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Surgical Neurology International

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SNI: Neuro-Oncology

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Case Report

Burkitt's lymphoma presenting as multiple nerve sheath tumors of the cauda equina

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Received: 15 July 2021
Accepted: 28 June 2024
Published: 04 April 2025

DOI
10.25259/SNI_701_2021

Quick Response Code:

ABSTRACT

Background: Primary central nervous system lymphomas comprise 0.8% of all lymphomas. Burkitt's lymphoma is a B-cell lymphoma with highly aggressive features that rapidly progress and have early hematogenous spread. Sporadic Burkitt's lymphoma accounts for 1–2% of adult lymphomas worldwide. Extranodal sites are involved in approximately 40% of cases of B-cell lymphomas. Central nervous system disease is found in <15% of sporadic cases. Less than 5% of cases of all non-Hodgkin's lymphomas present with radiculopathy, with the thoracic spinal region being most commonly affected.

Case Description: In this report, we discuss the case of a patient with extranodal Burkitt's lymphoma within the nerve roots of the cauda equina who underwent surgical intervention due to the presence of lumbar radiculopathy and weakness. This patient also exhibited hematogenous spread with evidence of an intracranial lesion.

Conclusion: The diagnosis of Burkitt's lymphoma was atypical, given the presentation, clinical indicators, and the appearance of the masses on magnetic resonance imaging being similar to that of a neurofibroma or schwannoma. Surgical decompression provided tissue for biopsy. However, nerve root decompression was not possible due to diffuse involvement and infiltration of the nerve roots.

Keywords: Burkitt, Cauda equina, Lymphoma, Nerve sheath

INTRODUCTION

Primary central nervous system lymphomas comprise 0.8% of all lymphomas. Burkitt's lymphoma is a B-cell lymphoma with highly aggressive features that rapidly progress and have early hematogenous spread.[4] Sporadic Burkitt's lymphoma accounts for 1–2% of adult lymphomas worldwide.[2] Extranodal sites are involved in approximately 40% of cases of B-cell lymphomas.[1] Central nervous system disease is found in <15% of sporadic cases.[1] Less than 5% of cases of all non-Hodgkin's lymphomas present with radiculopathy, with the thoracic spinal region being most commonly affected.[3] A handful of case reports have detailed B-cell lymphoma as infiltrating nerve roots and ganglia.[5] Here, we present a patient with extranodal Burkitt's lymphoma involving the roots of the cauda equina with suspected hematogenous spread. Although the initial differential diagnosis included neurofibroma and schwannoma, the final pathological diagnosis was lymphoma, creating an emphasis that these lesions should be

considered when assessing multifocal intradural nerve root tumors in all regions of the spine.

CASE DESCRIPTION

A 40-year-old male presented with 6 weeks of left foot drop and multidermatomal radiculopathy of the lumbar spine. The patient also had a recent history of left-sided peripheral nerve palsy of the seventh cranial nerve (Bell's palsy) with bilateral lateral rectus palsy following a viral respiratory illness. He exhibited 2/5 dorsiflexion and extensor hallucis longus and 4/5 plantar flexion strength on the left. The patient was also noted to have axillary freckling of the right axilla. However, no family history of neurofibromatosis was evident.

Magnetic resonance (MR) image of the lumbar spine revealed enhancing intradural masses along the course of the L3, S1, and S2 nerve roots, resulting in severe canal stenosis, as shown in Figures 1 and 2. MR brain revealed a 1.8-cm enhancing mass of the right internal auditory canal consistent with vestibular schwannoma with no other notable lesions or abnormalities [Figure 3]. A positron emission tomography (PET) scan revealed uptake within the left supraclavicular node. A lumbar puncture was performed, and cerebrospinal fluid analysis was determined to be inconclusive. Given the presence of radicular symptoms, weakness, and the need for tissue diagnosis, surgical intervention was pursued with the goal of decompression, diagnosis, and resection of the nerve root lesions.

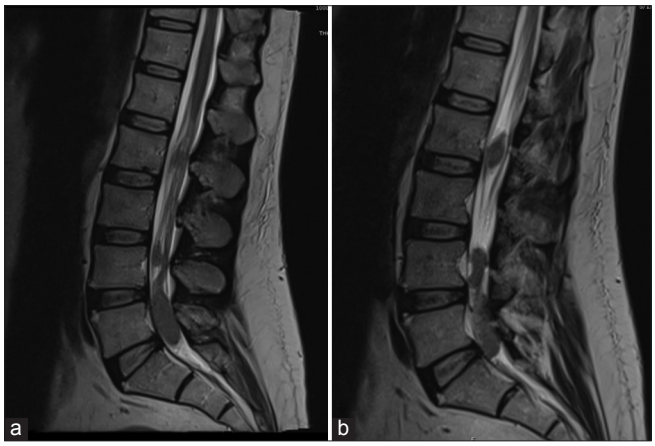

Figure 1: (a) T2 magnetic resonance imaging (MRI) lumbar spine. Sagittal view. Intradural masses located at L2, L4, and L5/S1 vertebral body levels. (b) T2 MRI lumbar spine. Left parasagittal view. Intradural masses located at L2, L4, and L5/S1 vertebral body levels.

Surgery and postoperative status

The patient underwent L1–S1 laminectomies and expansion duraplasty with subtotal resection of the L3 mass. Each mass appeared to be infiltrative and expansile of their respective nerve roots, as shown in Figure 4. Intraoperative neuromonitoring was used to identify nerve roots with viable function. Only a small portion of the L3 mass was able to be resected without causing additional loss of function. The intraoperative assessment was consistent with neurofibroma, given the appearance and infiltrative nature of the lesions, as well as the locations. Due to limited resection and tissue volume, a frozen section was not obtained for intraoperative analysis. Postoperatively, the patient was noted to have a persistent and stable weakness with retained bowel and bladder function. The final pathology confirmed Burkitt's lymphoma [Figure 5].

Chemotherapy and postoperative follow-up

The patient was initially started on high-dose methotrexate and rituximab in addition to intrathecal cytarabine, followed by a regimen of rituximab, etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin, and continued methotrexate.

Follow-up imaging at 1 month revealed decreased size of the intracranial lesion and decreased size of the lumbar

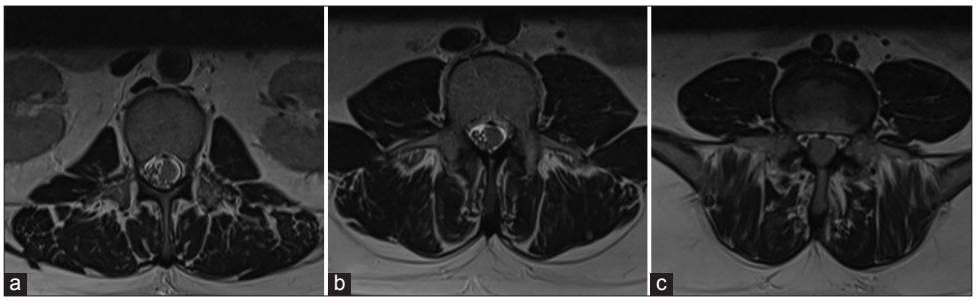

Figure 2: (a) T2 magnetic resonance imaging (MRI) lumbar spine. Axial. Intradural masses are located at the L2 vertebral body level. (b) T2 MRI lumbar spine. Axial. Intradural masses are located at the L4 vertebral body level. (c) T2 MRI lumbar spine. Axial. Intradural masses are located at the L5/S1 vertebral body level.

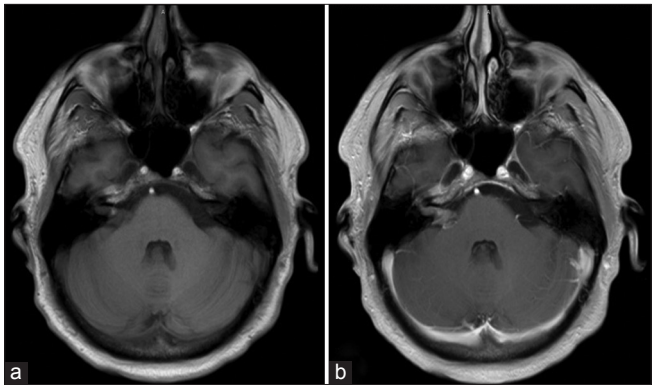

Figure 3: (a) T1 magnetic resonance imaging (MRI) without contrast. (b) T1 MRI with contrast. Enhancing mass of the right internal auditory canal.

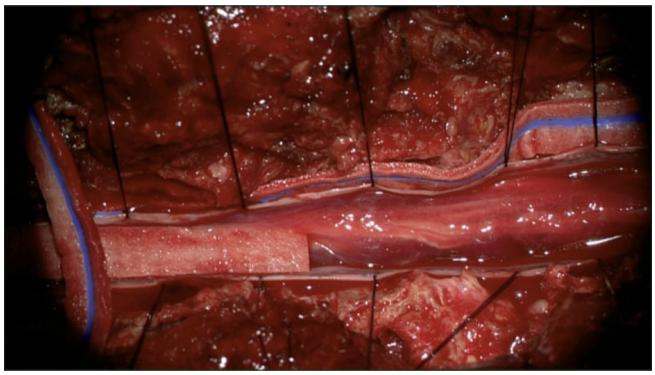

Figure 4: Intraoperative view of intradural expansile nerve root lesions.

masses, as shown in Figure 6. One month after beginning pharmacologic therapy, an Ommaya reservoir was placed to continue intrathecal methotrexate therapy, which was made. His neurological examination remained unchanged for 7 months, at which time he reported severe headaches and vision changes. MR brain revealed recurrence and progression of his disease. He resumed the previously noted chemotherapy regimen. He subsequently developed sepsis from myelosuppression and passed 1 year after beginning treatment.

DISCUSSION

Primary central nervous system lymphoma is a rare clinical entity but should be included within the differential diagnosis in which multiple nerve sheath lesions are present. The patient, in this case, presented with acute radiculopathy and weakness, which was likely indicative of the infiltrative and aggressive nature of the pathology. In addition, the prodromal symptoms of cranial nerve dysfunction should serve as an indication of more systemic

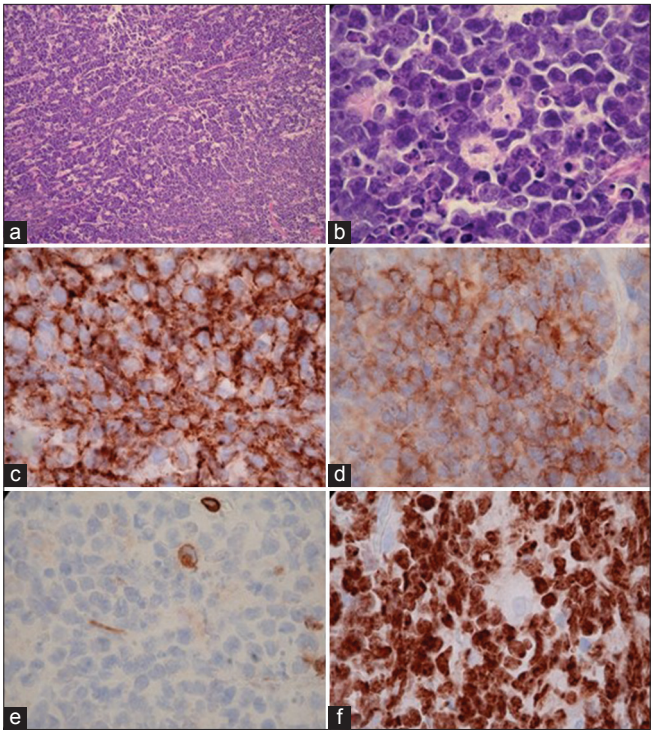

Figure 5: (a) Burkitt. ×100 is a lower power view that shows densely packed dark lymphoma cell nuclei and scattered light-colored spaces. This so-called "starry sky" pattern is characteristic of Burkitt lymphoma. (b) Burkitt. ×1000 is a high-power oil immersion view. The light-colored spaces are macrophages ("stars") surrounded by lymphoma cells ("sky"). (c) CD20: The cell membranes of lymphoma cells are positive for this B-cell marker by immunohistochemistry. (d) CD10: Weaker than CD20 but still positive on the cell membranes. (e) Bcl-2: Negative in the lymphoma cells. We use a light blue hematoxylin counter stain to see the nuclei in immunohistochemical slides. (f) Ki-67: 100% of lymphoma cells are positive for this proliferation marker. Note that this localizes to the nucleus rather than the cell membrane or cytoplasm. The pale blue nucleus in the central macrophage is negative for Ki-67.

disease despite the absence of radiographic evidence of cranial lesions to corroborate the symptoms. These clinical manifestations, along with increased PET update within the left supraclavicular lymph node, were likely, not unrelated aberrations following a respiratory viral illness but were the initial manifestations of the disease process. This is notable given the propensity of hematogenous spread of B-cell lymphomas.

A thorough evaluation with a comprehensive differential diagnosis is critical. A complete history may direct the differential diagnosis toward a rarer clinical entity, such as in this case. Although the clinical impression intraoperatively was incorrect, the course of treatment and method of diagnosis was appropriate given the history, presentation, and imaging findings.

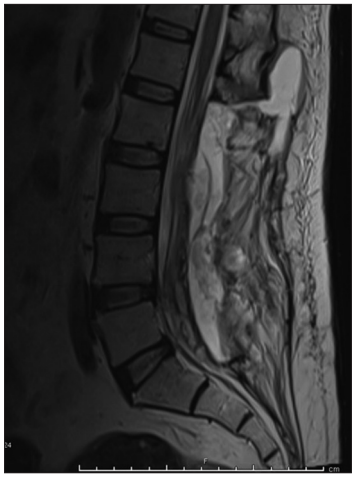

Figure 6: T2 magnetic resonance imaging lumbar spine. Sagittal view. Reveals a significant decrease in the size of the intradural masses located at L2, L4, and L5/S1 vertebral body levels. Also noted is a postoperative pseudomeningocele.

CONCLUSION

Primary central nervous system lymphoma is rare with only a handful of case reports available. We previously published a similar case of a patient with extranodal diffuse large B-cell lymphoma involving cervical nerve roots. Although the initial differential diagnosis included neurofibroma, schwannoma, and meningioma, the correct pathological diagnosis was lymphoma.[4] Based on our experience, lymphoma should be considered when assessing multifocal intradural nerve root tumors.

In the case presented, a left-sided foot drop correlated with MR-confirmed cauda equina masses. Decompressive laminectomy with expansion duraplasty revealed Burkitt's lymphoma treated with chemotherapy.

Ethical approval: The research/study complied with the Helsinki Declaration of 1964.
Declaration of patient consent: Patient's consent not required as patient's identity is not disclosed or compromised.
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. Hughes RA, Britton T, Richards M. Effects of lymphoma on the peripheral nervous system. J R Soc Med 1994;87:526-30.
2. Kalisz K, Alessandrino F, Beck R, Smith D, Kikano E, Ramaiya NH, *et al*. An update on Burkitt lymphoma: A review of pathogenesis and multimodality imaging assessment of disease presentation, treatment response, and recurrence. Insights Imaging 2019;10:56.
3. Kim YS, Lee JK, Choi KY, Jang JW. Spinal Burkitt's lymphoma mimicking dumbbell shape neurogenic tumor: A case report and review of the literature. Korean J Spine 2015;12:221-4.
4. Milton J, Renner J, Awuor V. B-cell lymphoma presenting as multiple nerve sheath tumors. Surg Neurol Int 2017;8:142.
5. Shibata-Hamaguchi A, Samuraki M, Furui E, Ishida C, Kitagawa S, Nakao S, *et al*. B-cell neurolymphomatosis confined to the peripheral nervous system. J Neurol Sci 2007;260:249-52.

How to cite this article: Milton J, Pandya P, Ulloa S, Kudithipudi V, Sarwar S, Awuor V. Burkitt's lymphoma presenting as multiple nerve sheath tumors of the cauda equina. Surg Neurol Int. 2025;16:125. doi: 10.25259/SNI_701_2021