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Case Report

B-cell lymphoma presenting as multiple nerve sheath tumors

Jason Milton¹, Julie Renner², Victor Awuor^{1,2}

¹Grant Medical Center, Ohio Health, Columbus, ²Department of Neurosurgery, Genesis Healthcare System, Zanesville, Ohio, USA

E-mail: *Jason Milton - Jason.Milton@ohiohealth.com; Julie Renner - julie.dobos@gmail.com; Victor Awuor - Victor.Awuor@ohiohealth.com *Corresponding author

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Abstract

Background: Diffuse Large B-cell Lymphoma (DLBCL) is the most common form of Non-Hodgkin lymphoma (NHL), accounting for 25-30 percent of cases in the United States.1 Extranodal sites are involved in approximately 40% of cases of DLBCL.

Case Description: In this report, we discuss the case of a patient with extranodal DLBCL within the cervical nerve roots that underwent surgical intervention due to the presence of cervical radiculopathy.

Conclusion: The diagnosis of DLBCL was surprising given the appearance of the masses on MRI being similar to that of a neurofibroma or schwannoma. Surgical decompression provided a tissue sample for biopsy as well as an opportunity for decompression of the nerve roots and restoration of function of the patient's left upper extremity.

Key Words: B-cell, lymphoma, spine, tumor



INTRODUCTION

Diffuse large B-cell lymphoma (DLBCL) accounts for 25–30% cases of non-Hodgkin lymphoma (NHL) in the United States.^[1] The most common sites include the gastrointestinal tract and bone marrow. Extranodal sites are involved in approximately 40% of the cases. Primary central nervous system lymphomas, however, comprise only 0.8% of all lymphomas.^[2] Few case studies report the primary manifestation of DLBCL as infiltrating nerve roots and ganglia. Here, we present a patient with extranodal DLBCL involving the cervical nerve roots. Although the initial differential diagnosis included neurofibroma, schwannoma, and meningioma, the correct pathological diagnosis was lymphoma, and clearly these lesions should be considered when assessing multifocal intradural nerve root tumors.

CASE DESCRIPTION

For 7 months, a 63-year-old female complained of left-sided neck pain and left upper extremity radiculopathy accompanied by progressive numbness and weakness. She exhibited 4/5 strength proximally and distally in the left arm alone.

Electromyography (EMG) of the bilateral upper extremities demonstrated a left C6 and C7 radiculopathy. Magnetic resonance image (MRI) of the cervical spine revealed enhancing masses involving the left neural foramina at the C5-6, C6-7, and C7-T1 levels accompanied by nodular thickening of the left exiting nerve root at T1-2 level (T1 root) as revealed in Figure 1. In addition, there was diffuse enhancement and thickening of the proximal left brachial plexus and upper trunk at the level of the left proximal pulmonary apex.

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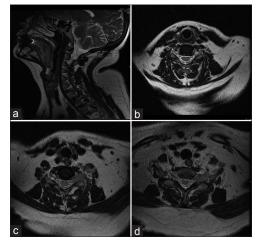


Figure 1: T2 MRI Cervical Spine. (a) Left parasagittal view, C5 and C6 isointense masses noted, (b-d) axial views of C5/6, C6/7, C7/TI with left isointense masses noted

Surgery and postoperative status

The patient underwent a C6-7-T1 hemilaminectomy and facetectomy with subtotal resection of the C7 and T1 intradural nerve root tumors; this was accompanied by an instrumented fusion from C4-T7 with intraoperative neuromonitoring. The frozen section was equivocal. Postoperatively, the patient noted residual proximal left upper extremity hyperesthesias, with proximal LUE strength of 4/5 but distal increased weakness to the 3/5 level. Motor function in the right upper extremity (RUE) was 4/5 and both lower extremities were neurologically intact.

The pathology confirmed B-cell lymphoma as shown in Figure 2. Furthermore, cerebral spinal fluid sampling (lumbar puncture) showed no malignant cells.

Chemotherapy and postoperative follow-up

The patient underwent chemotherapy consisting of 6 cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP). In addition, she received two doses of systemic methotrexate and two doses of intrathecal methotrexate. Prior to beginning pharmacologic therapy, a postoperative MRI was obtained, which was of poor quality due to the presence of hardware. Six months after her diagnosis, emission tomography-computed repeat positron tomography was performed without evidence of residual mass or abnormal metabolic activity. Her neurological examination remained unchanged. She will undergo oncologic surveillance at 12 months and then will undergo follow-up yearly thereafter. At 6 months, the patient was considered to be in remission.

CONCLUSION

Nerve root infiltration has been reported in both T-cell and B-cell NHL, however, nervous system involvement

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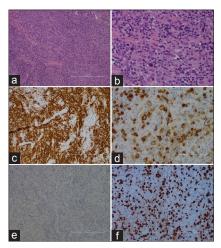


Figure 2: Histology. (a and b) H and E permanent sections reveal pleomorphic population of mononuclear cells including small and large forms at x10 and x40 magnification respectively, (c) CD20 immuno-histochemical staining reveals strong CD20 immunoreactivity, (d) CD79a immunohistochemical staining reveals strong CD79a immunoreactivity, (e) tissue biopsies are without GFAP immunoreac-tivity, (f) MIB-1 highlights increased mitotic activity

does not usually occur in the absence of widespread disease.^[3] In the literature, there were only a handful of case reports of patient s presenting with DLBCL of a nerve root without disseminated disease.^[2-7]

In our patient, a left-sided C6 and C7 radiculopathy correlated with MR-confirmed multifocal left-sided foraminal lesions. The decompressive laminectomy with instrumented fusion yielded B-cell lymphoma followed by appropriate chemotherapy.

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Conflicts of interest

There are no conflicts of interest.

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