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Medular Compression Syndrome as a Presentation of a Spinal Epidural Lynphoma.

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Abstract

Introduction*: Lymphomas are solid tumors of the immune system. Spinal cord compression by this tumor is a late manifestation and not always present in all of patients. Sometimes, lymphomas can be present only in the spinal epidural space but this is a very rare condition.*

Case Report*: This is a 23 years old patient who 6 months ago started with dorsal pain after a sudden movement. With time, the pain got worse in intensity and frequency. Four months after his first manifestation noted a progressive loss of weight and weakness in both lower limbs.*

Discussion*: In the CT and MRI studies, we found images suggestive of lymphadenopathy in the mediastinum and abdominal cavity, as well as in the dorsal spine an intra-spinal extradural image that enhancement with contrast. Thinking in a paraparesis caused by a extradural lymphoma, the patient was taken to the operating room and a three-space laminectomy was performed with a gross total resection of the tumor.*

Histopathological examination revealed a Hodkin lynphoma and 5 days after surgery the patient presented improvement in the muscular strength of the lower limbs. He was transferred to the oncology service to begin with adjuvant chemotherapy and radiotherapy treatment.

***Conclusion**: Despite the low incidence of lymphomas with spinal epidural presentation, it is a diagnosis to bear in mind when differentiating intra-spinal extradural tumors. Surgery in patients with signs of medullary compression that are clinically expressed with progressive neurological deterioration should be performed as soon as possible to achieve an adequate improvement of the neurological picture*

***Kye words:** Epidural lymphoma, Primary spinal epidural lymphoma, spastic paraparesis, laminectomy, Hodgkin lymphoma, non-Hodgkin lymphoma.*

Introduction

Lymphomas are solid tumors of the immune system. Hodgkin's lymphoma accounts for about 10% of all lymphomas, and the remaining 90% are referred to as non-Hodgkin lymphoma. **(**1**)** Hodkins lymphomas has a bimodal distribution, affecting young patients around 20 years old and then after the 55 years, it´s incidence is increased again. Most of these patients present with supradiaphragmatic lymphadenopathy and one third of this has high fevers, night sweat and weight loss. Commonly, this disease involves regionals lymph nodes but can involve extranidal sites by direct or hematogenous innovation. Common sites that may be involved include the spleen, liver, lungs and bone. **(**2**)**

By the other hand, spinal cord compression is a late manifestation and not always present in all of patients with lymphomas. **(**3**)** Sometimes, lymphomas can be present only in the spinal epidural space but this is a very rare condition. About 10% of all of spinal epidural tumors are represented by primary spinal epidural lymphomas and those represented 0.1-3.3% of all of them. **(**4**)** Primary spinal epidural lymphoma **(**PSEL**)** is defined when the are no recognizable sites of disease at the time of diagnosis. The incidence of this subset of lymphomas is much less than others. The first Autor reported a Hodkin lymphoma in the epidural spinal space was Love in 1954 in seven patients, after that, only a few patients had been reported in literature **(**5**)**

We present a patient with progressive loss of strength in both lower limbs because a spine epidural lymphoma.

Case Report:

This is a 23 years old patient who 6 months ago started with dorsal pain after a sudden movement. With time, the pain got worse in intensity and frequency, however, it relived with analgesic drugs and for that reason he did not find medical advice. Four months after his first manifestation, the patients get a covid-19 infection and noted a progressive loss of weight. After that, he started to feel weakness in both lower limbs. On the next two weeks the patient let to walk and for that reason he came to our neurosurgery consultation.

Physical Exam:

Hemolymphopoietic system: no visible or palpable lymphadenopathy. No palpable spleen.

Nervous system:

Muscle strength: decreased muscle strength in both lower limbs. Barre and Mingazzini tests: positive for both lower limbs.

Sensitivity: thermal, painful and tactile superficial sensitivity: decreased in both lower limbs to D10 level. From D6 upwards sensitivity was totally normal.

Deep sensitivity was abnormal keeping the same levels as the superficial.

ASIA: C

Laboratory Exams:

Hto: 0.36% Hb 12

RCP 36.2H Eritro 34mm/h

The rest of the blood tests were negatives

Images:

Computation Tomography Scan (CTS):

Dorsal Spine and chest CTS: Chest CTS is made keeping special attention in dorsal Spine at D3-D8 osseous level but no alteration in the vertebrae was founded, however, in the vertebral canal between D4 and D7 vertebrae there was a small density change, very diffuse. In chest there is a retrosternal isodense image in correspondence with mediastinal lymphadenopathies.

Abdominal CTS: Images in correspondence with deep lymphadenopathy's close to lumbers vertebras.

Brain CTS: Totally normal.

Magnetic Resonance Image (MRI):

Dorsal Spine MRI: Between D4 and D7 there is a hyperintense image in T2 and isointense in T1, completely extraxial in the epidural space with enhancement after gadolinium contrast. No evidence of continuity from another image from chest or vertebral bone. **(**Fig 1 and 2**)**

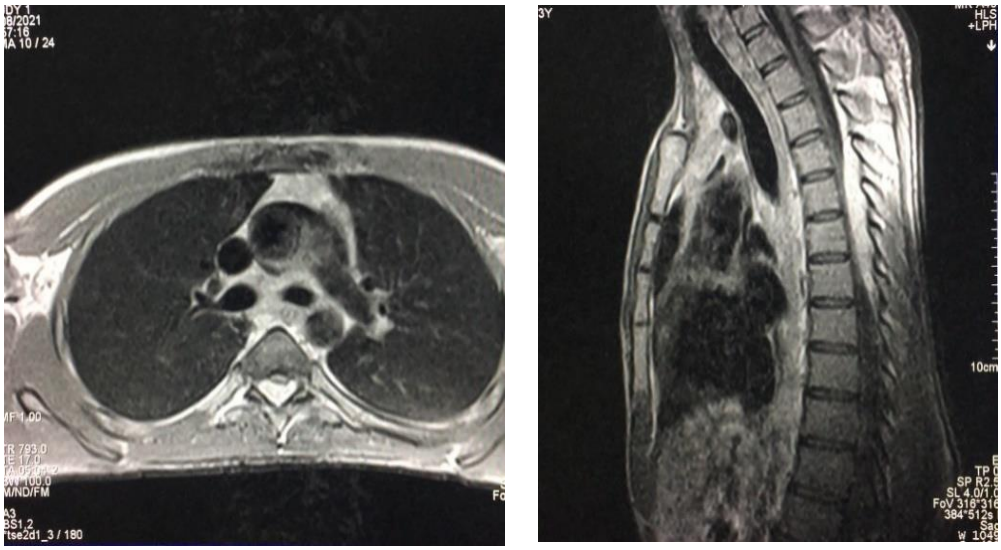

Fig 1 and 2: Dorsal Spine MRI with gadolinio enhancement. Imagenology department of Faustino Perez Hospital, Matanzas, Cuba.

Cervical and lumbar Spine MRI: Completely normal. Deep's lymphadenopathies in abdominal cavity.

Discussion

Because the clinic and complementary studies of the patient, we directed the diagnosis to a primary extradural lymphoma. However, mediastinal and abdominal lymphadenopathies mean that there is another recognizable site of disease. **(**6**)** For these reasons we cannot speak of a PSEL, it is a rare case since there are no intrathoracic masses that continue with the epidural tumor or metastases in the vertebrae. In addition, the two clinical phases proposed by Epelbaum for PSELs could be defined, characterized by the first one by back pain that can last for months to a year and a second phase that progresses in a few weeks with neurological deterioration. **(**5;7**)** The patient was taken to the operating room and a three-space laminectomy was performed, finding a well-defined tumor lesion poorly adhered

to the spinal dura, with hard and bleeding characteristics. For which, a microsurgical devascularization is performed, achieving total excision **(**Fig: 3 and 4**)**.

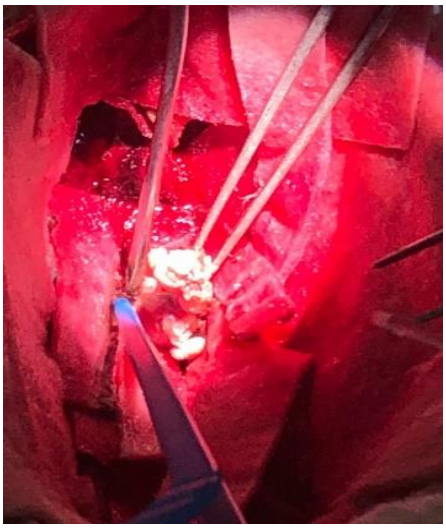

Fig 3: The tumor is being removed. Transurgical picture taken with a smartpfone camera.

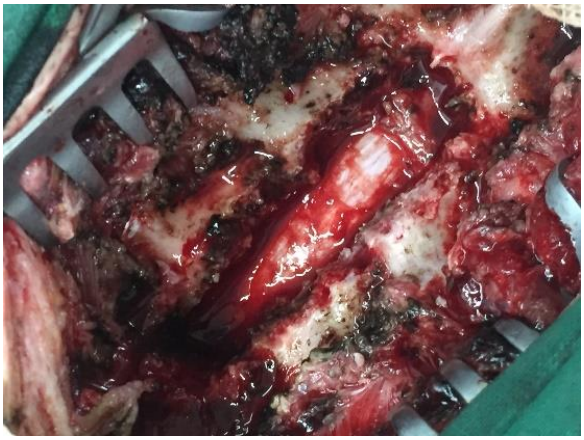

Fig 4: The tumor have been completely removed. Transurgical picture taken with a Smartphone camera.

We decided on surgery as the main treatment to avoid the progression of neurological symptoms and achieve a histological diagnosis. Despite the high sensitivity to radiotherapy and chemotherapy that these tumors have, we agree that most authors consider surgery as the main line of treatment in these patients, precisely to achieve these objectives. In a retrospective study published in Nature of 13 patients with malignant epidural lymphomas, surgery was recommended in 9 patients with signs of spinal cord compression, achieving satisfactory results in the improvement of neurological symptoms and in two

patients in whom surgical decompression was not recommended due to his advanced stage of the disease, the neurological deficit progressed rapidly. **(**8**)**

In the study published by Le Xiong where 130 patients with PSEL are analyzed, it is concluded that the best therapeutic option for these patients is surgery followed by radiotherapy and chemotherapy. Other case reports in the literature recommend surgery as the initial treatment in these patients **(**4; 5; 7; 8; 9; 10; 11**)**, which coincides with our decision. Histopathological examination revealed a Hodgkin lymphoma, classic variety, possible sclero-nodular. It is difficult to explain physiopathologically the appearance in the spinal epidural space of a Hodgkin lymphoma, since its extranodal origin is much less frequent than its nodular form, and even less when it occupies this location. **(**4;5;8**)** However, the Rubinstein's studies have suggested that there are lymphoid cells in the epidural space, which would be activated by antigenic stimulation. **(**12**)**

On the contrary, other authors are not agree with the presence of lymphoid tissue in the spinal epidural space and suggest that these lymphomas originate in the paravertebral space or retroperitoneal tissue reaching the spinal epidural space through foraminal foramina.**(**13;14**)** By the other hand, MgDonald proposes a classification system based on four groups for malignant epidural lymphomas and of this how to define the primary of the metastatic, Group 1: Primary origin in the bone; Group 2: Paravertebral tumor; Group 3: Paravertebral tumors with bone destruction and Group 4: Epidural tumors without bone destruction or paravertebral masses.**(**11;15**)**

If we are guided by this classification system, the PSELs would correspond to group 4, as did our patient. We ask, are we in the presence of a primary spinal epidural Hodgkin lymphoma? If we take into account that the patient did not present any other symptoms in any other system at the time of diagnosis and we rely on this classification system proposed by MgDonald et al. and that he is not in an advanced stage of his disease, then if we would be in the presence of a PSEL. However, the presence of deep lymphadenopathy in the mediastinum and abdominal cavity conspire against this diagnosis. Finally, we closed the case as a Hodgkin lymphoma with a clinical presentation of spinal cord compression, leaving its possible primary origin as a question, since there is only 8 patients reported with this origin. **(**9**)**

Five days after surgery the patient presented improvement in the muscular strength of the lower limbs, ASIA: D. He was transferred to the oncology service to begin with adjuvant chemotherapy treatment. At 30 days after surgery, the patient presented a complete recovery of neurological symptoms, ASIA: E.

Conclusion:

Despite the low incidence of lymphomas with spinal epidural presentation, it is a diagnosis to bear in mind when differentiating intra-spinal extradural tumors. Surgery in patients with signs of medullary compression that are clinically expressed with progressive neurological deterioration should be

performed as soon as possible to achieve an adequate improvement of the neurological picture and an accurate diagnosis by the histopathological study of the excised lesion. Patients should be treated with chemotherapy and radiation therapy after surgery to achieve favorable results in each patient.

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