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

Hidetoshi YANO, Takashi KOIKE, Mayuko SHIBATA, Yumiko SUGISHITA, Naoko KAWABATA, Sachio FUJITA, Kosuke AKIYAMA, Daisuke TOYAMA, Yoshiyuki YAMADA, Atsushi UCHIYAMA and Shohei YAMAMOTO

Department of Pediatrics, Tokai University School of Medicine

(Received December 15, 2022; Accepted February 3, 2023)

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 neurologic 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

Shohei YAMAMOTO, Department of Pediatrics, Tokai University School of Medicine, 143 Shimokasuya, Isehara, Kanagawa 259-1193, Japan
Tel: +81-463-93-1121 Fax: +81-463-94-3426 E-mail: shohei-y@tsc.u-tokai.ac.jp

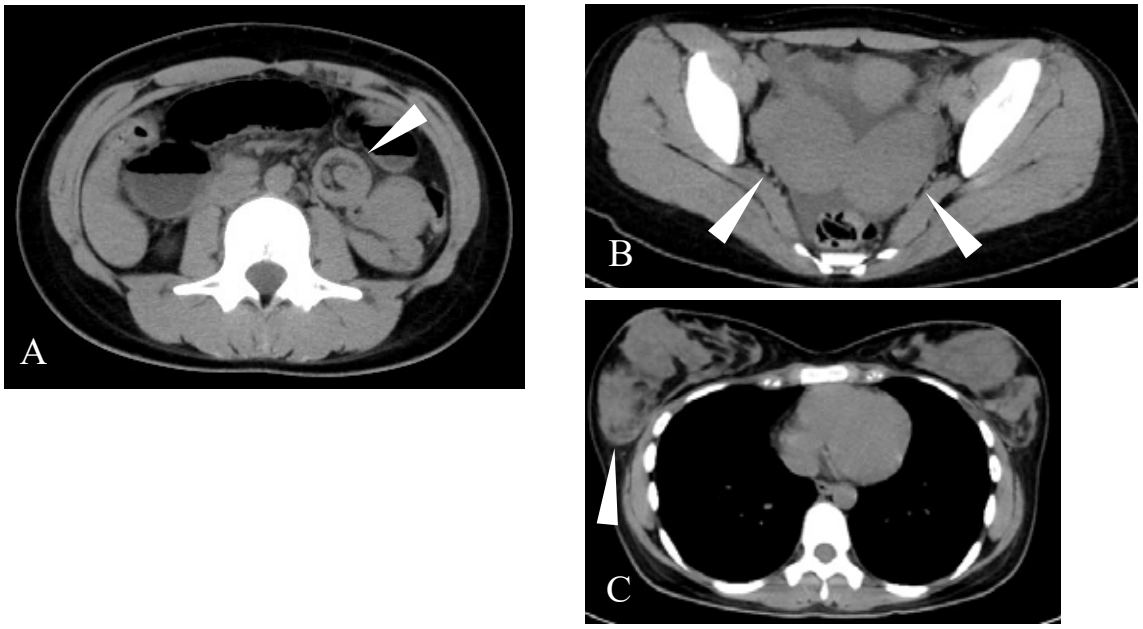

Fig. 1 Computed tomography (CT) findings on admission
A. Abdominal CT: Intussusception with a mass in her small intestine (arrow).
B. Pelvic CT: Bilateral ovarian swelling (arrows).
C. Chest CT: A mass in her right breast (arrow).

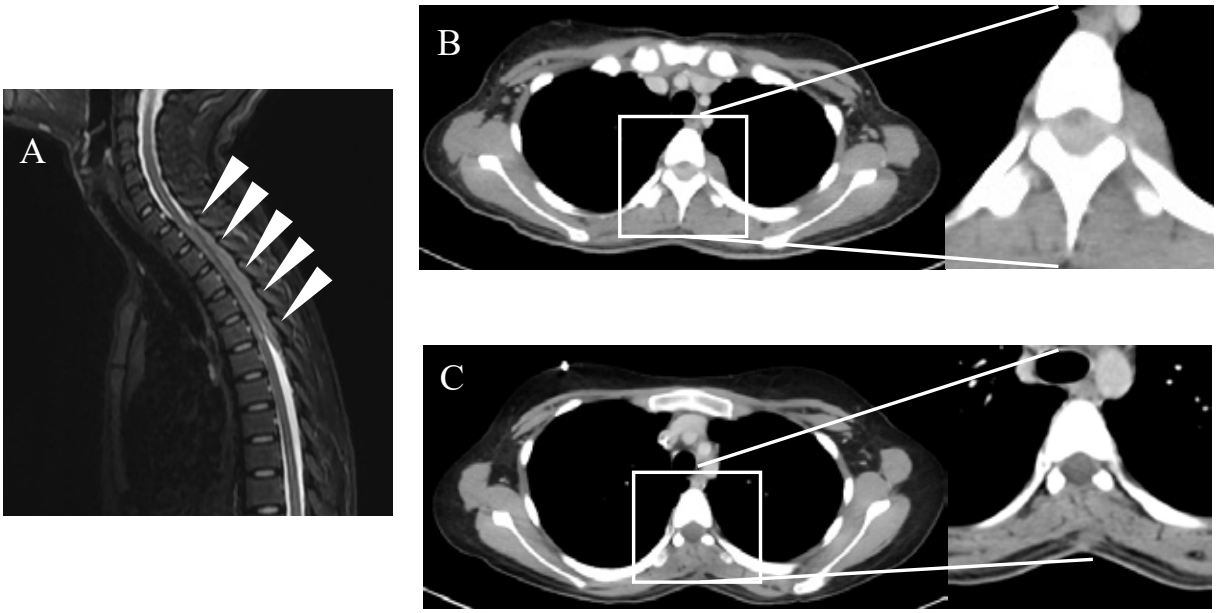

Fig. 2 A. T2-weighted MRI preoperative image showing an extensive mass in the epidural space from C7 to T6 (arrows).
B. Preoperative CT showing spinal cord compression by a mass.
C. Postoperative CT showing the removal of laminae, debulking of the mass, and resolution of the spinal cord compression.

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); prothorombin time (PT), 13.1 s (normal 10–15 s), prothrombin tine 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). Theprocedure was well-tolerated and without complication. The histopathologic findings ofthe lesion were the same as those of abdominal tumor cells. After surgery, we performed bone marrow (BM) aspiration and lumbar puncture. Although cerebrospinal fluid

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 to avoid subsequent spinal instability and there is a greater risk of kyphotic deformity in patients who require a laminectomy involving four or more levels [9], Dho *et al.* reported no differences in long-term function or deformity between epidural BL patients who underwent laminectomies and those who underwent laminotomies [5]. Fortunately, our patient did not require a laminoplasty since she did not experience spinal instability. Importantly, it has been well-understood that chemotherapy may provide a rapid decompressive response, and surgery is no longer preferred as a standard treatment option [4, 5, 10]. However, emergency decompressive laminotomy or laminectomy might be considered in the case of progressive myelopathy and paralysis since the effects of chemotherapy and radiation therapy are much more delayed than those of surgery [11, 12]. Therefore, indications for irradiation over surgery may include patients whose symptoms are subacute and mild. The time from the onset of symptoms to surgery and preoperative neurologic statuses are important prognostic factors for long-term postoperative outcomes [11]. Patients with lower extremity muscle strength scores of 0–1 often have poor neurological recovery after surgery, while most of those with muscle strength scores of ≥ 2 can often walk after surgery [5]. The time from the onset of neurological symptoms to diagnostic imaging was only one day in our patient. Her muscle strength score was 3 out of 5 for the left lower extremity and 2 out of 5 for the right lower extremity. Due to the rapidly-progressive myelopathy and paralysis, the orthopedic surgeon decided to perform emergency decompression 12 hours after the onset of neurologic symptoms. She can walk freely and has achieved complete neurological recovery. Although chemotherapy should be preferred to surgery, especially for patients with subacute forms and mild progression with neurological symptoms in BL patients with spinal lesions, surgical decompression including laminectomies as an oncologic emergency 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 neurological 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.

ETHICAL APPROVAL

Our institution does not require ethical approval for reporting individual cases or case series.

FUNDING

The authors received no financial support for the research, authorship, and/or publication of this article.

INFORMED CONSENT

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

REFERENCES

1) Bishop PC, Rao VK, Wilson WH. Burkitt's lymphoma: Molecular pathogenesis and treatment. Cancer Invest. 2000; 18: 574–583.
2) Mbulaiteye SM, Biggar RJ, Bhatia K, Linet MS, Devesa SS. Sporadic childhood Burkitt lymphoma incidence in the United States during 1992–2005. Pediatr Blood Cancer. 2009; 53: 366–370.
3) Saleh K, Michot JM, Camara-Clayette V, Vassetsky Y, Ribrag V. Burkitt and Burkitt-like lymphomas: A systematic review. Curr Oncol Rep. 2020; 22: 33.
4) Pui CH, Dahl GV, Hustu HO, Murphy SB. Epidural spinal cord compression as the initial finding in childhood acute leukemia and non-Hodgkin lymphoma. J Pediatr. 1985; 106: 788–792.
5) Dho YS, Kim H, Wang KC, Kim SK, Lee JY, Shin HY, *et al*. Pediatric spinal epidural lymphoma presenting with compressive myelopathy: A distinct pattern of disease presentation. World Neurosurg. 2018; 114: e689–97.
6) Pollono D, Tomarchia S, Drut R, Ibanez O, Ferreyra M, Cedola J. Spinal cord compression: A review of 70 pediatric patients. Pediatr Hematol Oncol. 2003; 20: 457–466.
7) Kurucu N, Akyuz C, Varan A, Akcoren Z, Aydin B, Yalcin B, *et al*. Primary paraspinal and spinal epidural non-hodgkin lymphoma in childhood. J Pediatr Hematol Oncol. 2021; 43: e395–400.
8) Dechambenoit G, Piquemal M, Giordano C, Cournil V, Zeze VB, Santini JJ. Spinal cord compression resulting from Burkitt's lymphoma in children. Child's Nerv Syst. 1996; 12: 210–214.
9) Fassett DR, Clark R, Brockmeyer DL, Schmidt MH. Cervical spine deformity associated with resection of spinal cord tumors. Neurosurg Focus. 2006; 20: E2.
10) Papagelopoulos PJ, Peterson HA, Ebersold MJ, Emmanuel PR, Choudhury SN, Quast LM. Spinal column deformity and instability after lumbar or thoracolumbar laminectomy for intraspinal tumors in children and young adults. Spine. 1997; 15: 442–451.
11) Rajz G, Cohen JE, Harnof S, Knoller N, Goren O, Shoshan Y, *et al*. Spontaneous spinal epidural hematoma: The importance of preoperative neurological status and rapid intervention. J Clin Neurosci. 2015; 22: 123–128.
12) 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: 221–224.