



## Case Report

# Primary epidural sporadic Burkitt lymphoma in a 3-year-old: Case report and literature review

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## ABSTRACT

**Background:** Burkitt lymphoma (BL) is a common tumor of childhood that usually arises in the abdomen or pelvis in its sporadic form. In a minority of cases, BL can present with CNS involvement, usually as a secondary site. Rarely, BL can arise primarily in the epidural space and present with back pain, or less commonly, acute myelopathy. This presentation is a surgical emergency and requires vigilant management.

**Case Description:** We describe a case of pediatric BL arising primarily within the epidural space and presenting with progressive difficulty walking in a 3-year-old boy. Progression to complete inability to walk, absent lower extremity deep tendon reflexes, and new urinary incontinence prompted MRI of the spine, which showed a lesion extending from T5 to T10 and wrapping around the anterior and posterior portions of the spine with evidence of spinal cord compression. The patient underwent decompressive laminectomies from T5 to T10 and partial debulking of the posterior portions of the tumor. Microscopic examination showed a prominent “starry sky” pattern with abundant mitotic figures. Immunohistochemistry confirmed the diagnosis of BL. The patient is 10 months post-op and continues to undergo chemotherapy with partial neurologic improvement. He was free of recurrence 10 months post-operative.

**Conclusion:** This appears to be the youngest described patient presenting with acute myelopathy in primary paraspinal BL. Management should include surgical decompression of the spinal cord followed by one of the various described chemotherapeutic regimens. Preoperative staging and neurologic function correlate with prognosis.

**Keywords:** Burkitt lymphoma, Pediatric lymphoma, Pediatric myelopathy, Pediatric paraspinal tumor, Spinal neoplasia

## INTRODUCTION

Burkitt's lymphoma (BL) is a common childhood tumor first described and eponymously named in 1958 by Denis Burkitt, after observing multiple children with jaw tumors in a Ugandan hospital.<sup>[7]</sup> It was originally thought to have two forms: lymphoid and leukemic (>25% bone involvement); however, it is now recognized as two phases of the same disease progression.<sup>[28]</sup> BL has three recognized clinical variants that have similar clinical presentations, oncogenetics, and immunophenotypes. Each of these variants has a characteristic rearrangement of the MYC oncogene, resulting in constitutive expression of c-myc.<sup>[41]</sup> This overexpression contributes to a high growth fraction and a doubling time of 24–48 h, making it the fastest growing human tumor.<sup>[10]</sup>

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The three distinct subtypes of BL currently recognized include endemic, sporadic, and immunodeficiency-associated BL. Endemic BL is highly prevalent in malaria-endemic regions such as equatorial Africa, with ~3–6 cases per 100,000 children per year.<sup>[23]</sup> Nearly all cases of endemic BL are associated with the Epstein-Barr virus (EBV), but the mechanism of oncogenicity is not fully understood.<sup>[19,45]</sup>

BL occurs at a 10-fold lower incidence outside of malaria-endemic regions.<sup>[19]</sup> Sporadic BL is predominantly found in North America, Western Europe, and East Asia, accounting for 30–40% of non-Hodgkin lymphoma (NHL) in these regions.<sup>[41]</sup> It is less commonly (10–20%) associated with the EBV infection and has no specific demographic association. Sporadic BL is much more common in younger patients, with a peak incidence in pediatric patients at age 11 and at age 30 for adults.<sup>[23]</sup> One review reports a 3.5:1 M: F sex predilection.<sup>[41]</sup> Although BL represents less than 5% of adult lymphomas, it makes up 40% of childhood NHLs.<sup>[4]</sup>

Immunodeficiency-associated BL is prevalent among individuals with HIV as opposed to other causes of immunodeficiency.<sup>[23]</sup> BL accounts for 40% of lymphomas associated with HIV, with a 10–20% lifetime incidence in HIV patients.<sup>[2]</sup> Unlike other NHLs associated with BL such as diffuse large B-cell lymphoma, the incidence of immunodeficiency-associated BL has not decreased with the introduction of antiretroviral therapy (ART).

The presentation and primary tumor sites of BL vary between the demographic subtypes. Unlike the endemic form of the disease, which most commonly arises in the jaw, the sporadic form arises from the abdomen in 60–80% of cases.<sup>[31]</sup> The second most common primary site of sporadic BL is the head and neck. The most common presentations of BL, therefore, are due to abdominal mass effect, including abdominal pain, nausea, vomiting, and gastrointestinal bleeding. Further symptomatology may reveal disseminated disease, including bone pain and anemia, suggestive of bone marrow involvement seen in 30% of sporadic BL at the time of diagnosis.<sup>[35]</sup> CNS involvement is present in 15% of patients at the time of diagnosis, although this includes tumors that have disseminated to the CNS from another primary site, which is common in sporadic BL.<sup>[35]</sup> Predilection for dissemination to the CNS is much more pronounced in the endemic and immunodeficiency-associated forms and is more often found at the time of diagnosis than in sporadic BL.<sup>[29]</sup> Regardless of form, however, most (~70%) of patients have advanced disease (Stage III or IV) at the time of diagnosis due to the rapid doubling time of BL.<sup>[24]</sup>

Pediatric epidural tumors causing spinal cord compression are rare and require emergent management. About 70% of childhood tumors causing spinal cord compression are extradural; lymphomas make up a minute percentage of the differential.<sup>[27]</sup> Although secondary involvement of the

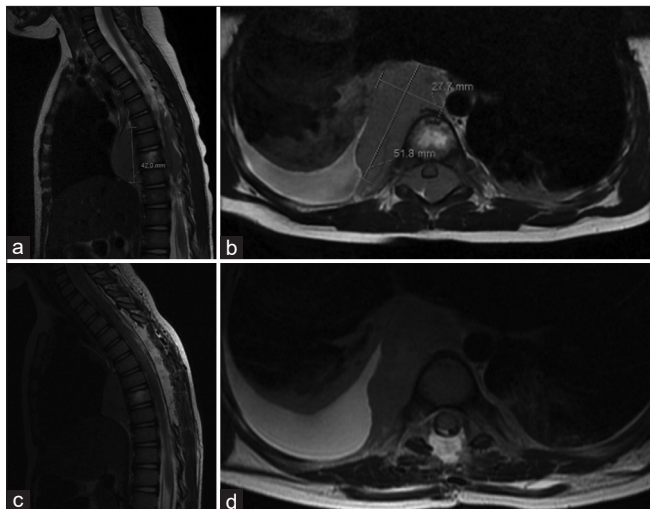
CNS is common in NHL, the spinal epidural space has been estimated to be the primary site for about 1% of NHLs in various case series.<sup>[27,36,38]</sup> This primary site is even rarer in the pediatric population.<sup>[16,36,38]</sup> Because most reviews report NHL as a single entity, the incidence of primary epidural sporadic BL in the pediatric population is unclear but is likely exceedingly rare. The youngest previously described case is in a 5-year-old boy.<sup>[30]</sup> This paper will report the case of a previously healthy 3-year-old boy who presented with spinal cord compression by a sporadic epidural BL.

## CASE PRESENTATION

A 3-year-old Caucasian male presented to an outside hospital with 2 weeks of progressively worsening trouble walking. He was initially prescribed azithromycin and prednisone at an outside clinic for a presumed infectious etiology; however, his gait instability continued to worsen. Two weeks before presentation he had an unwitnessed fall down a flight of stairs without acute injury. Pelvic X-rays showed no irregularities and CBC and blood chemistry were normal. Progression to complete inability to walk as well as areflexia in the lower extremities and new-onset urinary incontinence prompted further workup, for which the patient was then transferred to University of South Dakota Sanford Sioux Falls Hospital.

On arrival, the patient was noted to have significant weakness in his lower extremities bilaterally and was uncooperative with physical examination. Strength was 0 out of 5 in the left and 1 out of 5 in the right lower extremity. Deep tendon reflexes and sensation to light touch were absent bilaterally. The patient had no pertinent medical, family, or social history. He did not have any history suggestive of immunocompromised. Further workup included a normal CBC and CRP. An MRI of the brain, as well as the cervical, thoracic, and lumbar spine were completed under anesthesia and revealed a large soft tissue mass extending in the epidural space from T5 to T10 [Figure 1]. The mass measured  $2.8 \times 5.2 \times 4.2$  cm and involved the body of T8. Epidural involvement of the soft tissue mass resulted in visible severe thecal sac stenosis from T7-T9. Cervical lymph node enlargement could also be seen at the level of C2. CT chest/abdomen was ordered at this time and showed the mass to be uniform in attenuation with mild diffuse enhancement. No calcifications were noted. These characteristics suggested malignancy; lymphoma was considered prominent in the differential diagnosis, although the location was uncommon.

Due to the rapidly progressive myelopathic course, the decision was made to perform an emergent decompression. The extension of the tumor to the anterior surface of the vertebral bodies made complete resection improbable, so we elected to remove only the posterior portions of the tumor to achieve acute symptomatic relief and obtain a histologic



**Figure 1:** Sagittal (a) and axial (b) T2-weighted MRI preoperative images show an extensive soft-tissue mass in the epidural space from T5 to T10 with involvement of the T8 vertebral body with extension anteriorly into the posterior mediastinum. Spinal cord compression can be seen in the axial view. Postoperative T2-weighted MRI (c and d) shows removal of laminae, debulking of the dorsal portion of the tumor, as well as resolution of the spinal cord compression.

diagnosis, with a plan to address the remaining tumor after diagnosis was obtained. Complete T5-T9 laminectomies and a partial T10 laminectomy were carried out and the mass visualized as fatty-appearing homogenous tissue. The mass was also noted to protrude dorsally after removal of the bone tissue, suggesting compression before the procedure. Using microscopy, the lesion was subsequently dissected and removed from the dorsal aspect of the spinal cord dura and sent to pathology. Decompression of the cord was achieved. As planned, the lesion was not completely removed, as it occupied the anterior aspect of the spine as well as parts of the lateral canal. The procedure was well tolerated and without complication.

Histopathology revealed a prominent “starry sky” pattern, the classic description for marked apoptosis with large amounts of tingible body macrophages. High mitotic activity and foci of necrosis were present. Flow cytometry revealed the cells to express CD45, CD20, CD10, CD19, and CD38. CD34 and TdT were not expressed. The cells expressed high levels of *c-MYC* (80–90%). These findings indicated a high-grade B-cell lymphoma, strongly suggestive of Burkitt lymphoma (BL). Paraffin block confirmed *c-MYC/IGH* fusion through fluorescence in situ hybridization panel, further confirming the diagnosis.

Initial chemotherapy included intrathecal methotrexate and hydrocortisone. He is currently undergoing a regimen according to ANHL1131 with COP-R reduction phase including cyclophosphamide, vincristine, prednisolone, and rituximab. His course has been complicated by bacteremia

and mucositis with perirectal ulceration, which delayed his chemotherapeutic schedule temporarily and required diverting colostomy; however, his clinical picture is overall improving. He has regained walking ability, lower extremity deep tendon reflexes, and sensation to light touch. He has continued to have some postoperative neuropathy of the lower extremities that is under control with gabapentin. Positron emission tomography 3 months post-op showed regression of the tumor compatible with response to chemotherapy. Repeat imaging 9 months post-op showed no evidence of residual tumor or metastasis. The patient is now 10 months post-op and has returned home after intensive inpatient physical therapy and social interventions.

## DISCUSSION

Although CNS involvement is relatively common in BL at the time of diagnosis, compressive myelopathy as the presenting symptom is rare observed in less than 3% of patients with NHL.<sup>[16]</sup> Further, only some of these cases are due to primary CNS lymphoma and may be considered separately as primary spinal epidural lymphoma as opposed to systemic lymphoma with involvement of the spine. Primary epidural BL is also associated with higher mortality than systemic disease.<sup>[13]</sup> To the best of our knowledge, the youngest previously described patient with epidural spinal BL was a 5-year-old boy. Our case of a 3-year-old boy appears to be the youngest yet described and provides further data about presentation, course, and management of pediatric spinal lymphomas.

BL is generally divided into three groups based on patient demographics: endemic (African), immunodeficiency-associated, and sporadic. One retrospective study found that BL accounted for 30% of pediatric lymphomas and 40% of NHL in the United States with an incidence of three cases per million persons per year.<sup>[42]</sup> The African subtype is considered endemic as the incidence is 50 times greater in equatorial Africa than in the United States<sup>[5]</sup> and is almost always associated with EBV infection.<sup>[33]</sup> It was previously thought that the immunodeficiency-type had decreased in prevalence since the advent of highly active ART for HIV,<sup>[14]</sup> but newer studies have shown that lifetime prevalence of BL is not correlated with CD4 T cell counts.<sup>[2]</sup> The sporadic subtype is most commonly found in Eastern Europe, North America, and Asia and is less commonly (10–20% of cases) associated with EBV. Among Americans, Caucasians have a higher incidence of sporadic BL than those of African or Asian descent. Interestingly, sporadic BL has the most pronounced male predilection of all B-cell neoplasms, regardless of race.<sup>[34]</sup> Peak incidence for pediatric sporadic BL is at 11 years old.<sup>[33]</sup> The demographics of our patients fit those for sporadic BL in the United States, although he is much younger than the average age at diagnosis. To the best

of our knowledge, this is also the youngest patient reported to present with sporadic BL causing acute myelopathy.

The presentation and symptomatology of BL are highly dependent on the epidemiologic subtype. Pediatric sporadic BL most commonly presents in the abdomen (60–80%), followed by the head and neck.<sup>[41]</sup> Involvement of the CNS is present in 15% disseminated BL, but primary involvement of the epidural space is exceedingly rare, especially in children. In sporadic BL at large, the most common presentation involves mass effect within the abdomen, such as abdominal pain, palpable abdominal mass, constipation/obstruction, and hematochezia. Head and neck involvement most commonly occurs within the facial and jaw bones.<sup>[5]</sup> One review of pediatric lymphomas in a Korean center found that only 4% of patients had spinal involvement; of these, the most common presenting symptom was back pain.<sup>[16]</sup> Lower extremity weakness and myelopathy were the initial symptoms in only three of 302 patients. Due to the rapid growth capacity of BL, most patients present within days of symptom onset, with a quickly progressive course. Myelopathy and radiculopathy are often indicators for urgent surgical decompression.<sup>[25]</sup> Kurucu *et al.* report a series of 84 cases of pediatric primary paraspinal epidural lymphomas, including Hodgkin and NHL.<sup>[27]</sup> BL comprised just 33 of these cases. The presentation of our patient with frequent falls and progressive myelopathy, therefore, is very rare and not previously described in a patient this young.

The pathogenesis of BL classically involves the translocation of the *c-myc* gene on chromosome 8 to another locus that undergoes prolific transcription in mature B cells, most commonly nearby the *IgH enhancer* on chromosome 14.<sup>[21]</sup> This t(8;14) is found in 80% of BL cases regardless of epidemiological classification, although the translocation breakpoint may occur within different exons depending on the variant. Less common translocations that account for the remaining 20% of cases include t(2;8) and t(8;22), which place *c-myc* near either kappa or gamma light chain loci or enhancer elements.<sup>[5]</sup> *c-myc* protein is an important transcription factor for the regulation of cell cycle regulation, growth, and apoptosis. Increased transcription as a result of the pathologic translocation affects downstream transcription and regulation of cyclins, cyclin-dependent kinases, p53, GLUT1, collagen, and other influential proteins.<sup>[6]</sup> Further mutations in tumor suppressor genes, such as p53, have been associated with contribution to disease progression.<sup>[20,37]</sup> EBV has been linked to multiple distinct malignancies, especially in the immunocompromised HIV patient or immunosuppressed transplant recipient.<sup>[45]</sup> Analysis of endemic cases has shown that tumor lineage can be traced back to a single EBV-infected B cell, which is the case in only 10–20% of sporadic cases.<sup>[46]</sup> This suggests that EBV increases risk for BL development by promoting anti-apoptotic mutations<sup>[44]</sup> but is not required for pathogenesis.

Paravertebral tumors constitute just 4.8% of pediatric cancers.<sup>[22]</sup> Neuroblastoma and soft-tissue sarcomas make up over 72% of these. Paravertebral BL is a very rare presentation but must be considered on the differential of malignancies within the pediatric paravertebral spine.<sup>[9]</sup> Due to the acute presentation of BL, CT is a commonly used initial imaging modality and is often helpful in determining the severity and spread of the disease for staging.<sup>[15]</sup> Ultrasound may be considered initially for abdominal and pelvic masses in the pediatric population.<sup>[3]</sup> No recommendations could be found regarding imaging modalities of pediatric primary paraspinal BL due to the paucity of reported cases; MRI may often be necessary to determine the extent of CNS involvement due to its better visualization of soft tissue compared to CT. Due to the vast variety of epidemiologic and pathogenetic sources of BL, imaging characteristics are often unpredictable and dependent on anatomic location of the tumor.<sup>[43]</sup> Biko *et al.* reviewed common and uncommon radiographic appearances of pediatric BL.<sup>[3]</sup> On MRI, lymphomas in general are often hyperintense, ill-defined, single or multifocal lesions on T2-weighted and FLAIR images, and homogeneously enhanced on T1-weighted images.<sup>[1,26]</sup> In the present case, the lesion was isointense with hyperintense involvement of the vertebra on T2-weighted MRI, demonstrating the variability of BL imaging characteristics.

The classical morphology of BL is described as a “starry sky” appearance, composed of atypical lymphoid cells interspersed with tingible body macrophages ingesting the remnants of apoptotic cells due to the rapid growth of the tumor exceeding vasculogenesis.<sup>[40]</sup> Hemorrhage and coagulative necrosis are often seen. The tingible body macrophages (also referred to as histiocytes) are composed of abundant clear cytoplasm and therefore form the “stars,” while the basophilic tumor cells form the “sky” background. These tumor cells resemble normal lymphoid progenitors within germinal centers, medium-sized cells composed of basophilic cytoplasm, round nuclei, and multiple dark nucleoli.<sup>[5]</sup> Also due to the rapid growth of BL, multiple mitotic figures may be seen, and the Ki-67 index may approach 100% of cells. This unique pathognomonic appearance often makes the diagnosis of BL much clearer after biopsy can be obtained. Immunohistochemical staining of BL is most often positive for IgM, CD19, CD20, CD22, CD10, and CD79a. Negative staining for CD5, CD23, and TdT may help differentiate BL from other lymphomas and leukemias, such as B-cell acute lymphoid leukemia, which would express TdT and not surface immunoglobulin. All three epidemiologic variants of BL have been confirmed to arise from germinal center B cells, causing tumor cells to express Bcl-6 and CD10.<sup>[11,12]</sup> In the present case, the morphologic appearance along with positive B-cell protein expression without TdT expression was strongly suggestive of Burkitt’s lymphoma, which was

not the favored diagnosis prior to pathologic evaluation due to the patient demographics and presentation.

Treatment for epidural BL differs from typical recommendations due to the emergent need to decompress the spinal cord. In a patient who presents with compressive myelopathy, emergent multilevel decompressive laminotomy/laminectomy should be considered. Often, BL that either presents primarily in or metastasized to the epidural space involves the anterior portion of the spinal canal; in these cases, the goal of surgery must be to decompress the neuroaxis, with understanding that the remaining lymphoma is often sensitive to immunochemotherapy. Extraction of the posterior portions of the tumor also allows for pathologic diagnosis, which further guides treatment. Although some authors have suggested that laminotomy or laminoplasty may be preferred to laminectomy to avoid subsequent spinal instability, one review showed no difference in long-term function or deformity between epidural BL patients treated with laminectomy and those treated with laminotomy.<sup>[16]</sup> There is, however, greater risk of kyphotic deformity in patients who require laminectomy of four or more levels.<sup>[17]</sup> As such, if the spine surgeon is considering a large multilevel laminectomy, avoiding dissection and disruption of the facet joints will help prevent thoracic instability. Importantly, time from onset of symptoms to surgery and preoperative neurologic status are important prognostic indicators for long-term postoperative outcomes.<sup>[39]</sup> Patients who had lower extremity strength of 0–1 often have poor neurologic recovery after surgery, while a majority of those with strength of 2 or greater are able to walk after surgery.<sup>[16]</sup> This association is not reinforced by the current case, in which the patient motor strengths deteriorated to 1 and 0, and the patient has now been regaining walking capacity with physical therapy. Radiation therapy may be considered for decompression of these tumors, although its effects are much more delayed than surgery. Indications for radiation over surgery may include patients whose symptoms are subacute and mild and those that are not medically cleared for surgery.

Immunochemotherapeutic regimens for BL often vary between institutions, but most commonly include rituximab, a monoclonal antibody directed against CD-20, to target B-cells. Historically, the BL regimen followed that of acute lymphoblastic leukemia, meaning long, and intense cycles. This was not found to be effective in BL and does not target the unique behavior of the tumor. Because BL exhibits such rapid growth, prolonged courses of chemotherapy are likely to promote re-entrance of malignant cells into the cell cycle in between regimen cycles, causing drug resistance. For this reason, regimens were developed specifically for BL that had shortened durations of about 48–72 h; these were found to be more efficacious.<sup>[5]</sup> Recent advances in chemotherapeutic agents with improved efficacy and reduced toxicity have greatly improved outcomes in BL patients; one commonly used regimen is COPAD, which includes cyclophosphamide,

vincristine, prednisolone, and doxorubicin. A longitudinal study of the COPAD regimen on pediatric patients with BL found the 4-year event-free survival to be over 98% and the overall survival to be over 99%.<sup>[18]</sup> This regimen without doxorubicin and with rituximab is used in the current case. Importantly, rituximab has been shown to be a beneficial addition to any of the described regimens.<sup>[32]</sup> Pediatric NHL patients with CNS involvement at the time of diagnosis, whether primary or metastatic, have been shown to have poorer prognosis, often requiring further CNS prophylaxis in the form of intrathecal chemotherapy including methotrexate and/or cytarabine.<sup>[5,8]</sup> There is no evidence that additional prophylactic CNS radiation improves outcomes compared to intravenous and intrathecal chemotherapy alone. The length of treatment is often determined by the initial stage as well as the individual patient response to the first courses of chemotherapy.

## CONCLUSION

BL is a common childhood malignancy. Sporadic BL occurs in immunocompetent hosts from non-endemic areas, and most commonly arises within the abdomen or pelvis. BL arising primarily in the epidural space and presenting as acute compressive myelopathy in a pediatric patient is exceedingly rare. To the best of our knowledge, this is the youngest described patient with sporadic primary paraspinal BL. A paraspinal mass with epidural extension causing myelopathy is a surgical emergency. Chemotherapeutic regimens vary between institutions but should include neurotropic agents. Initial grade and neurologic status are important prognostic indicators.

## Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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## Conflicts of interest

There are no conflicts of interest.

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