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Case Report

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Surgical Resection with Boyd Technique for Metastatic Ewing Sarcoma of the Bone Plus Docetaxel/Gemcitabine, Associated with Improved Outcomes in Tumor Activity.

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Abstract

Ewing sarcoma is a malignant bone tumor that mainly affects children, adolescents and young adults with more than 1.5 cases per million worldwide. Approximately 20-25% of patients present metastatic disease at the diagnosis, that is often resistant to intensive therapy.We present the case of a 19-year-old male with history of epilepsy who started his condition with weight loss, increased volume, pain, swelling after receiving surgical treatment due to a left distal femur fracture, showing tomographic evidence of a 50-cm tumor with multiple lung lesions, so disarticulation was performed with the Boyd technique, obtaining histopathological result of Ewing's Sarcoma, and was subsequently sent to the medical oncology service for follow-up and adjuvant treatment with significant clinical and radiological improvement in pulmonary metastatic activity.

Key Words: Ewing Sarcoma, Pelvic Limb Pain, Pathological Fracture, Metastasis, Multidisciplinary Treatment.

Abbreviations

ES: Ewing's sarcoma
DOX: Doxorubicin
CPM: Cyclophosphamide
VCR: Vincristine
ACT-D: Actinomycin-D
IFO: Ifosfamide
ETO: Etoposide

Introduction

Ewing's sarcoma is a malignant bone tumor that occurs predominantly in the pelvis, femur, tibia and ribs or as a soft tissue tumor located in the chest wall, gluteal muscle, pleural cavities, and cervical muscles, with more than 1.5 cases per million worldwide. Approximately 20-25% of patients have metastases at diagnosis that are often resistant to intensive therapy [1]. Ewing's sarcoma (ES) is the second most common malignant primary bone tumor in children and adolescents, with a peak incidence at the age of 15 years; boys and men are slightly more affected than girls and women (sex ratio of 3:2) The origin of this tumor was unclear until recently, when electron microscopic and immunohistochemical analyzes suggested that it is of neurogenic origin, ES tumors often express a balanced translocation involving the EWS gene on chromosome 22 and a member of the ETS family of transcription factors [2-6]. As with other primary bone sarcomas, pain is the most common initial symptom of patients with Ewing's sarcoma of bone. As the tumor destroys bone, patients may notice a deep, dull, aching pain in the involved region or extremity, it is not uncommon for patients to present with fever or weight loss, which in the presence of bone pain may lead the physician into misdiagnosing the cause as osteomyelitis [7,8]. Histologically, Ewing's sarcoma appears as sheets of homogenous densely packed small round blue cells. They have a high nuclear to cytoplasm ratio and the nucleus is associated with fine granular chromatin and pinpoint nucleoli. The cytoplasm typically has few or small organelles and abundant glycogen [9]. In ES of the bone, plain radiographs exhibit permeative and infiltrative destruction of the affected bone. In addition, an onion skin-like appearance and spiculae are indicative of periosteal reactions [10].

Current standard of localized or regional treatment for patients with ES includes systemic chemotherapy and surgery and/or radiation therapy for local control [11]. Due to improvements in intensive chemotherapy, the prognosis of ES patients has improved markedly. The current chemotherapy protocols used to treat ES include various combinations of the following six drugs: doxorubicin (DOX), cyclophosphamide (CPM), vincristine (VCR), actinomycin-D (ACT-D), ifosfamide (IFO), and etoposide (ETO) [2]. Gemcitabine in combination with docetaxel was found to be well tolerated and demonstrated antitumor activity in children and ado-

lescents with recurrent or refractory disease [12].

Whether surgical excision of the primary bone tumor improves the survival of patients who present with metastatic ES is unknown, there are no large series covering this issue. Some authors advise amputation as the "gold standard" treatment [13,14]. Therefore, local surgery is an appropriate treatment for patients with metastatic ES. Raciborska et al. reported that treatment of isolated lung metastases may have a role in improving prognosis in patients with ES. Letourneau et al [15,16].

Materials and Methods

We present the case of a 19-year-old male patient, with a history of epilepsy, who suffers a diaphyseal fracture of his left distal femur, an open reduction and internal fixation were performed using an anatomical plate for the distal femur. The patient comes to the consultation because after the surgical intervention, the pain becomes incapacitating and presents a progressive increase in volume. Upon physical examination (Figure 1), a collateral venous network is observed, he presents hyperthermia in the thigh and a palpable mass of approximately 50 cm, which is painful, of rigid consistency and is fixed to deep planes, in addition to this, the patient reports loss of 10 kg weight in a month, therefore, we requested an X-ray (Figure 2) and pelvic limb tomography (Figure 3) in the face of findings compatible with a tumor lesion, a chest tomography was also requested to perform a scan, (Figure 4) finding lesions compatible with metastatic pulmonary activity. When faced with advanced disease, the left pelvic limb was disarticulated using the Boyd technique, obtaining a histopathological diagnosis of Ewing's Sarcoma and infiltration of muscle fibers and atrophy (Figure 5). With adequate post-operative evolution and no data of infection in the surgical wound, he was referred to the medical oncology service for treatment with adjuvant chemotherapy based on Docetaxel and Gemcitabine; currently in the fourth cycle, clinically and radiologically corroborating significant improvement in dyspnea and lung lesions as well as improvement in respiratory symptoms and quality of life.

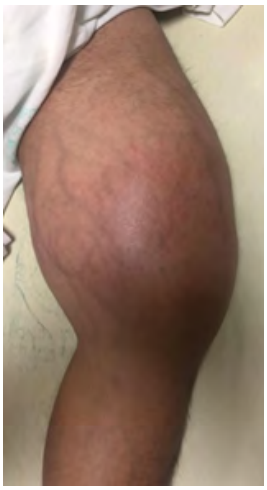

Figure 1: Tumor in the left pelvic limb on physical examination.

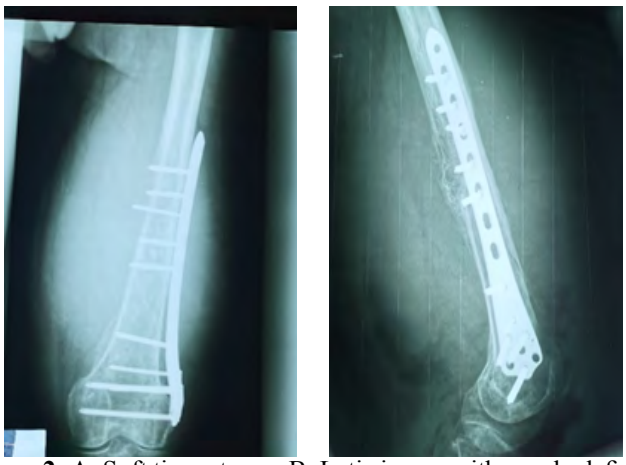

Figure 2: A: Soft tissue tumor B: Lytic image with poorly defined edges, with permeative and moth-eaten pattern, spiculated and multilaminar discontinuous periosteal reaction.

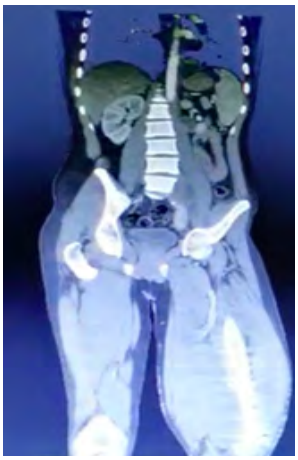

Figure 3: Heterogeneous tumor with irregular, poorly defined edges, 198X240 mm at the level of muscle structures with infiltration and bone remodeling that breaks the cortex, predominantly in the middle and lower third of the femur

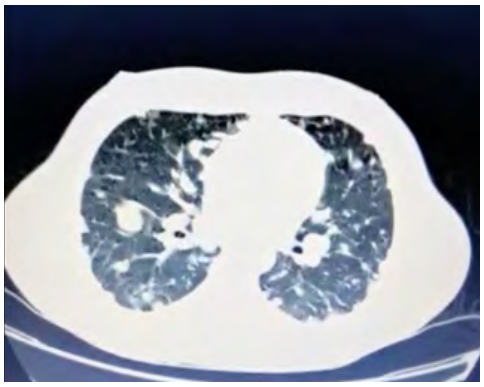

Figure 4: Lung parenchyma with multiple diffuse nodular lesions in both hemithorax and infiltrating mediastinal and parahilar adenomegalies that form 32 mm conglomerates with hypodensity suggestive of necrosis.

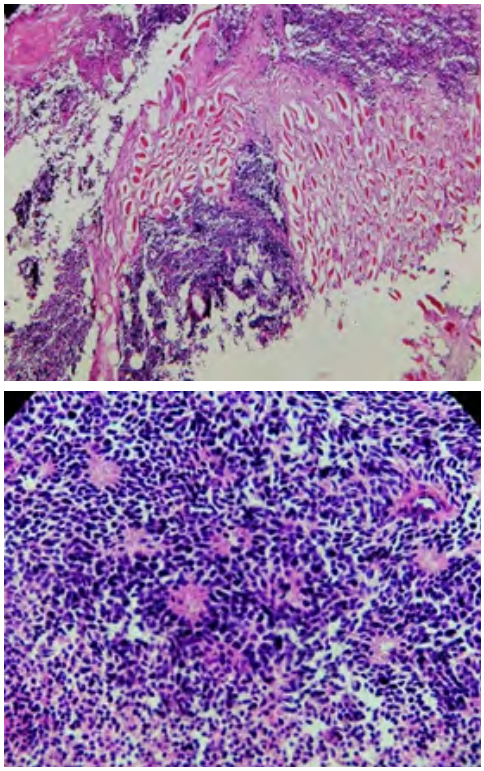

Figure 5: A: Histological section with hematoxylin-eosin stain, in which tumor infiltration of atrophic muscle fibers is observed. B: Homer-Wright type rosette formation in Ewing's sarcoma.

Results

The surgical technique is described below. The subcutaneous cellular tissue is dissected until the femoral neurovascular bundle is located, which is ligated and cut, the sartorius and rectus femoris muscles are disengaged from the anterosuperior and anteroinferior iliac spine respectively and are retracted distally, the pectineus muscle is dissected distal to the pubis, external rotation of the limb is performed and the psoas muscle is removed from the lesser trochanter. The adductor muscles of the hip and the gracilis muscle in their proximal insertions are dissected, the obturator artery is located, ligated and cut. Subsequently, the limb is rotated internally, the gluteus minimus, medius, and the most distal fibers of the gluteus maximus are disengaged from the rough line and the sciatic nerve is then located to perform ligation and cutting. The external rotators of the hip are detached from their origin. The section of the articular capsule and the round ligament are carried out, the gluteal flap is carried towards the anterior region, the distal part of these muscles is sutured at the insertion site of the pectineus and the obturator (Figure 6).

Figure 6.- Macroscopic piece in cross section where infiltration to muscle fibers is observed.

Discussion & Conclusion

Ewing's sarcoma is an entity that occurs especially in a young population, so it is of most importance to make the correct diagnostic approach in the presence of pathological or suggestive fractures in this type of patient due to the high index of presence of metastases at the time of diagnosis, likewise, perform a multidisciplinary treatment and a correct individualized surgical approach to each patient, in which the objective is the total resection of the tumor, as well as a higher quality of life. In the case presented, it was decided to perform a disarticulation with the Boyd technique

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