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RESEARCH ARTICLE

LOCALLY ADVANCED BLADDER EWING'S SARCOMA: AN EXCEPTIONAL CASE REPORT

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Abstract

Extra skeletal Ewing's sarcoma is a rare entity. The primary bladder localization is exceptional, their diagnosis is difficult because it is extremely rare in this place and because of diagnostic difficulties posed by a group of very heterogeneous tumors. Its clinical presentation is similar to that of transitional cell carcinoma, however the two entities must be distinguished because the protocols for treatment are different. The diagnosis is dependent on the pathological examination with the immunohistochemical and cytogenetic study. We report the case of a 36-year-old man with locally advanced Ewing sarcoma of the bladder.

Introduction:-

Extraskeletal Ewing's sarcoma is a rare disease. It is a malignant mesenchymal tumor with small round cells, belonging to the group of primary neuroectodermal tumors with a genetic predisposition. Primary bladder localization is exceptional and poses a problem of differential diagnosis because it is an extremely rare disease in this localization, their main revealing sign was macroscopic hematuria sometimes associated with signs of bladder irritation. The diagnosis is based on the pathological examination coupled with immunohistochemical and cytogenetic study. We report an exceptional case of a 36-year-old patient with locally advanced Ewing sarcoma of the bladder.

Patient And Observation:-

Mr. HB, 36 years old, chronic tobacco user at a rate of one pack per day was hospitalized in the urology department for treatment of intermittent clot hematuria associated with irritative bladder signs without fever and weight loss not quantified. The clinical examination found a conscious patient stable hemodynamic and respiratory constants with a preserved general condition, the abdomen is flexible, on the digital rectal examination there is a prostate estimated at 30g with a mobile infiltrated bladder base. Ultrasound and cystoscopy show a large budding tumor occupying the left anterior and lateral wall measuring 08cm with moderate dilatation of the left ureteropyolcalialis (UPC) without affecting renal function, the urine cytology is negative. Endoscopic resection of the tumor confirmed on pathological examination with immunohistochemistry the diagnosis of vesical Ewing sarcoma (Figure 1).

The extension assessment comprising: a TAP CT scan performed showed a large abdomino-pelvic mass invading the anterior abdominal wall without a separation border, invading the sigmoid colon, invading the left external iliac vein (figure 2). The patient was then referred to the medical oncology department for chemotherapy. The clinical evolution after 03 courses of chemotherapy finds a patient in a general condition preserved without hematuria, radiologically we note a stability of the lesion hence the decision to add 03 other courses. The patient is still under surveillance in the oncology department to continue the management of his condition.

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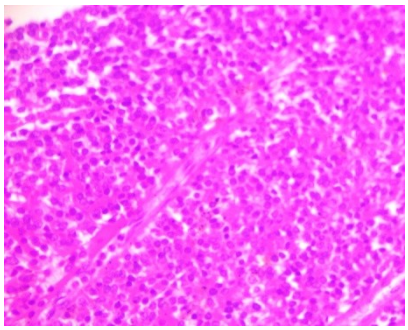

Figure 1:-

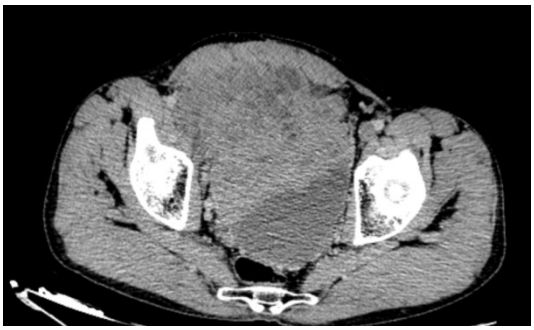

Figure 2:-

Discussion:-

Ewing's sarcoma is a malignant tumor with small round cells located mainly in the bones and exceptionally in the visceral area (1). Rare cases of primary Ewing's sarcoma of the bladder have been reported. The age of the patients ranges according to the authors between 10 and 80 years with an average age of 44 years (2). It appears earlier than urothelial tumors like our patient, where the age is 36 years. These clinical manifestations depend essentially on the location of the tumor and its extension to surrounding organs (3). Hematuria is the most frequent symptom found in bladder localization as is the case in our study. Its differential diagnosis includes other malignant tumors with small round cells (4) but the confirmation remains pathological with an immunohistochemical study. This family of tumors is characterized by a translocation most often involving the EWS gene of chromosome 22, and the FLI-1 gene carried by chromosome 11 encoding a new fusion protein which functions as a potent activator of transcription (5.6). Much more rarely, t (7; 22), t (17; 22), t (2; 22) translocations have been described. In all cases, an abnormal protein is produced leading to continuous activation of the IGF-1 membrane receptor responsible for cell proliferation. Their diagnosis is often made at an already advanced stage, as is the case in our study, or even metastatic. Adequate treatment of these tumors would combine radical surgery with adjuvant or neoadjuvant chemotherapy (7). Although the response of sarcomas to high-dose radiation therapy is well defined, its use in Ewing's sarcoma of the bladder is not yet well established. Our case was a 36-year-old patient with no comorbidity, he had locally advanced Ewing's sarcoma of the bladder at the time of diagnosis. the patient underwent endoscopic tumor resection with chemotherapy. The most indicative prognostic factor is the stage of the disease. The clinico-radiological outcome was unfavorable after a follow-up of 02 years with persistent lesions and the appearance of distant metastasis.

Conclusion:-

Ewing's sarcoma of the bladder is a rare entity. Raising a problem of differential diagnosis with a group of very heterogeneous tumors. The diagnosis is based on pathological examination with immunohistochemical and cytogenetic study. Surgical treatment with chemotherapy should be considered as an option, especially in the advanced Ewing sarcoma family of the bladder.

Tables and figures:

Figure 01: Histological appearance of the resected mass: presence of conformal ovoid cells with an enlarged nucleus of fine chromatin size with scant cytoplasm

Figure 02: Abdomino-pelvic CT scan showing a huge vesico-pelvic mass with invasion of the anterior abdominal wall

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