Case report

Fracture, beyond trauma

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Abstract
Case presentation. A 19-year-old patient, with no previous medical history, with a history of trauma to the right thigh, presented with shortening, edema, rotation of the right lower limb and pain. Radiography identified a fracture in the proximal third of multiple fragments of the diaphysis with thickening and periosteal reaction that generated a suspicion of a bone tumor. MRI confirmed a bone neoplasm of malignant characteristics in the upper third of the femur with destruction of the cortex and invasion of the endomedullary canal without signs of metastatic lesions. The biopsy confirmed the diagnosis of localized Ewing’s sarcoma. Treatment. The intrahospital management consisted of anti-inflammatory drugs and immobilization of the right lower limb for 21 days. Then, she received three cycles of chemotherapy with the Ewing sarcoma phase 1 scheme. In addition, physical therapy, outpatient treatment with analgesic, radiotherapy and finally partial hip resection was indicated. Outcome. There was a decrease in local edema, pain control with oral medications, mobility was recovered, although the functional limitation of the right lower limb was maintained, making it impossible to stand and walk.

Keywords
Ewing’s sarcoma, Leg bones, sarcoma, needle biopsy.

Introduction
Ewing sarcoma is a malignant primary bone tumor that presents as a necrotic or hemorrhagic mass mainly located in the metaphysis of the bones¹. Microscopically, it is presented as a collection of rounded, monomorphic cells¹. This tumor was described as a round cell sarcoma by James Ewing in 1921; It was initially described as a diffuse endothelioma of bone and later as endothelial myeloma².

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Resumen
Presentación de caso. Se presenta el caso de un paciente de 19 años, sin antecedentes médicos, con historia de traumatismo en el muslo derecho, que tuvo acortamiento, edema, rotación del miembro inferior derecho y dolor. En la radiografía se identificó una fractura en el tercio proximal de fragmentos múltiples de la diáfisis con engrosamiento y reacción perióstica que generaron una sospecha de un tumor óseo. La resonancia magnética confirmó una neoplasia ósea de características malignas en el tercio superior del fémur con destrucción de la cortical e invasión del canal endomedular sin signos de lesiones metastásicas. La biopsia confirmó el diagnóstico de sarcoma de Ewing localizado. Intervención terapéutica. El manejo intrahospitalario consistió en antiinflamatorios e inmovilización del miembro inferior derecho por 21 días. Luego, recibió tres ciclos de quimioterapia con el esquema para sarcoma de Ewing fase 1. Además, se indicó terapia física, tratamiento ambulatorio con analgésico, radioterapia y finalmente se practicó la resección parcial de cadera. Evolución clínica. Se evidenció disminución del edema local, control del dolor con medicamentos orales y recuperación de la movilidad, aunque mantuvo la limitación funcional del miembro inferior derecho que imposibilita la bipedestación y la deambulación.

Palabras clave
Sarcoma de Ewing, huesos de la pierna, sarcoma, biopsia con aguja.
Primary bone cancers are clinically heterogeneous and their cure depends on the opportunity of appropriate treatment. In the adult population, Ewing sarcoma ranks fourth, being the most common with chondrosarcoma, followed by osteosarcoma and chordoma, unlike the incidence in children and adolescents, which is presented as the second cause of bone cancer in the population among children and adolescents after osteosarcoma.

The skeletal distribution is wide, although the most common location is the femur, tibia and fibula, as well as the pelvis in the axial skeleton. Symptoms are usually nonspecific such as pain and sometimes tumor, without others that suggest a local problem or metastasis. The initial diagnostic method is simple radiography, in which the aggressive characteristics and high-grade nature of this malignant lesion are presented. Recommended imaging studies are contrast computed tomography, which provides information on cortical, changes in bone structure, and magnetic resonance imaging (MRI) as a technique of choice for identifying bone and extra osseous extent of the tumor and defining local stage definition.

The diagnosis is confirmed with biopsy of the tumor and should be supplemented with leukogram and blood chemistry.

The objective of describing this case is assessing the importance of establishing timely and adequate management in patients with suspected pathological fractures, exploring certain characteristics suggestive of malignancy and using the available diagnostic methods.

Case presentation

This is a 19-year-old male patient, who had a medical consultation at San Rafael National Hospital for severe pain in the right lower limb for two months, followed by episodes of rhabdomyolysis. It was decided to admit him to hospital, where he received treatment with analgesics and muscle relaxants.

Three months later, he consulted at the previous hospital emergency unit for a fall on same level. This generated a trauma to his right thigh. He was treated with 50 mg of diclofenac intramuscularly and then referred to the General Hospital of the Salvadoran Social Security Institute (ISSS).

On physical examination, the patient was conscious, oriented, complaining, unable to roam, with a temperature of 36.7°C, heart rate of 68 beats/minute and blood pressure of 120/80 mmHg. As far as cardiopulmonary auscultation, he presented 12 breaths per minute, without pathological findings in both pulmonary fields, with oxygen saturation of 95%. There was not any abnormality in the abdominal evaluation. The right lower limb was rotated to the right, shortened, with edema in the middle and upper third of the thigh and presented pain on palpation and when performing the abduction and adduction movements and inability to walk. The popliteal and dorsal pulses of the foot remained normal.

Laboratory analytical data reported hemoglobin of 12.0 gr/dL without leukocytosis, with neutrophils of 72.5 % and platelets of 443 000. Creatinine, potassium, sodium and glucose were detected at normal values, while C-reactive protein was at 21.5 mg/L; the prothrombin time, in 14.8 seconds and the INR in 1.3.

He was clinically evaluated and an X-ray of the pelvis and right femur was indicated, in which a fracture of multiple fragments of the diaphysis was observed, in the proximal third of the right femur, with thickening and periosteal reaction of the cortical in “Codman’s triangle” with heterogeneous density and poorly defined central radiolucent areas. These signs generated the suspicion of a bone tumor with malignant radiological characteristics, a pathological fracture of the right femur was diagnosed and the study was continued with hospital management. The X-ray of the left femur, without abnormalities, allowed to compare both lower limbs. Also, to discard lung metastasis, a chest computed axial tomography (CT) was indicated, and metastatic lesions were not found.

The study of the thigh was continued by means of a magnetic resonance in sagittal slices, in which isointense was described in T1 sequences, heterogeneously hyperintense in T2 and STIR with marked restriction to diffusion and heterogeneous
enhancement after the administration of paramagnetic contrast material. A mass that destroys the cortical and invades the endomedular canal was described, which produced the complete and displaced pathological fracture of the femur, with displacement of the muscular structures of the anterior and medial compartment, towards superior and medial with edema in the associated soft tissues. It was concluded that these findings were highly suggestive of Ewing's sarcoma (Figure 3).

Treatment

Initially, he received hospital management with ketorolac 30 mg through a vein every eight hours for seven days; then, it was switched to tramadol 100 mg every 12 hours orally and immobilization of the right lower limb was maintained for 21 days. After one week of diagnosis, he was transferred to the ISSS cancer hospital for the start of chemotherapy with the schedule for Ewing sarcoma phase 1, with ifosfamide 2800 g, etoposide 160 mg, vincristine 2 g, MESNA 400 mg intravenously and filgastrim 300 μg subcutaneously from the fourth to the seventh day. Besides, a course of treatment was indicated each month until all three cycles were completed. Besides, physical therapy, oral treatment with acetaminophen 325 mg and codeine 16 mg every eight hours and tramadol 100 mg every 12 hours were indicated, received radiotherapy and finally partial hip resection will be performed.

Ultrasound-guided percutaneous biopsy was performed after extending the proximal femur (Figure 4), where 5 filiform samples varied from 0.3 to 1.7 cm were taken, with hematoxylin and eosin staining. Bone fragments and fibrous connective tissue with multiple foci of hepatocellular malignant neoplasm, arranged in nests or lobes were observed. The cells were small, round, with hyperchromatic nuclei with thick chromatin, of scanty cytoplasm densely packed with little mitosis and coagulation necrosis in 30 %. Immunohistochemistry was performed with the presence of high activity when applying CD99, Vimentin positive and CD45 negative, which confirmed Ewing's sarcoma in the right femur (Figure 5).

Outcome

The patient evolved with a decrease in local edema, pain control with oral medications, recovered mobility, although he maintained the functional limitation of the right lower limb, which makes standing and ambulation impossible.

Clinical diagnosis

Diagnosis of localized Ewing sarcoma (stage IA) was confirmed by radiological and
Discussion

Ewing sarcoma is a rare tumor that occurs most often in males aged five to 20 years. Although it can occur at any time of life, the highest incidence occurs at the age of 15. The estimated incidence is more than one million cases of children and adolescents worldwide. In Spain, it is the first cause of bone tumor in this population, while in the United States of America, it is the second cause between 1% and 2% of cancers diagnosed in children under 20 years of age.

The most common location of this tumor is the pelvis, femur and chest bones. When this tumor affects the long bones, it is most often located in the diaphysis. The clinical presentation of Ewing sarcoma is usually nonspecific. Symptoms such as tumor...
with pain on palpation or intermittent and progressive and edema are usually present for more than six months prior to diagnosis. The appearance of a soft mass must lead to a case study. Pathological fractures are unusual as an initial clinical presentation. They occur in 16 % of cases.

Imaging studies are important to locate the tumor, define its extent or volume and determine if there is metastasis to decide treatment and predict the evolution and outcome. The origin of bone lesions is the central medullary canal and it affects the soft tissues in more than 80 % of cases; the masses in these tissues are usually large and surround the affected bone. Cortical destruction can be generated that facilitates communication between the medullary canal and soft tissue components.

The radiological aspects of these tumors are very variable. The most common findings are the Codman’s triangle and the lamellated periosteal reaction or onion skin periosteal reaction, which result from the displacement of the periosteum and the proliferation of bone tissue. The periosteal reaction is the most frequent and has an aggressive behavior, followed by the destruction of the cortex associated with a mass of soft parts, and the least common is the destruction of the bone with a wide transition zone.

The radiological technique that allows to assess the bone and extraosseous extension of the tumor is magnetic resonance spectroscopy to confirm or discard an involvement of the bone marrow, that is, in phase T1; in these cases, the existence of foci far from the primary tumor should be discarded. If a cortical and soft tissue invasion is found, this is classified as phase T2.

Computed tomography presents the aggressive periosteal reaction and bone destruction, so it is considered useful in complex anatomical areas such as the pelvis, spine and base of the skull. Also, tomography in the chest area allows to identify a lung metastasis, which together with bone marrow and liver metastases are the most common ones. These are presented by hematogenous dissemination and are associated with alterations in laboratory tests, including high levels of alkaline phosphatase, erythrocyte sedimentation rate or lactic dehydrogenase, especially when the latter has extreme values.

The diagnosis is confirmed by histological analysis by a closed needle biopsy. In this procedure, local anesthesia is used and experience on the part of the doctor in charge of it is required not to produce pathological fractures and avoid tumor contamination of the surrounding tissues. In this case, ultrasound-guided needle biopsy was performed, which was timely for beginning medical treatment consisting of systemic chemotherapy. This is considered the mainstay of treatment and it is sometimes combined with surgery or radiotherapy since these play a role depending on the location and size of the tumor.

Survival rates have increased from 10 % to about 70 % over the past 40 years. The prognosis in the evolution of these cases depends on factors such as the presence or otherwise of metastasis, the location and size of the primary tumor, age, response to treatment and the presence of certain chromosomal translocations.

In patients older than 10 years, a higher frequency of negative prognostic factors has been described, including the presence in males, the axial location of the tumor, such as the head, spine, chest, pelvis or neck, and the presence and location of metastasis.

The survival rate is also related to the stage of the tumor at the time of diagnosis, with a five-year survival greater than 60 %, when the disease is localized, and about 20 % when there is metastasis. A survival analysis identified that 50 % of the cases with localized sarcoma died during six years of follow-up, with a median survival of 25 months, while cases with metastasis had a median survival of 9,75 months.

Patients with metastasis who have had a lower survival are cases of bone metastasis or a combination of lung and bone metastases.

It is expected that in cases of tumors without metastases, chemotherapy will generate a reduction of the tumor and surgical resection can be continued. Aggressive chemotherapy treatment has led to an increase in survival rates up to 65 % over the past five years.

Ethical aspects

For the presentation of this case, the patient was asked for informed consent respecting the principles of privacy and dignity according to the Helsinki guidelines.

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