ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Professor of Clinical Neurosurgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Spine

Editor
Nancy E. Epstein, MD
Professor of Clinical Neurosurgery, School of Medicine, State U. of NY at Stony Brook

SNI
Open Access

Case Report

Rare histiocytic neoplasm: A case report

Sabrina Maria Genovese[1], Jakov Tiefenbach[2], Ravi A. Nunna[3], Andrew Youkilis[3]

[1]Department of Neurosurgery, University of Missouri, School of Medicine, Columbia, Missouri, [2]Department of Neurosurgery, University of Illinois, College of Medicine, Chicago, Chicago, Illinois, [3]Department of Neurosurgery, University of Missouri, Columbia, Missouri, United States.

E-mail: *Sabrina Maria Genovese - sgnnm@umsystem.edu; Jakov Tiefenbach - jakov.tiefenbach@yahoo.com; Ravi A. Nunna - nunnars@health.missouri.edu; Andrew Youkilis - ayoukilis@health.missouri.edu

***Corresponding author:**
Sabrina Maria Genovese, Department of Neurosurgery, University of Missouri, School of Medicine, Columbia, Missouri, United States.

sgnnm@umsystem.edu

Received: 21 September 2024
Accepted: 29 October 2024
Published: 22 November 2024

DOI
10.25259/SNI_795_2024

Quick Response Code:

ABSTRACT

Background: Histiocytic neoplasms are defined by too many histiocytes accumulating in various tissues, including the skin, bones, lymph nodes, and central nervous system. They are uncommon blood-related disorders that constitute <1% of cancers found in soft tissues and lymph nodes. Most referred to as Langerhans cell histiocytosis (LCH) or non-LCH, there are over 100 different sub-types that are divided into five groups. Here, a 76-year-old male presented with an intramedullary thoracic LCH.

Case Description: A 76-year-old male presented with the month of slowly progressive bilateral lower extremity weakness (i.e., right > left) accompanied by decreased left-sided sensation below the T7 level. The enhanced thoracic magnetic resonance (MR) imaging documented an intradural intramedullary nodule at the T5 level with a syrinx extending from C7 to T10. The patient underwent a T4–T6 laminectomy for complete resection of the lesion. CD163 and CD68 studies highlighted a small, spindled-shaped tumor with occasionally enlarged histiocytes without co-positivity for S100. Pathologically, the lesion was considered an isolated intramedullary thoracic LCH.

Conclusion: A 76-year-old male presented with progressive paraparesis of 1 month's duration attributed to an enhanced MR-documented T5 single intramedullary T5 thoracic LCH that was successfully resected.

Keywords: Histiocytic neoplasm, Histiocytosis-X, Langerhans cell histiocytosis, Rosai–Dorfman disease, Spine

INTRODUCTION

Histiocytic neoplasms make up <1% of cancers found in soft tissues and lymph nodes.[5] They include over 100 different sub-types divided into five distinct groups: Langerhans-related, cutaneous and mucocutaneous, malignant histiocytosis, Rosai–Dorfman disease, hemophagocytic lymphohistiocytosis, and macrophage activation syndrome.[4] Langerhans cell histiocytosis (LCH) occurs in 6.5–25% of all osteolytic skeletal cases involving, in descending order, the thoracic (54%), lumbar (35%), and cervical spine (11%).[1] Metastatic spread of LCH to the spinal cord is occasionally noted, but isolated intramedullary lesions are exceptionally rare.[8,6,12] Here, a 76-year-old male presented with a 1-month progressive paraparesis attributed to a magnetic resonance (MR)-documented isolated intramedullary T5 LCH that was successfully excised.

CASE REPORT

A 76-year-old male presented with progressive bilateral lower extremity paraparesis of 1 month's duration accompanied by a left-sided T7 pin level. His neurologic examination showed mild

right-sided (i.e., 4/5 hip flexion) but predominantly left-sided weakness (i.e., left 2/5 hip flexion/knee extension and 3/5 dorsiflexion/plantar flexion), accompanied by decreased pin appreciation on the left below the T7 level. The contrast thoracic MR showed a single intramedullary nodule at the T5 level with a syrinx extending from C7 to T10 [Figures 1a and b]. The chest computed tomography was normal. The original differential diagnoses included metastases to the spine, hemangioblastoma, astrocytoma, or ependymoma.

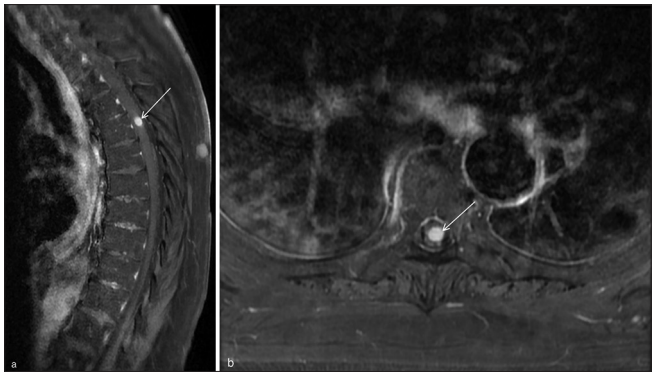

Figure 1: (a) Magnetic resonance imaging (MRI) of thoracic spine, sagittal view. The white arrow points to the intradural intramedullary nodule at T5, (b) MRI of the thoracic spine, axial view. The white arrow points to the intradural intramedullary nodule at T5.

Surgery

The patient underwent a T4–T6 laminectomy for complete resection of the intradural intramedullary C5 spinal cord lesion under neuromonitoring (Motor-evoked potential/Somatosensory-evoked potentials). The mass was pale yellow-whitish in color, well-encapsulated, and firm. Total *en bloc* excision was accomplished as there was a clear margin of demarcation of the tumor from the cord.

Pathology

Pathologic findings were diagnostic for an isolated intramedullary thoracic histiocytic neoplasm comprised of chronic inflammation with histiocytes. CD163 and CD68 studies highlighted small, spindled-shaped, and occasionally enlarged histiocytes [Figures 2a and 2b] without co-positivity for S100 [Figure 2c].

Postoperative course

Postoperatively, the patient was nearly paraplegic, requiring transfer to an inpatient rehabilitation center. When

Table 1: Cases of primary intramedullary histiocytic neoplasms.

Classification	Age LCH or non-LCH	Location	Symptoms	Treatment	Pathology	Adjunct treatment pursued
Hamilton *et al*.[6] JNS 1995	39 LCH	C4	Bilateral hand numbness followed by left leg numbness and muscle spasms	B/Dec	Histiocytes positive for S-100 protein	RT 20, GY local
Yamagata *et al*.[12] Neurol Med Chir (Tokyo) 2013	28 LCH	T3	Gait disturbance, back pain, and difficulty with sphincter control	Local LDRT	Histiocytes positive for CD1a	Steroid Pulse Therapy
Present Case Surg Neurol Int 2024	76 Non-LCH	T5	Bilateral lower extremity weakness and numbness	LAM/GTR	Lesional cells were positive for CD68 and S100	Steroids
Osenbach[9] JNS 1996	35 Non-LCH	T5	Paraplegia	LAM/GTR	Histiocytes positive for S–100	None
Jones and Rueda-Pedraza[7] Am J Hematol 1998	34 Non-LCH	T5	Paraplegia	LAM/GTR	Histiocytes positive for S–100 protein and Kp1 antigen	None
El Molla *et al*.[3] World Neurosurgery 2014	76 Non-LCH	C3–C4	Right hemiparesis	LAM/GTR	Histiocytes positive for S-100	None
Sandoval-Sus *et al*.[11] Medicine (Baltimore) 2014	53 Non-LCH	C5–C6	Unsteadiness, recurrent falls	B	Histiocytes positive for CD163, CD68, and S-100 but negative for CD1a	Steroids and RT
Rocha-Maguey *et al*.[10] Surg Neurol Int 2016	27 Non-LCH	C7–T1	Gait disturbances and decreased strength	LAM/GTR	Histiocytes positive for CD60, CD45/CD68	None

B: Biopsy, Dec: Decompression, LAM: Laminectomy, GTR: Gross total resection, LD: Low dose, RT: Radiation, LCH: Langerhans cell histiocytosis

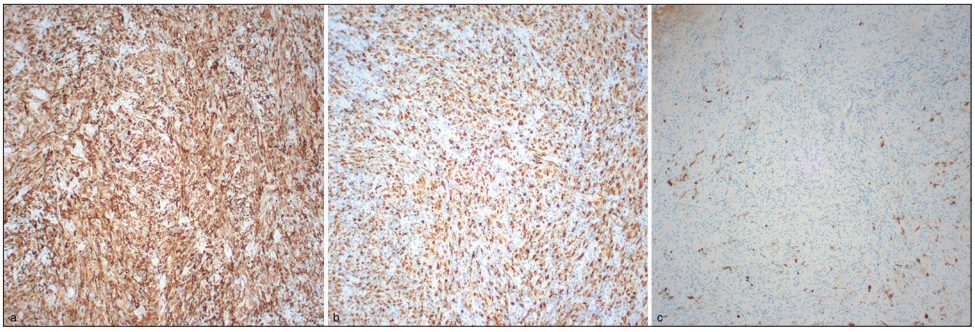

Figure 2: (a) Immunohistochemistry staining CD163 positivity of spindle shaped histiocytes. (b) CD68 positivity of spindle shaped histiocytes. (c) Histiocytes showed no S100 co-positivity.

reexamined at 4 postoperative months, left lower extremity motor function had improved to 3–4/5 proximal/distal, with the right-sided function now at 4–5/5. Nevertheless, he exhibited persistently decreased pin appreciation bilaterally below the T8 level.

DISCUSSION

Pathological diagnosis of histiocytosis

Confirmation of histiocytosis requires both biopsy and histological analysis. H&E stains revealed large, foamy histiocytic cells, while the basic immunohistochemistry panel included CD163/CD68, S100, CD1a, Langerin/CD207, cyclin D1, and factor XIIIa.[5,8]

Treatment of histiocytosis

The treatment for histiocytic neoplasms includes surgical resection, steroids, imiquimod, radiation therapy, and complex chemotherapy regimens [Table 1].[2] From the literature, we found two similar cases of isolated spinal histiocytic tumors. Hamilton *et al*., described a 39-year-old male who presented with bilateral hand numbness, leg numbness, and hyperreflexia.[6] A biopsy established the diagnosis of LCH; following partial tumor resection and postoperative radiation (i.e., 20 GY in fractions of 2 GY), the patient showed symptomatic improvement and reduced tumor size 1-year later. Yamagata *et al*. 28-year-old male with paraparesis and incontinence, underwent a biopsy of an histiocytic neoplasm, and T3-T5 osteoplastic laminotomy for partial tumor resection; postoperatively, steroid pulse therapy halted the further progression of the lesion and stabilization of his symptoms.[12] One year following the gross total resection of an isolated T5 intramedullary histiocytic neoplasm, our 76-year-old male demonstrated significant partial neurological recovery.

CONCLUSION

A 76-year-old male with 1 month of progressive paraparesis/left-sided T7 pin level underwent a T4–T6 laminectomy to remove an MR-documented enhancing single intramedullary T5 lesion; pathologically, the tumor was a LCH. Four months postoperatively, the patient exhibited an improved although moderate residual paraparesis, but with residual decreased bilateral pin appreciation below the T8 level.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Abu-Bonsrah N, Goodwin CR, Crane GM, Abiola G, Sciubba DM. Langerhans cell histiocytosis of the cervical spine. Spine J 2016;16:e11-2.

2. Allen CE, Ladisch S, McClain KL. How I treat Langerhans cell histiocytosis. Blood 2015;126:26-35.
3. El Molla M, Mahasneh T, Holmes SE, Al-Khawaja D. Rare presentation of rosai-dorfman disease mimicking a cervical intramedullary spinal cord tumor. World Neurosurg 2014;81:442.e7-9.
4. Emile JF, Abla O, Fraitag S, Horne A, Haroche J, Donadieu J, *et al*. Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. Blood 2016;127:2672-81.
5. Go RS, Jacobsen E, Baiocchi R, Buhtoiarov I, Butler EB, Campbell PK, *et al*. Histiocytic neoplasms, version 2.2021, NCCN clinical practice guidelines in oncology. J Natl Compr Canc Netw 2021;19:1277-303.
6. Hamilton B, Connolly ES, Mitchell WT. Isolated intramedullary histiocytosis-X of the cervical spinal cord: Case report. J Neurosurg 1995;83:716-8.
7. Jones MP, Rueda-Pedraza ME. Extranodal sinus histiocytosis with massive lymphadenopathy presenting as an intramedullary spinal cord tumor: A case report. Am J Hematol 1997;54:253-7.
8. Nayef RF, Ossama AM, John TG, John RJ, John LF. Histiocytosis-X of the spinal cord: A case report. Neurosurgery 1986;19:837-40.
9. Osenbach RK. Isolated extranodal sinus histiocytosis presenting as an intramedullary spinal cord tumor with paraplegia: Case report. J Neurosurg 1996;85:692-6.
10. Rocha-Maguey J, Felix-Torrontegui JA, Cabrera-López M, Gutiérrez-Castro M, Montante-Montes D. A new case of cervical intramedullary sinus histiocytosis causing paraplegia and review of the literature. Surg Neurol Int 2016;7:9.
11. Sandoval-Sus JD, Sandoval-Leon AC, Chapman JR, Velazquez-Vega J, Borja MJ, Rosenberg S, *et al*. Rosai-Dorfman disease of the central nervous system: Report of 6 cases and review of the literature. Medicine (Baltimore) 2014;93:165-75.
12. Yamagata T, Takami T, Yamamoto N, Tanaka S, Wakasa K, Ohata K. Primary intramedullary langerhans cell histiocytosis of the thoracic spinal cord. Neurol Med Chir (Tokyo) 2013;53:245-8.

How to cite this article: Genovese SM, Tiefenbach J, Nunna RA, Youkilis A. Rare histiocytic neoplasm: A case report. Surg Neurol Int. 2024;15:431. doi: 10.25259/SNI_795_2024