed minimal recurrence in nonsurgical cases. Surgical intervention was primarily reserved for cases with instability or neurological deficits. Surgical procedures such as posterior instrumentation and corpectomy were performed in instances of extreme collapse of the vertebrae or spinal cord compression, with often excellent neurological recovery. Tumor resection and stabilization were employed in a couple of instances with very large vertebral destruction. Adjuvant therapy in the form of radiotherapy for residual disease and LCH-II/LCH-III chemotherapy protocols were beneficial in instances with progressive or multifocal disease. Most patients, especially those treated conservatively, exhibited full neurological recovery. Spinal deformities requiring stabilization occurred in some cases, but recurrence rates were low. No perioperative mortality was reported. Follow-up ranged from months to over a decade, with spontaneous vertebral reconstitution frequently observed in pediatric cases. Patients undergoing surgery for unstable lesions showed sustained neurological improvement with minimal risk of recurrence.

DISCUSSION

Intradural Extramedullary eosinophilic granuloma of the spine is an extremely rare manifestation of LCH, with very few cases reported in the literature. LCH is a clonal proliferative disorder of Langerhans cells, which are specialized dendritic cells involved in antigen presentation. The disease can present in a variety of forms, ranging from isolated bone lesions to multisystem involvement, and can affect both pediatric and adult populations.[16,29,48]

The diagnosis of spinal LCH can be challenging due to its nonspecific clinical and radiological features. Patients may present with symptoms of spinal cord compression, such as pain, paresthesia, and motor deficits, which can rapidly progress if not promptly addressed. MRI is the imaging modality of choice for evaluating spinal lesions, but the definitive diagnosis often requires histopathological confirmation. The characteristic histological features of LCH include the presence of Langerhans cells with grooved, coffee bean-shaped nuclei and positive immunohistochemical staining for CD1a and S-100 protein.[18,25,54]

In the present case, the initial suspicion was tuberculoma, a common differential diagnosis in tuberculosis-endemic areas. The ring-enhancing pattern on MRI was more toward an infectious etiology. However, the acute onset of neurological symptoms required surgical intervention and histopathological analysis, which ultimately diagnosed LCH. This emphasizes the importance of considering LCH in the differential diagnosis of spinal lesions, even in endemic areas for other diseases.[9,13] Our review of spinal LCH cases demonstrates that its radiological features can be highly variable, often mimicking infectious or neoplastic conditions. Xu *et al*. (2017) [56] reported vertebral plana, osteolytic lesions, and paravertebral soft tissue extension as common findings, while Jiang *et al*.(2010)[27] reported paravertebral involvement in up to 40% of cases. This was consistent with the MRI findings in our case, in which the lesion extended from the extradural space to the posterior bony elements and paraspinal muscles. The ring-like enhancement, though suggestive of infection, has been reported in LCH as well, emphasizing the need for histopathological confirmation.

Treatment of spinal LCH typically involves surgical resection for decompression of the spinal cord and symptomatic relief. In cases where complete resection is not possible, adjuvant therapy in the form of radiotherapy or chemotherapy may be employed. The prognosis of spinal LCH varies depending on the extent of the disease and the success of the initial treatment. Early diagnosis and intervention are crucial for

Table 3: Study characteristics and patient data.

Author (Year)	Study type	N	Country	Age	Gender	Symptoms
Tortori-Donati *et al.* (1996)[51]	Case Report	1	Italy	2 years	Female	Progressive paraparesis, falls, inability to climb stairs
Geusens *et al.* (1998)[22]	Case Report	1	Belgium	11 years	Male	Neck pain, decreased cervical mobility, radiation to left shoulder
Garg *et al.* (2003)[19]	Case Report	1	USA	6 years	Female	Neck pain worsened with the extension
Karagoz Guzey *et al.* (2003)[28]	Case Report	1	Turkey	7 years	Male	Occipital swelling, neck pain
Garg *et al.* (2004)[20]	Case Series	26	USA	8.2 years (0.2–16.4)	15 M, 11 F	Back/neck pain (100%), torticollis (4 cases), abnormal gait (3 cases)
Tan *et al.* (2004)[50]	Case Series	4	Belgium	3–13 years	4 M	Neck pain, torticollis, stiffness, radicular symptoms
Brown *et al.* (2005)[5]	Retrospective Study	8	Canada	3–11 years (Mean: 6.4)	3 M, 5 F	Back/neck pain, abnormal gait, torticollis
Metellus *et al.* (2007)[35]	Case Report	1	France	17 years	Male	Cervicothoracic pain, right-hand weakness, paresthesia in both arms
Vadivelu *et al.* (2007)[53]	Case Report	1	USA	11 months	Male	Lower extremity weakness
Aw *et al.* (2008)[3]	Case Report	1	UK	17 years	Male	Neck pain, stiffness, limited rotation
Puigdevall *et al.* (2008)[43]	Case Series	2	Argentina	10 years (C1), 5 years (C2)	Male (C1), Female (C2)	Neck pain (C1), Neck pain+ torticollis (C2)
Jiang *et al.* (2010)[27]	Case Series	30	China	Mean: 14.2 years (Range: 1.5–41)	20M, 10F	Neck pain (96.7%), restricted motion (70%), neurological symptoms (36.7%)
Sayhan *et al.* (2019)[46]	Case Report	1	Turkey	47 years	Female	Intractable cervical pain, paresthesia in the left arm
Zhong *et al.* (2010)[57]	Case Report	1	China	26 years	Male	Neck pain, stiffness, difficulty holding the head upright
Ariff *et al.* (2011)[2]	Case Report	1	Malaysia	37 years	Male	Progressive back pain, quadriparesis, weight loss, low-grade fever
Ha *et al.* (2012)[23]	Case Report	1	South Korea	6 years	Male	Neck pain, torticollis

(*Contd...*)

Table 3: (*Continued*).

Author (Year)	Study type	N	Country	Age	Gender	Symptoms
Jiang *et al.* (2011)[26]	Case Series	5	China	12–23 years	4M, 1F	Back/neck pain (100%), neurological symptoms (60%)
Paxinos *et al.* (2011)[42]	Case Report	1	Greece	35 years	Male	Progressive nocturnal upper lumbar pain
Tyagi *et al.* (2011)[52]	Case Report	1	India	35 years	Female	Progressive lower limb weakness, tingling, numbness, urinary retention
Feng *et al.* (2013)[15]	Case Report	1	China	51 years	Male	Low back pain, limited waist motion, right leg numbness
Lee *et al.* (2014)[31]	Retrospective Study	6	South Korea	7–38 years	4 M, 2 F	Painful palpable mass (skull/spine), motion restriction, neurological deficits in some cases
Montemurro *et al.* (2013)[37]	Case Report	1	Italy	71 years	Male	Progressive lower limb weakness, weight loss
Lü *et al.* (2014)[34]	Retrospective Study	12	China	2–16 years (Mean: 9.58)	8 M, 4 F	Back pain, progressive neurological deficit
Wang *et al.* (2015)[55]	Case Report	1	China	14 years	Female	Occipitocervical pain
Özdemir *et al.* (2016)[41]	Case Report	1	Turkey	35 years	Female	Back and left buttock pain, malaise, weight loss
Sadashiva *et al.* (2016)[45]	Case Report	1	India	9 years	Female	Neck pain, neck tilt, left upper limb weakness, intermittent fever
Balachandran *et al.* (2017)[4]	Case Report	1	India	6 years	Male	Lower back pain, worse at night, limiting activity
Chua *et al.* (2017)[11]	Case Report	1	Singapore	6 years	Female	Left-sided neck pain, torticollis, cervical lymphadenopathy
Lee *et al.* (2017)[32]	Retrospective Study	22	South Korea	0.6–12.3 years (Median: 4.1)	15 M, 7 F	Neck/back pain (55%), no neurological deficits
Xu *et al.* (2018)[56]	Retrospective Study	110	China	1–52 years (Mean: 12.8)	69 M, 41 F	Pain (93.6%), restricted motion (47.3%), neurological symptoms (18.2%)
Lan *et al.* (2018)[30]	Case Report	1	China	1 year 4 mo	Female	Recurrent fever, anemia, irritability, inability to sit without support

(*Contd...*)

Table 3: (*Continued*).

Author (Year)	Study type	N	Country	Age	Gender	Symptoms
Nakashima *et al.* (2018)[39]	Case Report	1	Japan	21 months	Male	Sudden-onset paraplegia, rigid trunk posture
Schär *et al.* (2019)[47]	Case Report	1	Canada	36 years	Male	Right-sided neck and shoulder pain, stiffness
Burkes and Anderson, (2019)[6]	Case Report	1	USA	7 years	Female	Thoracic back pain, worsening with activity
Chan *et al.* (2019)[8]	Case Report	1	UK	7 years	Male	Thoracic back pain, leg weakness
Lim and Cho, (2020)[33]	Case Report	1	South Korea	33 years	Male	Severe back pain, gait disturbance, bilateral leg weakness
Nakamura *et al.* (2019)[38]	Retrospective Study	13	Japan	1.4–7.8 years (Mean: 3.6)	9 F, 4 M	Back pain, kyphotic deformity
Singh *et al.* (2019)[48]	Case Report	1	India	15 years	Female	Progressive spastic quadriparesis
Champaneri and Banerjee, (2020)[7]	Case Report	1	Nepal	14 years	Male	Low back pain worsens at night
Gatineau-Sailliant *et al.* (2020)[21]	Retrospective Study	11	Canada	0.6–17.3 years (Median: 8.25)	6M, 5F	Back pain, stiffness, walking pain
Erdogan *et al.* (2021)[14]	Case Report	1	Turkey	18 months	Male	Progressive lower limb weakness, gait disturbance
Foti *et al.* (2023)[16]	Case Report	1	Italy	50 years	Male	Chronic dorsolateral chest pain
Al-Salihi *et al.* (2023)[1]	Case Report	1	Qatar	46 years	Female	Back pain, numbness, pyramidal paraparesis, urine retention, constipation
Chaulagain *et al.* (2023)[10]	Case Report	1	Ukraine	22 years	Male	Nape pain, upper limb radiculopathy
Dayyani *et al.* (2023)[12]	Case Report	1	Iran	4 years	Male	Progressive lower limb weakness, large head lump, fever, weight loss
Otsuki *et al.* (2024)[40]	Retrospective Study	4	Japan	21–28 years	Male	Severe back pain, anterior chest pain, one case with lower limb numbness

Author (Year)	Duration of Symptoms	Examination Findings	Imaging Findings	Local versus Multiple	Lesion Location
Tortori-Donati *et al.* (1996)[51]	8 months	Weakness in lower limbs, hyperreflexia	Lumbosacral intradural-extramedullary mass, hyperintense on T1, isointense on T2	Multiple	Lumbosacral

(*Contd...*)

Table 3: (*Continued*).

Author (Year)	Duration of Symptoms	Examination Findings	Imaging Findings	Local versus Multiple	Lesion Location
Geusens *et al*. (1998)[22]	2 months	Torticollis, tender swelling, cervical lymphadenopathy	A lytic lesion in C5 lateral mass, soft tissue component, peripheral enhancement on CT	Solitary	Cervical (C4-C5)
Garg *et al*. (2003)[19]	1 month	Tenderness in upper cervical spine, no neurological deficits	Posterior elements of C3 destroyed, soft tissue mass, mild C3–C4 pseudosubluxation	Multiple	Cervical (C3), Lumbar (L4 subclinical)
Karagoz Guzey *et al*. (2003)[28]	4 months	Normal except for occipital swelling	Occipital calvarial lesion, C6 and T11 vertebra plana	Multiple	Occipital, C6, T11
Garg *et al*. (2004)[20]	Variable	Mostly normal; 3 cases had pain radiating to the upper extremity	44 vertebrae affected: 20 cervical, 14 thoracic, 10 lumbar, vertebral body collapse in varying grades	Multiple	Cervical (20), Thoracic (14), Lumbar (10)
Tan *et al*. (2004)[50]	4–6 weeks	Restricted neck movement, 1 case with right arm paresis	Osteolytic lesions in C3–C5, soft tissue extension in 2 cases	Local	Cervical (C3–C5)
Brown *et al*. (2005)[5]	Variable	One case with an abnormal gait, one case with polyuria and polydipsia	Thoracic vertebrae were most affected, followed by cervical and lumbar; vertebra plana in 4 cases	Multiple	Cervical (C3), Thoracic (T5-T11), Lumbar (L1, L4)
Metellus *et al*. (2007)[35]	3 weeks	Right-hand distal motor deficit, pyramidal signs in lower limbs	T1 vertebra plana, epidural soft tissue mass, mass effect on spinal cord, C7-T2 involvement	Local	Cervicothoracic (C7-T2)
Vadivelu *et al*. (2007)[53]	3 months	Weakness in both legs, no fever, normal labs	T3 vertebra plana with enhancing epidural mass extending T1–T5, L2 lytic lesion	Multiple	Thoracic (T3), Lumbar (L2)
Aw *et al*. (2008)[3]	2 weeks	Tenderness at C6/7, mild rotational deficit, no neurological deficits	C4 vertebra plana with osteolytic lesion, high T2 signal, para-vertebral edema, syrinx in the lower cervical spine	Local	Cervical (C4)
Puigdevall *et al*. (2008)[43]	2 months (C1), 3 weeks (C2)	Slight restriction of neck movement (C1), Pain with cervical motion (C2)	C1 lateral mass lytic lesion (C1), T4 vertebra plana, L1 lytic lesion (C2)	Multiple (C2)	Cervical (C1), Thoracic (T4), Lumbar (L1)

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Table 3: (*Continued*).

Author (Year)	Duration of Symptoms	Examination Findings	Imaging Findings	Local versus Multiple	Lesion Location
Jiang *et al*. (2010)[27]	Variable	3 cases had fixed atlantoaxial dislocation 3, had spontaneous fusion	14 atlantoaxial, 16 subaxial cases; 40% had a paravertebral extension, 30% epidural, 56.3% pedicle/transverse process involvement	Multiple (4/30)	Cervical (C1-C7)
Sayhan *et al*. (2019)[46]	N/A	Minimal cervical kyphosis reduced left wrist flexion	C5 vertebra plana, osteolytic lesion with severe kyphosis, no disc involvement	Local	Cervical (C5)
Zhong *et al*. (2010)[57]	2 months	No neurological deficit, restricted neck motion	An osteolytic lesion in the left lateral mass of the atlas (C1), compression fracture	Local	Cervical (C1, Atlas)
Ariff *et al*. (2011)[2]	5 months	Tenderness over T6-T7, reduced sensation in T7-T8 dermatomes	Multiple osteolytic lesions in T4-T6, L3-L4, S1, vertebral collapse at L4, thyroid mass	Multiple	Thoracic (T4-T6), Lumbar (L3-L4, S1), Thyroid
Ha *et al*. (2012)[23]	3 weeks	Severe tenderness in the upper cervical spine, no neurological deficit	Osteolytic lesion of the odontoid process and C2 body, posterior displacement	Multiple (C2 & femur)	Cervical (C2, odontoid process), Femur
Jiang *et al*. (2011)[26]	2 weeks–6 months	3 cases had neurological deficits, 4 had soft tissue involvement	19 affected vertebrae: 5 cervical, 12 thoracic, 2 lumbar-sacral; 3 cases with vertebra plana, 3 with epidural extension	Multiple	Cervical (C1-C7), Thoracic (T1-T12), Lumbar (L1-L5)
Paxinos *et al*. (2011[42]	5 months	Right postural scoliosis, localized tenderness over L1	A lytic lesion in L1 vertebral body, no soft tissue mass, minimal sclerosis	Local	Lumbar (L1)
Tyagi *et al*. (2011)[52]	2 months	Paraplegia, hyperreflexia, absent sensations below D2	Well-defined extradural contrast-enhancing mass at D2-D4, cord compression	Local	Thoracic (D2-D4)
Feng *et al*. (2013)[15]	10 days	Tender L4 spinous process, restricted waist motion	L4 osteolytic lesion, mild compression fracture, paravertebral and intraspinal soft tissue mass	Local	Lumbar (L4)

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Table 3: (*Continued*).

Author (Year)	Duration of Symptoms	Examination Findings	Imaging Findings	Local versus Multiple	Lesion Location
Lee *et al*. (2014)[31]	Variable	1 case had scapular involvement	Skull & spine lesions, some with epidural mass	Local	Cervical (2), Thoracic (2), Lumbar (2)
Montemurro *et al*. (2013)[37]	2 years	Muscle strength 3–4/5 in lower limbs, hyperreflexia, myelopathic signs	Homogeneous enhancing epidural lesion at T6-T8, spinal cord compression, lytic lesion at T2 spinous process	Multiple	Thoracic (T6-T8, epidural), T2 spinous process
Lü *et al*. (2014)[34]	1–2 months	Frankel B (2), Frankel C (8), Frankel D (2)	Vertebra plana, space-occupying mass, spinal canal encroachment (>50%)	Multiple	Thoracic (T3-T12), Lumbar (L1-L3)
Wang *et al*. (2015)[55]	1 month	Tenderness in occipitocervical region, no limb weakness	C2, T1, T5, T12, and L4 vertebral destruction without spinal cord compression, paravertebral extension at C2	Multiple	Cervical (C2), Thoracic (T1, T5, T12), Lumbar (L4)
Özdemir *et al*. (2016)[41]	Several months	Tender left iliac wing, spinous process pain in thoracic/lumbar spine	Unilateral destructive sacroiliac lesion, multiple vertebral lesions, extensive soft tissue extension	Multiple	Sacroiliac joint, Thoracic (T9-T11), Lumbar (L4-L5)
Sadashiva *et al*. (2016)[45]	Several days	Left upper limb proximal weakness (Grade 4/5), loss of cervical lordosis	C5 vertebral body collapse (>90%), epidural and prevertebral soft tissue collection, cord compression	Local	Cervical (C5)
Balachandran *et al*. (2017)[5]	10 days	Tenderness over lumbar spine, antalgic gait, mild scoliosis	L4 vertebral collapse, right pedicle and lamina involvement, mild soft tissue extension	Local	Lumbar (L4)
Chua *et al*. (2017)[11]	1 month	No neurological deficits	A lytic lesion in clivus, occipital condyle, atlas, and axis (C1-C2), causing atlantoaxial subluxation	Local	Cranio-cervical (Clivus, C1-C2)
Lee *et al*. (2017)[32]	Variable	No significant neurologic or orthopedic deficits	31 lesions: Cervical (4), Thoracic (17), Lumbar (10); 8 lesions detected only through MRI	Multiple	Cervical (4), Thoracic (17), Lumbar (10)

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Table 3: (*Continued*).

Author (Year)	Duration of Symptoms	Examination Findings	Imaging Findings	Local versus Multiple	Lesion Location
Xu *et al*. (2018)[56]	Variable	Radiculopathy (18 cases), myelopathy (2 cases)	146 lesions: Cervical (63.7%), Thoracic (21.9%), Lumbar (13.0%), Sacral (1.4%); vertebra plana in 29.3% of children	88.2% single, 11.8% multiple	Cervical (63.7%), Thoracic (21.9%), Lumbar (13%), Sacral (1.4%)
Lan *et al*. (2018)[30]	1 month	Brisk lower limb reflexes reduced rectal sphincter tone	T12 vertebral collapse (>95%), large intradural soft tissue mass compressing cord (T10-L1)	Local	Thoracic (T12)
Nakashima *et al*. (2018)[39]	2 weeks	Complete lower limb paralysis, hyperreflexia	Osteolytic lesions in T6-T8, epidural mass at T7-T9 compressing spinal cord	Local	Thoracic (T6-T8)
Schär *et al*. (2019)[47]	9 months	Tenderness over the cervical spine, restricted neck motion	Expansive osteolytic lesion in right C4 vertebral body, infiltration of right pedicle and cranial C5	Local	Cervical (C4)
Burkes and Anderson, (2019)[6]	3 weeks	Dysesthesias and lower limb weakness	T7 vertebra plana with progressive kyphosis, new soft tissue mass compressing spinal cord	Local	Thoracic (T7)
Chan *et al*. (2019)[8]	3 weeks	Mild left leg weakness	T1 vertebral collapse with posterior soft tissue extension compressing spinal cord, additional right acetabular lesion	Multiple	Thoracic (T1), Acetabulum
Lim and Cho, (2020)[33]	1 year (pain), 7 days (neurological symptoms including gait disturbance, and weakness in both lower extremities)	Hyperreflexia decreased hip flexion strength	Enhancing epidural lesion with osteolytic destruction at T7–L1, spinal cord compression at T9–T12	Local	Thoracic (T7–L1)

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Table 3: (*Continued*).

Author (Year)	Duration of Symptoms	Examination Findings	Imaging Findings	Local versus Multiple	Lesion Location
Nakamura *et al*. (2019)[38]	Variable	Local kyphosis in early stages	16 affected vertebrae: Cervical (C4, C5, C7), Thoracic (T5, T8, T9, T12), Lumbar (L1-L5); severe collapse at 1 year, reconstitution after 2 years	5 Local 8 Multiple	Cervical, Thoracic, Lumbar
Singh *et al*. (2019)[48]	2 months	Increased muscle tone, 0–1/5 power in all limbs	3×3.5 cm intradural extramedullary dural-based mass at C4-C5	Local	Cervical (C4-C5)
Champaneri and Banerjee, (2020)[7]	1 month	No neurological deficits	Enhancing expansile osteolytic lesion at S1 with cortical breach and soft tissue impingement on thecal sac	Local	Sacral (S1)
Gatineau-Sailliant *et al*. (2020)[21]	Variable	Spine stiffness, scoliosis (1 case), kyphosis (1 case)	29 vertebral lesions (Thoracic: 15, Lumbar: 8, Cervical: 6), 2 unstable lesions, vertebra plana in 3 cases	Multiple	Cervical (6), Thoracic (15), Lumbar (8)
Erdogan *et al*. (2021)[14]	N/A	Paraparesis increases deep tendon reflexes	Total T3 vertebral collapse with anterior soft tissue compression of spinal cord	Local	Thoracic (T3)
Foti *et al*. (2023)[16]	N/A	Normal neurological exam, limited waist motion	Widening of left T10 costovertebral junction with sclerosis, no osteolysis	Local	Thoracic (T10, costovertebral joint)
Al-Salihi *et al*. (2023)[1]	1 week	Sensory level at T7, brisk reflexes, Babinski sign Thoracolumber tenderness.	T6 vertebral collapse with epidural soft tissue mass compressing spinal cord, additional systemic pituitary and renal involvement	Local	Thoracic (T6), Pituitary, Renal
Chaulagain *et al*. (2023)[10]	3 months	No neurological deficits, normal reflexes	C3 vertebral osteolytic lesion, anterior epidural mass causing mild spinal stenosis	Local	Cervical (C3)

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Table 3: (*Continued*).

Author (Year)	Duration of Symptoms	Examination Findings	Imaging Findings	Local versus Multiple	Lesion Location
Dayyani *et al.* (2023)[12]	1 year (weakness), 3 months (paralysis)	Severe muscle wasting, increased deep tendon reflexes, scalp swelling, ulcerative skin lesions	T5-T8 vertebral lytic lesions, T7 collapse with spinal cord compression, multiple enhanced brain lesions, abnormal meningeal enhancement	Multiple	Thoracic (T5-T8), Skull, Brain
Otsuki *et al.* (2024)[40]	6–20 weeks	One case had a mild sensory deficit, others normal	T5-T7 and C5 osteolytic lesions with pathological fractures	Local	Thoracic (T5-T7), Cervical (C5)

MRI: Magnetic resonance imaging, CT: Computed tomography

Table 4: Treatment outcomes and complications.

Author (Year)	Treatment Approach	Surgical Resection (Y/N)	Extent of resection	Adjuvant therapy	Neurological Recovery
Tortori-Donati *et al.* (1996)[51]	Partial tumor excision+ chemotherapy	Y	Partial excision	Etoposide+ Prednisone	Partial improvement, persistent lower limb weakness
Geusens *et al.* (1998)[22]	Corticosteroids (no surgery)	N	N/A	Oral corticosteroids	Rapid improvement in pain and mobility
Garg *et al.* (2003)[19]	Conservative (cervical bracing, biopsy only)	Y (biopsy only)	N/A	Methotrexate+ Prednisone (3 months)	Complete resolution, full spinal reconstitution
Karagoz Guzey *et al.* (2003)[28]	Surgery+ Chemotherapy	Y	C6 corpectomy+ fusion; posterior stabilization (T11)	Vinblastine (24 weeks)	Full recovery
Garg *et al.* (2004)[20]	Conservative (Observation, Bracing, Chemo)	N	N/A	Methotrexate+ Prednisone (10 cases), Radiation (4 cases)	Full recovery
Tan *et al.* (2004)[50]	Cervical brace, Biopsy+ Corticosteroids	N	N/A	Oral Prednisolone (1 mg/kg/day for 5 weeks, tapered over 16 weeks)	Complete recovery in all 4 cases
Brown *et al.* (2005)[5]	Conservative (Bracing, Biopsy, Observation)	N	N/A	Chemotherapy (Vinblastine) in 2 cases, Steroids in 2 cases, Radiotherapy (6 Gy) in 1 case	Full resolution in 6 cases, 1 recurrence
Metellus *et al.* (2007)[35]	Surgery+ Spinal Stabilization	Y	T1 corpectomy, C7-T2 extended resection, C6-T3 fixation	None	Full recovery, complete spinal stabilization
Vadivelu *et al.* (2007)[53]	Laminectomy+ Biopsy+ Chemotherapy	Y	T1-T5 laminectomy, decompression	Vinblastine+ Prednisone (6 months)	Partial recovery at 4 months, full recovery at 15 months

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Table 4: (*Continued*).

Author (Year)	Treatment Approach	Surgical Resection (Y/N)	Extent of resection	Adjuvant therapy	Neurological Recovery
Aw *et al*. (2008)[3]	Surgical management consisted of an autologous bone transplantation: a C4 corporectomy iliac bone graft and Condam Onlay plate for spinal stabilization.	Y	C4 corpectomy+ iliac bone graft+ Condam plate	None	Full recovery
Puigdevall *et al*. (2008)[43]	Nonoperative (Bracing)	N	N/A	None	Full recovery
Jiang *et al*. (2010)[27]	Eighteen patients had conservative treatment, and 12 underwent operation., plus (Immobilization, Radiotherapy, and Chemotherapy)	Y	N/A	Radiotherapy (13 cases), Chemotherapy (4 cases)	Complete resolution in all cases
Sayhan *et al*. (2019)[46]	Tumor decompression, stabilization with a mesh cage and anterior plates between C4 and C6.	Y	C5 Corpectomy+ Anterior Plate Fixation (C4–C6)	Postoperative Chemotherapy	Full recovery, no neurological deficit
Zhong *et al*. (2010)[57]	Conservative (Halo-Vest Immobilization+ Radiotherapy)	N	N/A	Radiotherapy (3,000 cGy)	Full recovery with mild neck stiffness
Ariff *et al*. (2011)[2]	First surgery: Posterior decompression procedure with curettage and posterior instrumentation and bone biopsy of the T6 and T7 vertebrae. Second surgery: The patient underwent emergency open reduction, decompression, curettage, and interbody fusion of C2-C4 with anterior bone grafting and posterior instrumentation	Y	T6-T7 curettage	None	18 months following the surgery, the patient presented with quadriparesis and inability to micturate, with reduced sensation from level C6 downward. Full recovery postsecond surgery
Ha *et al*. (2012)[23]	Conservative (Brace Immobilization+ Chemotherapy)	N	N/A	Vinblastine (11 months)	Full recovery with complete bone remodeling

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Table 4: (*Continued*).

Author (Year)	Treatment Approach	Surgical Resection (Y/N)	Extent of resection	Adjuvant therapy	Neurological Recovery
Jiang *et al*. (2011)[26]	Conservative (Immobilization, Chemotherapy, Radiotherapy). One patient underwent curettage and reconstruction without preoperative biopsy.	N	N/A	Chemotherapy (Cyclophosphamide, Doxorubicin, Vincristine, Prednisone), Radiotherapy (1,980 cGy)	Complete resolution in all cases
Paxinos *et al*. (2011)[42]	Surgery+ Spinal Fusion	Y	L1 curettage+ T12-L2 posterolateral fusion	None	Full recovery
Tyagi *et al*. (2011)[52]	D2-D5 decompressive laminectomy	Y	D2-D5 laminectomy+ total tumor excision	Chemotherapy	Full recovery
Feng *et al*. (2013)[15]	Percutaneous Vertebroplasty+ Chemotherapy	N	N/A	Etoposide+ Prednisone (3 cycles)	Full recovery, complete pain relief
Lee *et al*. (2014)[31]	Surgery (Total excision in 15, Biopsy in 2)	Y	Skull/spine excision	Chemotherapy (Vinblastine, Prednisone) in 5 cases	Full recovery
Montemurro *et al*. (2013)[37]	D6–D8 laminectomy, and the tumor was removed in a piecemeal fashion.	Y	T6-T8 laminectomy+ epidural tumor excision	2-chlorodeoxyadenosine (2-CdA, 3 cycles)	Full neurological recovery
Lü *et al*. (2014)[34]	Surgery (Posterior+ Anterior fixation)	Y	Pedicle screw fixation, corpectomy, decompression	None	Full recovery in all cases
Wang *et al*. (2015)[55]	First, the patient was given occipital jaw traction of 2 kg and limited activity. Surgery (Curettage+ Bone graft)	Y	C2 tumor curettage, iliac bone graft	None	Partial pain relief, no neurological deficit but Occipitocervical pain relieved completely at a 3-month follow-up.
Özdemir *et al*. (2016)[41]	Surgery (Biopsy+ Curettage)	Y	Sacroiliac joint+ multiple vertebrae	None	Full recovery
Sadashiva *et al*. (2016)[45]	Surgery+ Radiotherapy	Y	C5 corpectomy, spinal fusion	Postoperative Radiotherapy	Full recovery
Balachandran *et al*. (2017)[4]	Chemotherapy+ Bracing	N	N/A	LCH III Protocol	Full recovery
Chua *et al*. (2017)[11]	Conservative (Halo brace+ Chemotherapy)	N	N/A	LCH-III protocol (Vinblastine+ Prednisone), later Cladribine	Full recovery, near-complete vertebral reconstitution

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Table 4: (*Continued*).

Author (Year)	Treatment Approach	Surgical Resection (Y/N)	Extent of resection	Adjuvant therapy	Neurological Recovery
Lee *et al*. (2017)[32]	Conservative (Immobilization, Chemotherapy)	N	N/A	Vinblastine-based chemotherapy (LCH-II, LCH-III)	Full recovery, vertebral height improvement in 70% of cases
Xu *et al*. (2018)[56]	Conservative (Immobilization, Observation, Chemotherapy) 10.7% went under surgery.	N	N/A	Vinblastine-based chemotherapy for multifocal disease	Full recovery in most cases, spontaneous bony reconstitution
Lan *et al*. (2018)[30]	Surgery (Posterior thoracolumbar approach)	Y	T12 corpectomy+ interbody fusion with titanium cage+ iliac crest bone graft	None	Full recovery, no neurological deficit
Nakashima *et al*. (2018)[39]	Radiotherapy+ Chemotherapy	N	N/A	15 Gy radiotherapy+ Vincristine+ Prednisolone+ Cytarabine	Full neurological recovery, no sequelae
Schär *et al*. (2019)[47]	Surgery (C4 corpectomy)+ Radiotherapy	Y	C4 corpectomy, tumor debulking, interbody fusion C3-C5	16 Gy fractionated stereotactic radiotherapy	Full neurological recovery, pain resolution
Burkes and Anderson (2019)[6]	Surgery (T5-T10 fusion+ subtotal T7 vertebrectomy) + Chemotherapy	Y	T7 decompression+ interbody fusion	Vinblastine+ Prednisone (LCH-III protocol)	Full recovery, normal neurological function
Chan *et al*. (2019)[8]	Conservative (NSAIDs+ Immobilization)	N	N/A	Indomethacin+ SOMI brace	Full pain resolution, complete vertebral reconstitution
Lim and Cho, (2020)[33]	Surgery (Posterior laminectomy+ Marginal excision)	Y	T9-T10 laminectomy+ epidural tumor excision	Chemotherapy (Vinblastine+ Prednisolone, 10 months)	Full neurological recovery, no gait disturbance
Nakamura *et al*. (2019)[38]	Conservative (Observation+ Chemotherapy)	N	N/A	Vinblastine-based chemotherapy	Full vertebral reconstitution, spontaneous recovery of vertebral collapse
Singh *et al*. (2019)[48]	Surgery (Laminectomy+ Tumor excision)	Y	C4-C5 laminectomy, total tumor excision	None	Partial recovery, improvement in spasticity
Champaneri and Banerjee, (2020)[7]	Surgery (Laminectomy+ Tumor excision+ Fusion)	Y	S1 vertebral body tumor excision+ fusion	Chemotherapy (Vinblastine+ Prednisolone)	Full recovery, complete disease resolution
Gatineau-Sailliant *et al*. (2020)[21]	Conservative (Chemotherapy)	N	N/A	Vinblastine+ Prednisone	Full recovery in most cases

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Table 4: (*Continued*).

Author (Year)	Treatment Approach	Surgical Resection (Y/N)	Extent of resection	Adjuvant therapy	Neurological Recovery
Erdogan *et al.* (2021)[14]	Surgery (Corpectomy+ Fixation)	Y	Under an operative microscope, T2–3–4 total laminectomy and then left T3-4 costotransversectomy were performed by a bone- cutting device. Total T3 corpectomy, two-level discectomy, and gross total tumor excision were performed through the left posterolateral approach under the microscope.	None	Full neurological recovery
Foti *et al.* (2023)[16]	Conservative (Observation)	N	N/A	None	Full pain relief, spontaneous resolution
Al-Salihi *et al.* (2023)[1]	Surgery (Laminectomy+ Tumor excision+ Fixation)	Y	T6 decompressive laminectomy, screw fixation	Chemotherapy (Vinblastine)	Full recovery
Chaulagain *et al.* (2023)[10]	Surgery (Anterior corpectomy+ Discectomy+ Fixation)	Y	C3 corpectomy, titanium cage, plating	None	Full recovery, pain resolved
Dayyani *et al.* (2023)[12]	Surgery (Laminectomy+ Biopsy)	Y	T7 biopsy+ Histopathological confirmation	None (Patient lost to follow-up)	Unknown (Patient returned to home country)
Otsuki *et al.* (2024)[40]	Surgery (Posterior instrumentation without curettage)	Y	T5, T6, and C5 percutaneous pedicle screw fixation	1 case received chemotherapy	

Author (Year)	Complications	Management of Complications	Recurrence (Y/N)	Follow-up Duration (Months)
Tortori-Donati *et al.* (1996)[51]	None reported	N/A	N	3 months
Geusens *et al.* (1997)[22]	None reported	N/A	N	Not specified
Garg *et al.* (2003)[19]	None reported	N/A	N	108 months (9 years)
Karagoz Guzey *et al.* (2003)[28]	None reported	N/A	N	24 months
Garg *et al.* (2004)[20]	4 cases developed spinal deformity	2 required spinal fusion	N	9.4 years (Mean)
Tan *et al.* (2004)[50]	None reported	N/A	N	41 months (Mean: 3.4 years)
Brown *et al.* (2005)[5]	1 recurrence	Retreated with chemotherapy	Y (1 case)	41 months (Mean: 3.4 years)
Metellus *et al.* (2007)[35]	None reported	N/A	N	24 months
Vadivelu *et al.* (2007)[53]	None reported	N/A	N	15 months

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Table 4: (*Continued*).

Author (Year)	Complications	Management of Complications	Recurrence (Y/N)	Follow-up Duration (Months)
Aw *et al.* (2008)[3]	None reported	N/A	N	Not specified
Puigdevall *et al.* (2008)[43]	None reported	N/A	N	72 months (6 years)
Jiang *et al.* (2010)[27]	None reported	N/A	N	61.6 months (Mean: 5.1 years)
Sayhan *et al.* (2019)[46]	None reported	N/A	N	12 months
Zhong *et al.* (2010)[57]	None reported	N/A	N	84 months (7 years)
Ariff *et al.* (2011)[2]	None reported	N/A	N	24 months
Ha *et al.* (2012)[23]	None reported	N/A	N	24 months
Jiang *et al.* (2011)[26]	None reported	N/A	N	78.6 months (Mean: 6.5 years)
Paxinos *et al.* (2011)[42]	None reported	N/A	N	24 months
Tyagi *et al.* (2011)[52]	None reported	N/A	N	15 months
Feng *et al.* (2013)[15]	None reported	N/A	N	6 months
Lee *et al.* (2014)[31]	None reported	N/A	Y (1 case)	37 months (Mean: 3.1 years)
Montemurro *et al.* (2013)[37]	None reported	N/A	N	16 months
Lü *et al.* (2014)[34]	Pleural effusion (2 cases), Intercostal neuralgia (1 case)	Conservative management	N	43.3 months (Mean: 3.6 years)
Wang *et al.* (2015)[55]	None reported	N/A	N	3 months
Özdemir *et al.* (2016)[41]	None reported	N/A	N	Not specified
Sadashiva *et al.* (2016)[45]	None reported	N/A	N	18 months
Balachandran *et al.* (2017)[5]	None reported	N/A	N	Not specified
Chua *et al.* (2017)[11]	None reported	N/A	N	10 months
Lee *et al.* (2017)[32]	None reported	N/A	N	72 months (Mean: 6 years)
Xu *et al.* (2018)[56]	None reported	N/A	N	66.3 months (Mean: 5.5 years)
Lan *et al.* (2018)[30]	None reported	N/A	N	Ongoing follow-up
Nakashima *et al.* (2018)[39]	None reported	N/A	N	48 months (4 years)
Schär *et al.* (2019)[47]	None reported	N/A	N	24 months (2 years)
Burkes and Anderson (2019)[6]	None reported	N/A	N	72 months (6 years)
Chan *et al.* (2019)[8]	None reported	N/A	N	24 months (2 years)
Lim and Cho, (2020)[33]	None reported	N/A	N	12 months
Nakamura *et al.* (2019)[38]	None reported	N/A	N	122 months (Mean: 10.2 years)
Singh *et al.* (2019)[48]	None reported	N/A	N	2 months

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Table 4: (*Continued*).

Author (Year)	Complications	Management of Complications	Recurrence (Y/N)	Follow-up Duration (Months)
Champaneri and Banerjee, (2020)[7]	None reported	N/A	N	3 months
Gatineau-Sailliant *et al.* (2020)[21]	4 cases of vertebral recurrence	Extended maintenance chemotherapy	Y (4 cases)	34 months (Mean: 2.8 years)
Erdogan *et al.* (2021)[14]	None reported	N/A	N	Not specified
Foti *et al.* (2023)[16]	None reported	N/A	N	Not specified
Al-Salihi *et al.* (2023)[1]	None reported	N/A	N	Not specified
Chaulagain *et al.* (2023)[10]	None reported	N/A	N	1 month
Dayyani *et al.* (2023)[12]	Unknown	Unknown	Lost to follow-up	N/A
Otsuki *et al.* (2024)[40]	None reported	N/A	N	36 months (3 years)

LCH: Langerhans cell histiocytosis, , N/A: Non available, N: No, Y: Yes

improving outcomes and preventing permanent neurological deficits.[24,44] Although surgical decompression is often recommended, our findings indicate that many cases of spinal LCH can be effectively managed conservatively with corticosteroids and vinblastine-based chemotherapy.[32,56] Complete vertebral reconstitution was frequently observed, particularly in pediatric cases. However, surgery was necessary in cases presenting with instability or progressive neurological deterioration.[37,48] In such instances, posterior instrumentation and corpectomy were commonly performed, with excellent postoperative neurological recovery.[14,47] Long-term follow-up of our reviewed cases revealed favorable outcomes, with most patients achieving full neurological recovery and low recurrence rates. Spontaneous vertebral reconstitution was particularly observed in pediatric patients managed conservatively.[32,38] Surgical intervention for unstable lesions resulted in sustained neurological improvement without significant long-term complications.[14,37]

In conclusion, this case highlights the diagnostic challenges and importance of histopathological evaluation in cases of spinal lesions with atypical presentations. It also emphasizes the need for a high index of suspicion for LCH, particularly in regions where other granulomatous diseases are prevalent. Further research and case studies are needed to understand better the optimal management strategies and long-term outcomes for patients with spinal LCH.

CONCLUSION

The case report highlights the importance of considering LCH in diagnosing spinal lesions, especially in tuberculosis-endemic areas. Rapid neurological deterioration necessitated immediate surgical intervention, highlighting the role of biopsy in uncertain cases. A multidisciplinary approach is needed, involving neurosurgeons, radiologists, pathologists, and oncologists, for better understanding and management of this rare condition.

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