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CLINICAL CASE

Intestinal Ewing sarcoma: An unusual presentation in the pediatric age

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Abstract

Background: *Ewing's Sarcoma (ES) is the second most common type of bone cancer, with an annual incidence of 2.9:100,000. Extraosseous cases represent 15%; however, there are no reported cases of ES located in the intestine in the pediatric population.* **Case report:** *We describe the case of a 14-year-old male patient, previously healthy, who started with an anemic syndrome, weight loss, and diaphoresis of 8 weeks of evolution. After visiting a physician, who documented the presence of anemia, the patient was referred to the National Institute of Pediatrics. Physical examination showed grade III-IV systolic murmur, splenomegaly, and pain in the left hemiabdomen with no irradiation. Computed axial tomography showed a mass-dependent on the peritoneum and intestinal loop. A biopsy of the lesion showed intestinal ES. The lesion was completely resected, and the patient was treated with chemotherapy and radiotherapy. Thirty months after diagnosis, the patient has no evidence of tumor activity.* **Conclusions:** *Extraosseous presentation of ES in pediatric age is rare. There are no reports of intestinal ES in the Latin American pediatric population, although eight case reports were found in adults. ES is curable by a combination of chemotherapy, radiotherapy, and surgery. The medical literature indicates that the extraosseous presentation should receive the same treatment as the osseous presentation, which can provide a survival rate of up to 70% if there is no evidence of metastasis (which most frequently is observed in the lung).*

Keywords: *Abdominal neoplasm. Ewing sarcoma. Malignant mesenchymal tumor.*

Sarcoma de Ewing intestinal: presentación inusual en la edad pediátrica

Resumen

Introducción: *El sarcoma de Ewing (SE) es el segundo tipo cáncer más común de hueso, cuya incidencia anual es de 2.9:100,000. Los casos extraóseos representan el 15%; sin embargo, no existen reportes en la literatura de casos de SE ubicados en el intestino en la población pediátrica.* **Caso clínico:** *Se describe el caso de un paciente de sexo masculino de 14 años, previamente sano, que inició con síndrome anémico, pérdida de peso y diaforesis de 8 semanas de evolución. Acudió con un médico, quien documentó la presencia de anemia y lo refirió al Instituto Nacional de Pediatría. A la exploración física presentaba soplo sistólico grado III-IV, esplenomegalia y dolor en hemiabdomen izquierdo sin irradiaciones. La tomografía axial computarizada mostró una masa dependiente del peritoneo y asa intestinal. La biopsia de la lesión reportó SE intestinal. Se resecó por completo la lesión y el paciente recibió tratamiento con quimioterapia y radioterapia. Después de 30 meses del diagnóstico, el paciente se encuentra sin datos de actividad tumoral.* **Conclusiones:** *La presentación*

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extraósea del SE en edad pediátrica es rara. No existen reportes de presentación de SE intestinal en la población pediátrica latinoamericana, aunque se encontraron ocho reportes de caso en adultos. El SE es curable mediante la combinación de quimioterapia, radioterapia y cirugía. La literatura médica indica que la presentación extraósea debe recibir el mismo tratamiento que la ósea, lo cual puede proporcionar una sobrevida de hasta el 70% si no hay evidencia de metástasis (que ocurre más frecuentemente a pulmón).

Palabras clave: *Masa abdominal. Sarcoma de Ewing extraóseo. Tumor maligno mesenquimatoso.*

Introduction

Ewing's sarcoma (ES) is the second most frequent primary malignant bone tumor in the pediatric age group, with an estimated incidence of 2.9:100,000. ES represents 34% of primary bone tumors[1,2]. The axial skeleton, pelvis, femur, and ribs are the most common locations. The most frequently reported site of metastasis is the lungs. In children, ES is more frequent in males, with a ratio of 3:2 and a mean age of 14 years[1,3]. ES is a very aggressive cancer, with a survival rate of 70% to 80% for patients with localized, usual-risk disease and 30% for those with metastatic disease[1], representing 20-30% of cases. ES originates from mesenchymal cells. Therefore, 15% of ES are extraosseous as they arise from different tissues. Here, we describe an unusual case of intestinal ES in a pediatric patient.

Clinical case

We describe the case of a 14-year-old male with no relevant history. The patient presented with an anemic syndrome, unintentional weight loss of 6 kg, and diaphoresis of 8 weeks of evolution. The attending physician requested a complete blood count that revealed non-regenerative anemia, with hemoglobin (Hb) 5.3 g/dl. Subsequently, the patient was referred to the National Institute of Pediatrics.

Initial physical examination showed pallor, systolic murmur III-IV, splenomegaly, and pain in the left abdomen. Among the initial studies, abdominal ultrasonography (USG) showed a mass of 115 x 102 x 80 mm with hypodense areas on the left flank, and computed tomography (CT) showed evidence of a peritoneum-dependent tumor without abdominal lymphadenopathy. PET/CT with FDG (fluorodeoxyglucose positron emission tomography/CT) showed an 11.2 x 10.5 x 7.2 cm tumor in the left flank, extending longitudinally from the lower splenic border to the upper third of the proximal insertion of the ipsilateral psoas muscle and transversely from the anterior abdominal wall to the anterior border of the left kidney with no infiltrations. A USG-guided biopsy of the abdominal mass was performed, showing monotonous cells with round nuclei, inconspicuous nucleolus, and scant eosinophilic cytoplasm arranged in solid clusters intermixed with areas of tumor necrosis (Figure 1). Immunohistochemical staining with CD99 and Fli-1 was positive (Figure 2); CD-117, myeloperoxidase (MPO), Wilms tumor (WT-1), CD34, and extranodal spread (ENS) markers were negative. Due to these histological and immunohistochemical findings, Ewing's sarcoma was diagnosed.

Complete surgical resection was performed (Figure 3), and chemotherapy treatment with vincristine/doxorubicin/cyclophosphamide and etoposide/ifosfamide was started alternately for seven cycles. The patient also received radiotherapy to the abdomen and pelvis at 24 Gy. Later, a new cycle was administered to the pelvis at 26 Gy. Thirty months after diagnosis, the patient is under close follow-up and the PET scan has shown no evidence of tumor activity.

Discussion

As ES originates from mesenchymal progenitor cells, it has the ability to develop in multiple tissues. The most common clinical manifestation is mild pain that worsens with exercise and at night, including symptoms of spinal cord compression manifested by weakness or loss of sphincter control when the primary lesion is present in an axial location. Constitutional symptoms are present in 10-20% at diagnosis. Symptoms vary depending on the affected area[1]. This variation represents a challenge because the clinician must integrate the indolent manifestations until the precise diagnosis.

The final diagnosis always requires histopathological support to confirm the presence of CD99 and EWS-FLI1 (Ewing sarcoma breakpoint region 1/Friend leukemia integration 1 transcription factor) with immunohistochemical studies[4]. Extraosseous presentation of ES is infrequent[4]; most ES are reported in the

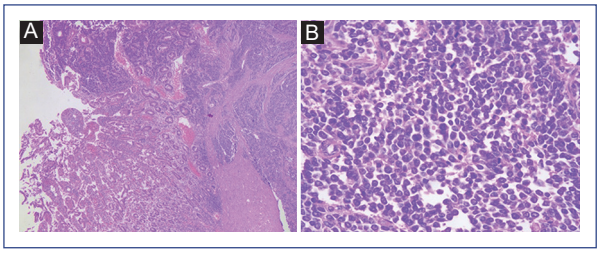

Figure 1. A: histological section showing the transition zone between the preserved intestinal wall and the tumor (H&E 5x). **B:** small monotonous cells with round nuclei and little eosinophilic cytoplasm (H&E 40x).

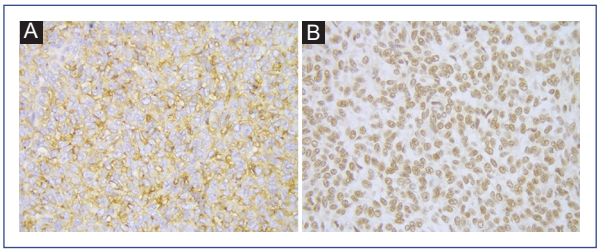

Figure 2. A: positive CD99 on the tumor cell membrane. **B:** nuclear positive FLI-1 in tumor cells.

trunk, head, neck, and extremities[2,4,5]. Some studies in the literature have reported ES in the adrenal glands, kidneys, intestine, liver, and peritoneum[3,4,6-10]. Intestinal presentation is more frequent than skeletal ES in adult women. In contrast, no reports were found on intestinal ES in the pediatric age group[4].

In this patient, the underlying data led to the initial diagnosis of an anemic syndrome with few abdominal symptoms that delayed the diagnosis. Most extraosseous ES can be cured with the combination of chemotherapy, radiotherapy, and surgery; the treatment scheme is decided with the same guidelines as in skeletal ES. The evidence so far indicates that extraosseous involvement is not a poor prognostic factor, as survival is affected by other factors such as the stage of the disease at diagnosis and the resectability of the primary tumor. In most series, the extraosseous presentation was mostly muscular, with greater involvement at the level of the pelvic limbs. Therefore, in the case presented here, the tumor's location did not play an essential role in the patient's prognosis. Hence, a 5-year survival of 70% is expected, with an adequate response to the first line of treatment[1].

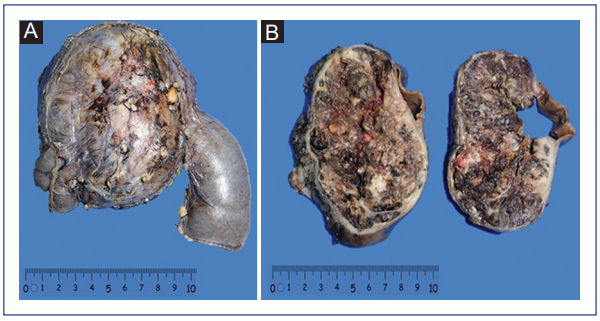

Figure 3. A: macroscopic image of the tumor adhered to the jejunal wall. **B:** after cutting, invasion and destruction of the intestinal wall are observed.

Extraosseous presentation of ES in pediatric age is rare. Therefore, it requires a multidisciplinary approach to diagnose it accurately. Until now, there are no reports in the Latin American literature of intestinal presentation of ES in the pediatric age group.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author has this document.

Conflicts of interest

The authors declare no conflict of interest.

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