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Paediatrics

CHILDHOOD EWING SARCOMA OF THE BLADDER: A RARE CANCER

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ABSTRACT Ewing sarcoma of the bladder is an exceptionally rare form of bladder tumor, accounting for less than 1% of all cases. The exact cause of this condition remains unknown, and molecular testing has shown that the presence of the EWSR1 gene rearrangement is essential to confirm the diagnosis. Treatment for Ewing sarcoma of the bladder typically involves a comprehensive approach, combining chemotherapy, surgery, and radiation therapy. The prognosis for patients with this condition can vary widely, depending on the stage of cancer at the time of diagnosis and the response to treatment. However, overall, Ewing sarcoma of the bladder is considered a highly aggressive cancer with a poor prognosis. This is often due to late-stage diagnosis and the high likelihood of recurrence. We report a case of a 1- year-old boy diagnosed with Ewing sarcoma of the bladder while discussing the appropriate literature.

KEYWORDS : Ewing sarcoma - Bladder - Chemotherapy

Introduction:

Ewing sarcoma (ES) is a rare type of bone and soft tissue cancer that typically occurs in children and young adults. However, in rare cases, it can also develop in other parts of the body, including the bladder. ES of the bladder is an aggressive and exceptionally uncommon cancer that necessitates specialized care and attention. In this article, we present a case of a 15-year-old boy diagnosed with ES of the bladder and treated in the Department of Medical Oncology at the military hospital in Rabat.

Case report:

A 15-year-old child with no significant medical history, who has been experiencing macroscopic hematuria with suprapubic pain for two weeks. Clinical examination revealed conjunctival and skin pallor, as well as hypogastric tenderness. Abdominopelvic ultrasound revealed a large intravesical tissular mass. Transurethral resection of the bladder was performed and the histopathological examination confirmed an undifferentiated round cell tumor infiltrating the bladder muscle (Figure 1).

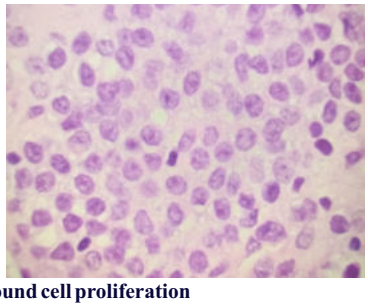

Figure 1: Round cell proliferation

Immunohistochemical phenotyping study was necessary, anti-CD99 antibody (Figure 2), anti-PS100 antibody and anti-synaptophysin antibody were positive. The anti-CD20 antibody, anti-CD3 antibody, anti-CD34 antibody, anti-Pancytokeratin antibody and anti-EMA antibody, were all negative. The immunohistochemical profile was in favor of a round cell tumor of the Ewing sarcoma type. Molecular analysis for the EWS fusion transcript was not performed.

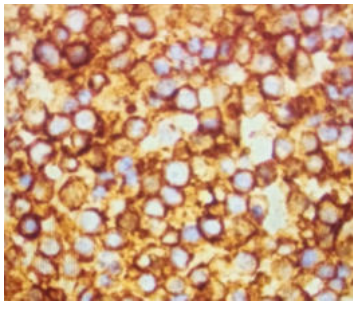

Figure 2: Anti CD99 antibody positive

A thoraco-abdomino-pelvic CT scan showed a tumoral process (73x65 mm) of the bladder floor budding into the bladder lumen without secondary lesions (Figure 3).

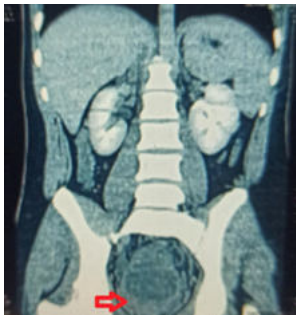

Figure 3: Tumoral process of the bladder

The patient received chemotherapy according to the VIDE protocol, with an improvement in symptoms after 2 cycles of treatment. Radiological control at 4 months of treatment revealed a spectacular response with a 75% reduction in tumor mass. The patient underwent radiotherapy to the bladder bed at a dose of 64 Gray, followed by 4 additional months of chemotherapy using the same protocol. Evaluation by CT scan showed a maintained complete response after 2 years of follow-up.

Discussion:

Ewing sarcoma is a malignant tumor that originates from primitive nerve cells and predominantly impacts the pediatric and young adult population. It most commonly arises in bones, such as the pelvis, femur, and ribs, but can also occur in other organs and tissues, including the bladder, though such occurrences are exceedingly rare, accounting for less than 1% of bladder tumors (1).

The exact cause of ES of the bladder remains unknown, but it is believed to be associated with genetic mutations that trigger abnormal cell growth in the bladder. Due to its rarity, risk factors are not well-established, and there are no known preventive measures for this type of cancer.

The presentation of symptoms in ES of the bladder can vary depending on the location and size of the tumor. Common symptoms include hematuria, lower abdominal or pelvic pain, frequent urination, urinary urgency, and a palpable mass in the lower abdomen. However, these symptoms are nonspecific and can be attributed to other conditions, often resulting in delayed diagnosis.

Diagnosing ES of the bladder necessitates a comprehensive evaluation involving medical history, physical examination, imaging studies, and

biopsy. Imaging studies such as X-ray, computed tomography (CT) scan, magnetic resonance imaging (MRI), and bone scan are used to identify the location, size, and extent of the tumor. Biopsy, which involves obtaining a small sample of the tumor for microscopic examination, is crucial for confirming the diagnosis. Molecular tests are essential for confirming the diagnosis, either through FISH or RT-PCR to detect the EWS rearrangement (2,3).

Once diagnosed, the management of ES of the bladder typically involves a multidisciplinary approach with a team of healthcare professionals, including oncologists, surgeons, and radiation oncologists. The treatment plan depends on various factors, including the size, location, and stage of the tumor, as well as the age and overall health of the patient. The main treatment modalities include surgery, chemotherapy, and radiation therapy.

Surgery aims to remove the tumor from the bladder and may involve partial or complete removal of the bladder (cystectomy). Chemotherapy is typically administered before surgery to shrink the tumor and after surgery to target any remaining cancer cells. VIDE (vincristine, ifosfamide, doxorubicin and etoposide) is the most used protocol. Radiation therapy, may be used in combination with surgery and/or chemotherapy to eradicate the tumor (4,5).

Despite aggressive treatment, ES of the bladder has a poor prognosis, with an overall survival rate at 5 years ranging from 20% to 60%. Prognosis depends on various factors, including the stage of the tumor, presence of metastasis, and response to treatment. Early diagnosis and prompt initiation of treatment are crucial for improving the prognosis (4,6).

Conclusion:

ES of the bladder is a rare and aggressive form of cancer that primarily affects children and young adults. It is characterized by the abnormal growth of cells in the bladder and can cause symptoms such as hematuria. Diagnosis requires a comprehensive evaluation, and treatment typically involves a multidisciplinary approach with surgery, chemotherapy, and radiation therapy. Despite challenges in prognosis, early detection and timely treatment are vital for improving outcomes.

Conflict of interest: None

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