Successful Emergency Decompressive Laminectomy for Burkitt Lymphoma with Metastatic Spinal Lesions: A Case Report

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Although Burkitt lymphoma (BL) usually arises in the abdomen or pelvis, it can also arise in the epidural space as a primary or secondary site and present with back pain or limb weakness. Emergency management is necessary to relieve spinal cord compression (SCC). Herein, we report a case of BL with metastatic spinal lesions in a 16-year-old female who presented with sudden-onset progressive walking difficulty. She was admitted to a previous hospital where she presented with abdominal pain and vomiting and was diagnosed with intussusception via a computed tomography scan. Laparoscopic small bowel resection was performed, during which a diagnosis of BL was made on the basis of pathological examination. Sudden numbness in the extremities and the complete inability to walk occurred ten days after surgery. Thoracolumbar MRI revealed a metastatic mass extending from C7 to T6 with evidence of SCC. Emergency decompressive laminectomies (from C7 to T6) and partial debulking of the tumor were performed 12 hours after the onset of her neurological symptoms.

She was subsequently treated with chemotherapy, and she made a complete neurologic recovery. Emergency decompressive laminectomies for BL with spinal lesions could effectively lead to the recovery of neurologic symptoms.

Key words: Burkitt lymphoma, spinal lesion, laminectomy, oncologic emergency

INTRODUCTION

Burkitt lymphoma (BL), a common childhood tumor, has three distinct subtypes, including the endemic, sporadic, and immunodeficiency-associated subtypes. Although BL represents less than 5% of adult lymphoma, it accounts for approximately 40% of childhood non-Hodgkin lymphoma (NHL) [1]. The pediatric sporadic form arises from the abdomen in 60%–80% of cases, while the incidence of primary epidural cases is unclear [2, 3]. Only 1%–4% of all pediatric NHL cases primarily originate from the spinal epidural regions; however, secondary involvement is more common [4, 5]. Pediatric tumors located in the spinal regions present with symptoms of spinal cord compression (SCC) and require urgent management because of the progressive neurological dysfunction they cause [6]. Herein, we present the case of a patient suffering from BL with metastatic spinal lesions who was successfully managed via an emergency decompressive laminectomy as an oncologic emergency.

CASE

A 16-year-old female was admitted to a previous hospital for abdominal pain and vomiting. Computed tomography (CT) scans revealed evidence of a target sign in the small intestine suggesting intussusception, bilateral ovarian swelling, and masses in her right breast (Fig. 1: A, B, C). Laparoscopic small bowel resection involving the mass was performed. Histopathology revealed lymphoid cells with a typical “starry sky” pattern, the classic description for marked apoptosis with many macrophages. Flow cytometry revealed that the cells expressed CD10, CD19, CD20, CD22, CD38, and HLA-DR. The cells expressed high levels of c-MYC (95%). Paraffin block confirmed c-MYC/IGH fusion through fluorescence in situ hybridization panel. A diagnosis of BL was made from these findings. No image evaluation of the thoracolumbar region was performed at this point. She left the hospital and prepared for chemotherapy at our hospital. However, she developed sudden numbness in her extremities and a complete inability to walk ten days after surgery; therefore, she was suddenly hospitalized in our hospital. She was noted to have significant bilateral weakness in her lower extremities and was not able to stand up. Her muscle strength per the manual muscle test (MMT) was 3 out of 5 for the left lower extremity and 2 out of 5 for the right lower extremity. Deep tendon reflexes were absent bilaterally. Thoracolumbar MRI revealed a metastatic mass extending to the epidural space from C7 to T6 (Fig. 2A). Epidural involvement of the soft tissue mass resulted in visible severe SCC from C7 to T6 (Fig. 2B). Hemogram, laboratory, and coagulation tests showed the following: white blood cell count, 6,100/μL; hemoglobin concentration, 12.9 g/dL; platelet

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count, $39.4 \times 10^4 / \mu L$; AST, 29 U/L; ALT, 30 U/L; LD, 575 IU/L; total bilirubin, 0.3 mg/dl; CRP, 0.76 mg/dl, activated partial thromboplastin time (aPTT), 29 s (normal 26–36 s); prothrombin time (PT), 13.1 s (normal 10–15 s), prothrombin time in international normalized ratio (PT-INR), 1.11 (normal 0.9–1.1). B symptoms such as fever, weight loss, and night sweats were not seen. Due to the rapidly-progressive neurologic deficits, the decision was made to perform an emergency decompression 12 hours after the onset of neurologic symptoms. Complete C7–T6 laminectomies were carried out and the mass was visualized. The mass was noted to protrude dorsally after the laminectomies, suggesting compression before the procedure. The mass was subsequently removed from the dorsal aspect of the spinal cord's dura. Pulsation of dura appeared and decompression of the cord was achieved (Fig. 2C). The procedure was well-tolerated and without complication. The histopathologic findings of the lesion were the same as those of abdominal tumor cells. After surgery, we performed bone marrow (BM) aspiration and lumbar puncture. Although cerebrospinal fluid

Fig. 1 Computed tomography (CT) findings on admission
A. Abdominal CT: Intussusception with a mass in her small intestine (arrow).
B. Pelvic CT: Bilateral ovarian swelling (arrows).
C. Chest CT: A mass in her right breast (arrow).

Fig. 2 A. T2-weighted MRI preoperative image showing an extensive mass in the epidural space from C7 to T6 (arrows).
B. Preoperative CT showing spinal cord compression by a mass.
C. Postoperative CT showing the removal of laminae, debulking of the mass, and resolution of the spinal cord compression.
infiltration involvement was not present, BM aspiration and histopathology revealed 90% of BL cells. Therefore, she was diagnosed with Stage IV BL and subsequently received chemotherapy including intrathecal injection according to the B-NHL03 regimen proposed by the Japan pediatric leukemia/lymphoma study group from the day after surgery. Her muscle strength per the MMT test revealed a score of 4 out of 5 in both lower extremities on postoperative day 3. She could stand up using lower extremity orthoses on day 5 and walk with support on day 10. Also, she could walk freely seven months after surgery. Complete remission was achieved; she is alive and has attained full neurological recovery 18 months after the operation.

**DISCUSSION**

The incidence of primary epidural sporadic BL in the pediatric population is unclear due to its rarity. Approximately 1–4% of all pediatric NHL cases primarily originate from the spinal epidural regions, and secondary involvement is more common [4, 5]. Although the primary site was unknown in our case, the spinal epidural regions were considered secondary involvement. Kurucu et al. reported a case series of 84 cases of primary paraspinal epidural lymphomas including 33 cases of BL [7]. From the report, the most common tumor location was the thoracic region (52.4%), followed by the cervical (13.1%), lumbar (10.7%), and thoracolumbar (8.3%) regions. Back pain was the most common presenting symptom, and weakness and numbness of the extremities were the second most common reason for admission to the hospital. The most common neurologic finding was motor weakness (92%), followed by sensory deficits (47.3%), bladder dysfunction (35.7%), changes in reflexes (18.4%), and bowel dysfunction (17.1%). Although the time from the onset of symptoms to diagnosis varies from one day to eight months in all cases of pediatric NHL that involve spinal epidural regions [7], Dechambenoit et al. reported that most patients present within approximately seven days of symptom onset in BL cases due to the rapid growth capacity [8]. Our patient suddenly experienced numbness and weakness in both of her lower extremities, which was consistent with the findings of the previous report.

Emergency management entails spinal cord decompression for BL with SCC. From the previous case series, 64 out of 81 patients (except three patients whose treatment data were not available) and 27 out of 31 BL patients (except two patients whose treatment data were not available) underwent decompressive laminectomies [7]. Among the 64 cases with neurologic outcomes, complete and partial recoveries of neurologic deficits were achieved in 36 (56.2%) and 11 (17.2%) patients respectively, while in 17 (26.5%) patients there was no recovery. Important factors such as laminectomy-related neurologic outcomes were not known because it was a case series. Although laminotomy and laminoplasty could be preferred to laminectomy as an oncologic emergency, indications for cases with rapidly-progressive myelopathy and paralysis might be effective in resolving neurologic symptoms for cases with rapidly-progressive myelopathy and paralysis.

**CONCLUSION**

We report the case of a BL patient with spinal lesions and SCC who was successfully treated via an emergency decompressive laminectomy. Urgent management is necessary to ameliorate the SCC. Although chemotherapy may provide a rapid decompressive response, surgical decompression might be considered in the case of progressive myelopathy and paralysis. Prospective studies are necessary to identify neurologic prognostic factors.

**DECLARATION OF CONFLICTING INTERESTS**

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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**INFORMED CONSENT**

Written informed consent was obtained from the patient for their anonymized information to be published in this article.
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