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Case Report

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Cord compression due to atypical T-cell lymphoma from paraspinal soft tissue: Report of a case

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Abstract

Background: During lymphoma's natural history of the disease, 5-10% of cases may develop Central Nervous affectation.

Case presentation: A 57-years-old man with less than 24 hours of onset symptoms of paraparesis, lower limb hypoesthesia, and sphincter dysfunction was operated due to dorsal tumor with an epidural component which caused severe cord compression.

Conclusions: Pathological analysis concluded atypical T-cell lymphoblastic lymphoma, a rare subtype of lymphoma which accounts for 1–2% of all Non-Hodgkin Lymphomas. Our case was particularly aggressive and atypical due to its origin in paraspinal soft tissue. Despite specific treatment, the patient presented an early epidural relapse, frequent in this lymphoma subtype.

Keywords: Lymphoma; Surgery; T-cell.

Abbreviations: ALL: Acute Lymphoblastic Leukemia; CNS: Central Nervous System; CT: Computerized Tomography; LBL: Lymphoblastic lymphoma; NHL: Non-Hodgkin lymphoma.

Background

Lymphomas are malign tumors of B, T, or NK cells that generally originate in the lymph nodes, which classically are divided into Hodgkin and Non-Hodgkin types. Particularly, T-cell lymphomas may be classified depending on the grade of cell maturation. Peripheral T-cell lymphomas are neoplasms derived from post-thymic lymphoid T cells at different steps of differentiation, whereas Lymphoblastic Lymphomas (LBL) derive from precursor thymic T-cells.

At least 5-10% of patients may develop Central Nervous System (CNS) affectation during the disease, which is commonly seen in late stages, when dissemination has appeared [1].

We present the case of an atypical aggressive T-LBL which arose from dorsal paravertebral soft tissue with epidural component causing severe cord compression.

Case presentation

A 57-years-old male patient with antecedents of recent kidney stone nephrostomy, severe drug abuse, and hepatitis C virus was transferred to our hospital due to suspicion of spinal cord compression of less than 24 hours of instauration. Onset symptoms were lower limbs hypoesthesia, urine difficulty, and progressive gait disturbance being impossible previous transfer. At the Emergency Department of our center, we confirmed lower extremities paresis, being 4/5 proximal bilateral and 4+/5

distal bilateral, severe D4-D5 sensory level and 3+/4 lower limbs reflexes. Gait was only possible with help of two people.

Urgent spinal Magnetic Resonance Imaging (MRI) was performed, revealing a D3-D5 paraspinal soft tissue mass which was enhanced with contrast. That mass infiltrated posterior bone elements, thorax cavity, and epidural space, causing severe cord compression (Figure 1). Thorax X-Ray and recent abdominal Computerized Tomography (TC) did not show other lesions.

Due to acute neurological deterioration, emergent surgery was considered. D4 laminectomy and D3 and D5 hemilaminectomy were performed. Paravertebral musculature tumor mass was seen; being friable and high vascularized. Lamina infiltration was remarkable and contiguous extension to facet joins encountered. The epidural invasion was slightly adhered to dura mater which was successfully resected allowing optimal thecal sac decompression (Figure 2). Intraoperative tumor samples were sent to further pathological study. Postoperative neurological improvement was achieved with early rehabilitation, being able to walk with one crutch during the first week.

No lymph nodes affectation or other tumors were observed in thorax and abdomen CT. Considering blood malignancies as the most plausible origin, bone marrow aspiration was scheduled, which concluded blastosis of 25%. There were elements of heterogeneous size: some of small size and more mature aspect and others of medium size, and lax chromatin, with visible nucleolus. The nuclei were irregular, with a high nucleus-cytoplasm ratio, some with fine granulation. The definitive pathological diagnosis was T-cell Lymphoblastic Lymphoma.

According to PETHEMA LAL 2019 (group of Lymphoblastic Acute Leukemia of the Spanish Program of Treatments in Hematology), he started corticoid preparation, firstly well-tolerated, but newly presenting at fourth-week neurologic deterioration with abrupt paraplegia. Early epidural recurrence was confirmed on MRI and surgery was ruled out. Radiotherapy was accepted and the patient received 30Gy radiation previous to induction treatment with Daunorubicin and Vincristine. During induction treatment, the patient presented different complications such as progressive cholestasis, persistent fever syndrome due to bacteremia and urine infection, mucositis, and refractory agitation. On day 23 of induction treatment, the Eastern Cooperative Eastern Group (ECOG) scale worsened to level 4, so it was dismissed from other curative therapeutic options such as Allogeneic Hematopoietic Stem Cell Transplantation and Nelarabine, and symptomatic treatment was established until death.

Pathological findings

The tumor samples showed diffuse atypical lymphoid infiltrate, consisting of small-medium lymphocytes, with irregular nuclei, chromatin ranging from condensed to more dispersed, with many nucleolus occasionally evident, and sparse cytoplasm. Abundant figures of mitosis and bodies of apoptosis were observed. The infiltration was accompanied by abundant dispersed epithelioid granulomas, of variable size, non-necrotizing, and not encapsulated (Figure 3a-c). A moderate number of eosinophils, abundant macrophages, and an endothelial network were also evident. The immunohistochemical study revealed that this atypical lymphoid infiltrate was positive for CD45, CD2, CD3, CD5, CD7, CD4, PD1 (faint and diffuse), TdT (intense and heterogeneous) (Figure 3d), CD99 (intense and diffuse) (Figure 3d-f), CD1a (heterogeneous and partial) and BCL6 (diffuse and heterogeneous). It was negative for B markers (CD20, CD79a and PAX5) (Figure 3g), CD8 (expressed in occasional small T lymphocytes), CD138, BCL2, CD10, CD30, Cyclin D1, MUM1, EBER (technique in situ hybridization EBV), CD21, CD23, CD15, ALK-1, CD68, CD123, CD163, S100, CD34, CD33, C-KIT, Myeloperoxidase, CD56, PGFA, CK AE1 / AE3 and CK Cam 5.2. The cell proliferation index (Ki67) was very high raised by approximately 75%. The molecular study presented clonal rearrangements for TCR-beta.

Conclusions

LBL is an infrequent and aggressive malignancy which represents 1-2% of all Non-Hodgkin Lymphomas (NHL). T-cell LBL (T-LBL) accounts for 90% of all LBL, predominantly develops in adolescents and young adults, and is mostlyseen in males [2,3]. Typically, T-LBL presents as advanced disseminated disease, usually revealing a mediastinum bulky mass, often with pleural and pericardial effusion, and with bone marrow not or partially involved (<25% infiltrating blast cells) [1]. Bone marrow involvement is what differs from Acute Lymphoblastic Leukemia (ALL), but due to morphological and immune-phenotypical resemblances, LBL is listed together with ALL [3].

NHL may compromise either the peripheral nervous system or the CNS, being spinal cord compression, leptomeningeal spread, or intracerebral mass lesions the way of presentation. In addition, the effects of chemotherapy, paraneoplastic syndromes, or opportunistic infections may be considered as other mechanisms of neuropathy in patients with lymphoma [4].

Leptomeningeal spread is the most common manner of metastatic dissemination of the nervous system.Hematogenous spread to brain parenchyma is less frequent, but lymphoma may develop nodular drops by infiltration from the subarachnoid space. Likewise, comparable production of tumor nodules may result in focal neurologic symptoms along the central neuraxis, including the brainstem and spinal cord [4].

The risk of CNS involvement or spinal cord compression is dependent on the lymphoma subtype (may be seen in both Hodgkin and Non-Hodgkin lymphomas) as well as other factors. High-grade lymphomas, such as Burkitt lymphoma or LBL, tend to have more CNS involvement at relapse than other histologic subtypes [5]. Other factors like cytogenetic abnormalities may alter the expression of T-cell oncogenes which influences the clinical course as well. The majority of these abnormalities in T-LBL are present in the 14q11-13 region, the site of the TCR-alpha and TCR-delta genes. Rearrangements of the regions encoding TCR-beta (7q34) and TCR-gamma (7p14.1) are also frequently seen [6,7].

Spinal cord compression is an infrequent presentation of NHL, appearing only in 0.1% to 3.3% of patients of primary extradural NHL. Primary intramedullary and intradural extra medullary are extremely rare. Otherwise, spinal cord compression may occur in 0.1% to 10.2% during the course of NHL disease and it is normally destructive in behavior [8]. It is usually caused by extradural disease, being a solid drop along the spinal canal, extension from an adjacent nodal mass, or bone involvement

in the patterns of compression. The affectation of lymph nodes is the most common pathogenesis, extending into the spinal epidural space through the neural foramina. If NHL has spinal epidural involvement, it usually means an aggressive disease.

Treatment is peculiarized by an elevated response rate to immediate chemotherapy; although there is usually an early recurrence, being CNS a frequent relapse location [2]. At present, treatment options imply intensive induction therapy, prevention of CNS relapse, and post-induction therapy purposing the reduction of subsequent recurrence. Other regimens include mediastinum radiation therapy in cases of high tumor burden at this site [6].

The difference in survival between cord compression resulting from lymphoma or other extradural malignancies is notorious. The global mean survival of patients with all types of extradural is 8-9 months, with less than 10% surviving more than 1 year [9]. Even with survivals rates of 50% at more than 3 years in some series, extradural lymphoma although localized, may be an aggressive disease [10].

We present the unusual case of an adult male patient with T-cell Lymphoblastic Lymphoma with no mediastinum mass and peripheral disease, whose dorsal paravertebral soft tissue was the primary location. Despite having an optimal neurological rehabilitation after surgical decompression, he presented an extremely poor response to immediate induction treatment and an early CNS relapse appeared with the consequent irreversible motor impairment.

Spinal cord compression is an emergency, which needs immediate measures to avoid neurological sequelae. Prompt diagnostic is required, being MRI the gold standard study. In the differential diagnosis of an epidural mass which compromises the spinal canal, it is important to rule out hematological tumors for their overall better treatment response compared with other malignancies. To ensure the correct subsequent therapy, large biopsy samples are often necessary for cellular morphology, molecular, immunophenotype, and karyotype analysis. Total-body computed tomography scan, bone marrow aspirate, and increasingly positron emission tomography are tools necessary to complete the diagnosis. In cases of T-LBL, special emphasis must be taken with CNS early relapses.

Declarations

Ethics approval and consent to participate: Not applicable.

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