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Spinal Cord Compression Revealing a Case of Metastasis of Burkitt Lymphoma

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Abstract **Case Report**

Burkitt lymphoma is the most frequent non-Hodgkin lymphoma in children and adolescents. The disease can involve all organs, although spinal cord involvement in the Burkitt lymphoma is exceptional and only a few cases are reported. We report a case of Burkitt lymphoma revealing by spinal cord compression in a 4-year-old patient. Magnetic resonance imaging (MRI) of the spine revealed a mass in the spinal intradural extramedullary space with severe spinal canal stenosis in T6-T9. Neuromeningeal involvement in Burkitt's lymphoma often represents a secondary location and defines an advanced stage of the disease. The child responded well to chemotherapy and is on remission.

Keywords: Burkitt lymphoma, Spinal cord compression, MRI.

INTRODUCTION

Burkitt's lymphoma (BL) is a lymphoma belonging to the group of B-cell malignant non-Hodgkin's lymphomas (NHL). The BL is a very aggressive lymphoma characterized by the deregulation and translocation of the C-MYC gene from chromosome [1]. BL is the most frequent non-Hodgkin lymphoma in children and adolescents, accounting for approximately 34% of the lymphoma cases in young age [1]. Maxillofacial and abdominal locations are the most common, although the disease can involve all organs, and studies report the rarity of neuro-meningeal locations including spinal cord involvement [2-5].

OBSERVATION

Our observation concerns a 4 year old child with no previous pathological history. The patient was admitted with flaccid paraplegia. The onset of symptoms was two months prior to his admission with the appearance of a cervical swelling. In addition, the child had presented three weeks before his admission with pain of the intense paresthesia type, of progressive onset, dorsolumbar, radiating to the buttocks, the posterior face of the thighs and legs, with progressive onset of functional impotence of both lower limbs evolving towards paraplegia. On admission, the neurological examination revealed a conscious patient with paraplegia and sharp osteotendinous reflexes. We also noted the presence of bilateral jugulo-carotid and sub-angulo-mandibular adenopathies, the largest measuring 2x1,5 cm, of solid consistency and mobile in relation to the deep and superficial planes responsible for respiratory distress. Magnetic resonance imaging (MRI) of the spinal cord showed an intradural and extramedullary lesional process developed posteriorly (T6-T9) and anteriorly (T12-L1) to the medullary cone, creating an extensive heterogeneous tissue masses. This process was T1-isointense and intermediate hypersignal T2 and STIR, and moderately enhanced after injection of Gadolinium (Fig. 1), it also revealed bilateral

nephromegaly with nodular infiltration enhanced after injection of contrast media. The brain MRI was without abnormalities. Lumbar puncture showed lymphocytic meningitis, culture was negative. Biopsy of the cervical adenopathy followed by pathological examination revealed squamous mucosa infiltrated by lymphomatous proliferation. As part of the lymphoma work-up, the blood count was normal. The thoracic-abdominal-pelvic CT scan had shown an hepato-splenomegaly associated with a magma of deep abdominal, inguinal and cervical adenopathies and pulmonary nodules (Fig. 2). The bone marrow biopsy (BOM) showed bone marrow hypoplasia without malignancy. The pre-treatment work-up was unremarkable. Multidrug therapy with cyclophosphamide, hydroxyadriamycin, vincristine and prednisone (CHOP) was started. The patient had received a total of 6 courses of CHOP. The clinical course after 8 months was favourable, marked by regression of adenopathy and recovery of functional impotence.

The above case describes a spinal cord compression syndrome with paraplegia in a 4-year-old child. The diagnosis of Burkitt's lymphoma stage IV was made.

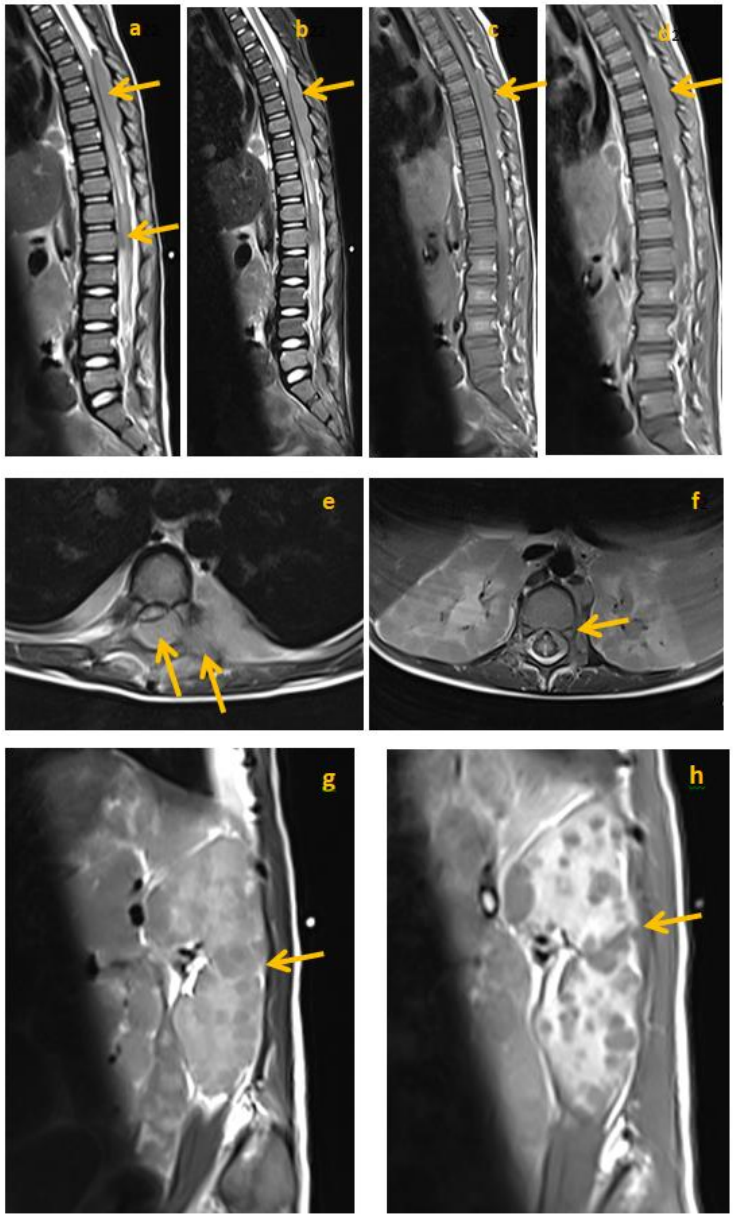

Figure 1: MRI of the thoracic and lumbar spine.

MRI T2 (a), STIR(b), T1(c) and T1 with Gadolinium (d) sagittal view of the spine revealing significant cord compression at T 6-9 due to dorsal epidural mass and T12-L1 ventral epidural mass heterogeneous measuring 10 cm. (e,f) MRI T2 axial view of the lumbar spine revealing a mass epidural measuring approximately 30mm with neural foraminal narrowing and direct extension to the left paraspinal musculature.

NB: it also revealed bilateral nephromegaly with nodular infiltration enhanced after injection of contrast media (g: T1; h: T1Gado)

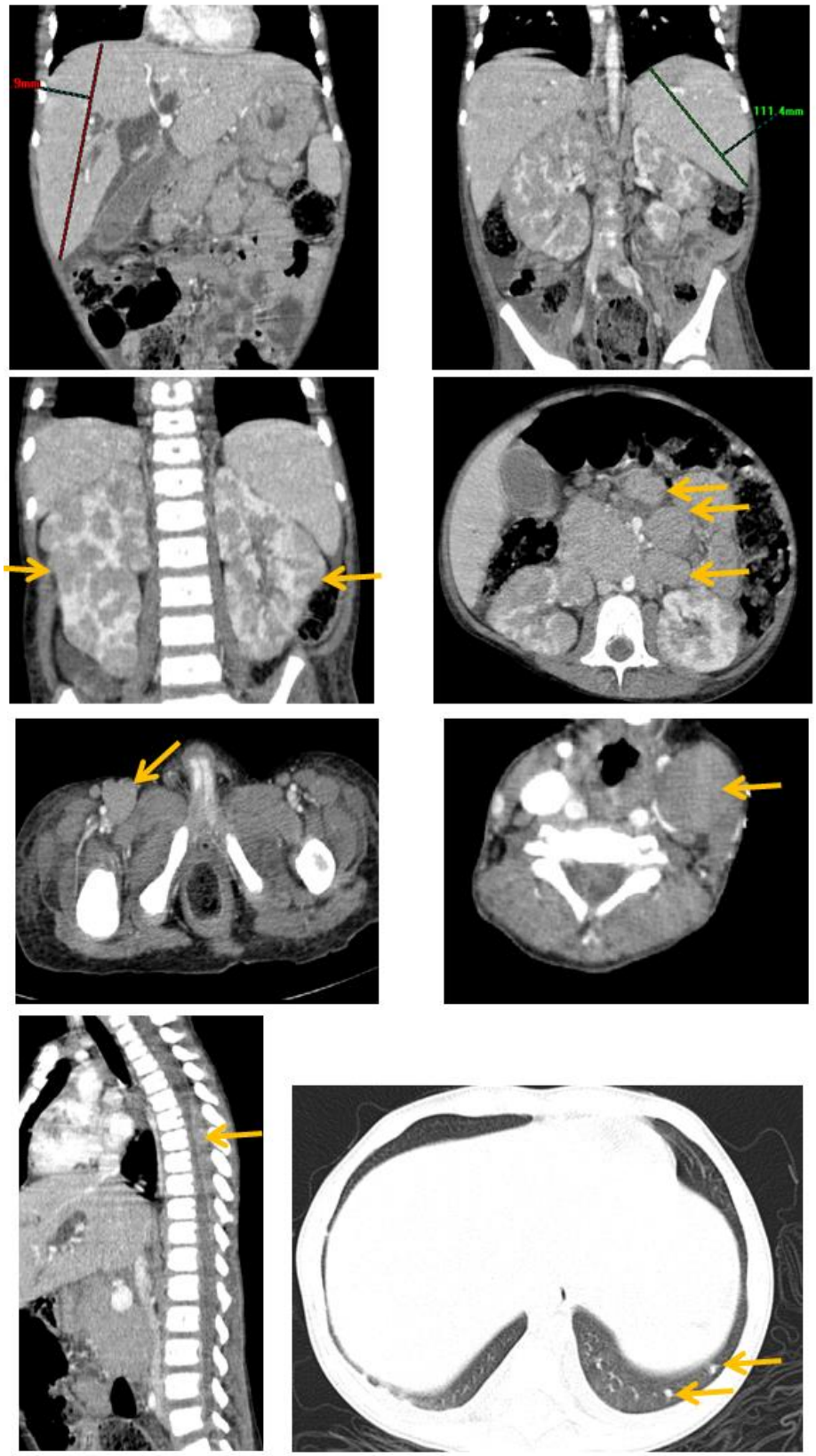

Figure 2: CT scan of the chest, abdomen, and pelvis

CT scan had shown an hepato-splenomegaly (a,b), bilateral nephromegaly (c) associated with a magma of deep abdominal(d), inguinal (e) and cervical adenopathies (f) and pulmonary nodules (h) is significant for metastatic disease. **NB**: intradural and extramedullary lesional process (g)

DISCUSSION

Burkitt's lymphoma (BL) is a non-Hodgkin's lymphoma of high malignancy grade. BL is common among the African children where the estimated incidence is 3 to 6 cases per 100,000 children/year [1]. Burkitt's lymphoma can be divided into three forms: endemic, sporadic and immunocompromised [6]. The endemic form affects mainly the facial bones, the sporadic form affects mainly the terminal ileum, the cecum and the intra-abdominal lymph nodes, and the form associated to immunodeficiency [1]. In young age, BL can appear as an abdominal mass associated to symptoms such as: gastrointestinal hemorrhage, abdominal pain, nausea, and bowel obstruction caused by the direct compression or the involvement of the bowel lumen [1]. Intussusception upon presentation can be present in up to 18% of the patients with primary abdominal BL [1]. Neuromeningeal involvement in Burkitt's lymphoma often represents a secondary location and defines an advanced stage of the disease [3,7,8,9]. Clinical presentation of spinal lymphoma typically includes weakness, numbness, and progressive difficulty in ambulation [10]. Cases involving spinal cord compression have been reported mainly in children, including the 7 cases reported by Ses et al, where the average age was 15 years [9]. Age, performance status, LDH value, bone marrow, and central nervous system involvement are the most frequently used prognostic factors [1].

The thoracic spine is the most commonly affected site, followed by the cervical, less commonly the lumbar region, and most of them are solitary [10]. Several hypotheses have been put forward. For some authors, it was a transformation of lymphomatosis of a pre-existing lymphoid tissue in the epidural space. For others, epidural masses are the result of tumour extension from bony and paravertebral locations [3]. In our case, it is indeed a Burkitt-type NHL, stage IV with epidural extension and compression of the spinal cone without vertebral involvement. Kidneys are affected in aggressive forms of non-Hodgkin lymphoma. Their infiltration is often present in terminal phases of non-Hodgkin lymphoma. In children, BL has the highest frequency of kidney infiltration. Hematogenic dissemination is the most common cause for renal lymphoma, though infiltration from adjacent ganglions can be present in 10% to 20% of the cases [1].

Magnetic resonance imaging (MRI) with contrast is the preferred modality for the evaluation of spinal cord and spinal canal tumors [10]. On MR, lymphoma appears as single or multifocal, ill-defined T2W/FLAIR hyperintense lesions with homogenous contrast enhancement on T1W images [10]. Because of high cell tumor density, diffusion-weighted MRI often demonstrates restriction and correspondingly hyperintensity.[10] In the present case, the lesion was multilevel and intermediate signal on T2 and mildly enhancing post gadolinium. There was no upper motor neuron lesion clinically and brain imaging was normal [10]. Currently, CT is widely used as an initial imaging method for staging BL. The chest and abdominal CT native and with contrast agent highlight the existence of the abdominal, mediastinal adenopathies, and pleural, pericardial, renal, muscle and peritoneal metastases. Upon admission, most children with BL come in an advanced stage of the disease and/or with metastases. The exact determination of the stage is essential for the appropriate treatment [1].

Decompressive laminectomy can be used to treat spinal cord compression in Burkitt's lymphoma. For Dechambenoit [3], surgery is only justified for diagnostic purposes. The combination of radiotherapy and chemotherapy has been shown to improve the overall survival of patients [2, 4].

CONCLUSION

Spinal medullary Burkitt’s lymphoma is a rare diagnosis that may often be difficult to differentiate radiologically from other causes of intradural extramedullary lesions. A thorough histological examination is warranted in such cases.

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Author contributions

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Disclosure

The authors declare that they have no competing interests.

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