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Paratesticular Ewing's sarcoma: A case report

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ABSTRACT

Introduction:
Ewing extra bone sarcoma is a rare entity accounting for 4% of soft tissue tumors[1] and 1.5 to 4% of sarcomas[2,3]. It usually occurs in the extremities or soft tissues deep, cutaneous and subcutaneous. Paratesticular localization is exceptional[4]. We are reporting a case of paratesticular Ewing's sarcoma in a 19-year-old patient by neoadjuvant chemotherapy followed by surgery.

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Observation:

We are reporting the case of a young man of 19 years chronical and occasional cannabis consumer. He initially consulted for a painful right scrotal perineo mass, increasing gradually volume measuring 5cm large diameter. A testicular ultrasound has beenmade, that showed an absurd infiltration of the scrotal envelopes without being able to eliminate a testicular origin. One month later, due to a significant increase in scrotal mass, the patient consulted an urologist, who requested a second testicular ultrasound. This one has found a intertesticular tissue mass of scrotal envelopes.

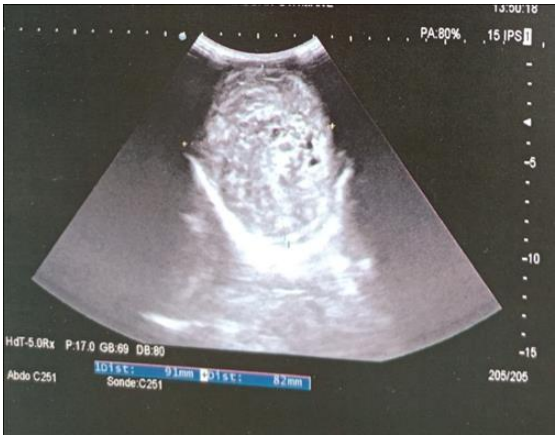
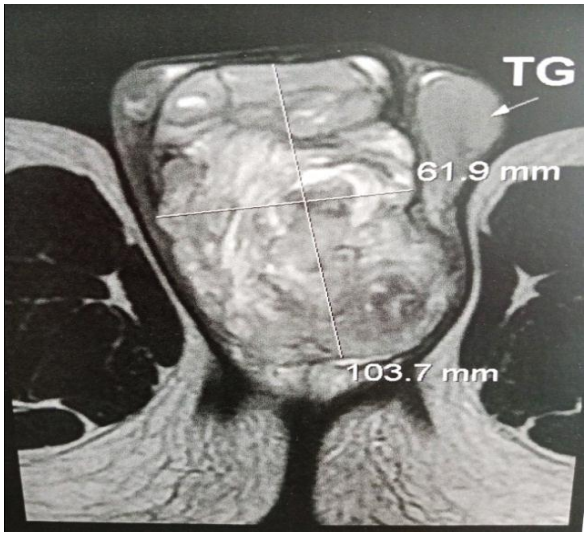


Figure 1 : ultrasound image of an inter-testicular tissue mass of the scrotal envelopes

The serum tumor markers BHCG and AFP were negative. The investigation was completed by aTesticular MRI which found: Straight intracotral mass extra testicular voluminous weakly enhanced by the contrast non-specific aspect that may be related to a tumor scrotal envelopes.

The patient was operated a week later, on the exploration there was a huge mass fistulized at ascrotal level and extending from the perineum forward of the anus, to the base of the penis intertesticular septum level. The testicles were intact with their vaginals.



Figures 2 : MRI appearance of an extra testicular scrotal mass measuring 61 X 103 mm in large diameter.

Histological examination of the resected mass found vaguely lobulated tumor proliferation, made of lobules dissociated by hemorrhagic suffusions, and sometimes separated by a fibrous tissue, dense, and desmoplastic. The bottom is myxoid. This proliferation is made of round cells, quite monomorphic, with poorly individualized cytoplasm and irregular dense nucleus with presence of foci of necrosis.

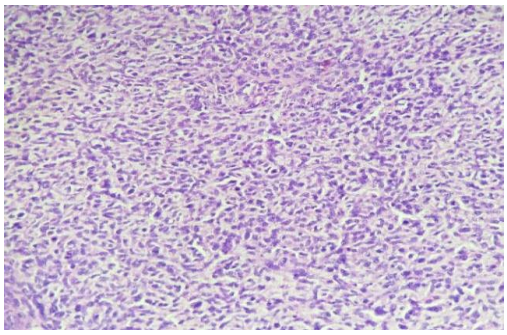


Figure 3 : histological section made of round cells, Hematein eosin staining, magnification X10

The immunohistochemical study carried out showed an intense and membranous expression of CD99 with no expression of the following markers: CKAE1 / AE3, Desmin, myogeny, Alpha-FP, PLAP, Oct ¾, CD3, CD20, CD30, STAT6, CD34, MDM2, and PS100. With these morphological and immunohistochemical arguments, the diagnosis of Ewing Sarcoma scrotale was held.

The patient was seen in consultation at the oncology center of Ibn Rochd University Hospital for the treatment surveillance, 2 months after the initial surgery.

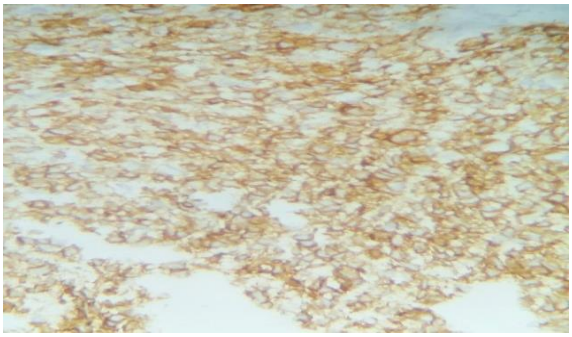


Figure 4 : Intense expression of CD99

The clinical examination found a hard mass measuring 6cm long axis with inguinal ADP Bilateral and infracentimetrics. Extension Thoraco-Abdomino-Pelvic CT showed at the pelvic level bilateral inguinal ganglia of 11 mm in diameter on the right and 7 mm in left. The file was discussed in a multidisciplinary consultation meeting whose decision was to start neoadjuvant chemotherapy according to the protocol VAC-IE, followed by surgery depending on the tumor response.

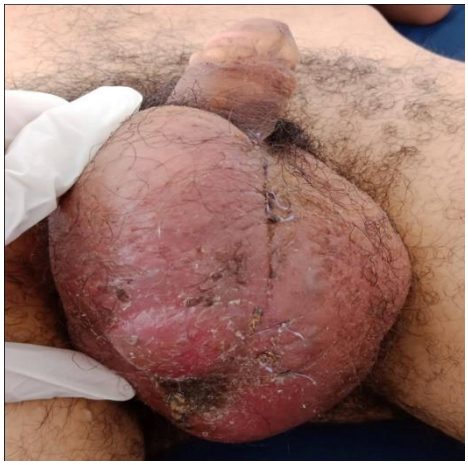


Figure 5: large scrotal mass

Discussion:

Primitive paratesticular Ewing's sarcoma was first described in 2014 in a case of a child of 3 years old. It was revealed by a scrotal mass of 2 cm which increased gradually increase in volume. The patient was without any notable pathological antecedents and completely asymptomatic. Testicular ultrasound showed a heterogeneous mass, vascularized, at the expense of scrotal envelopes, measuring 1.8 cm in diameter.

A surgical excision was performed whose histological examination objectified the presence of a infiltrating neoplasm, made of C round uniform cells with round CORES, compatible with a Ewing's sarcoma and confirmed by a cytogenetic study. The margins of resection have been tumors and the extension assessment was negative. The patient was put under chemotherapy adjutant, based on vincristine, doxorubicin and cyclophosphamide.

Our patient was a young adult, the scrotal mass was painful, huge and aggressive. Testicular ultrasound and histology data are consistent with the case described above. Ewing's sarcoma is a round-cell tumor with a variable degree of neuroectodermal differentiation. The clinical symptomatology is non specific, made through finding a scrotal mass, and or the appearance of testicular pain. [5]

Testicular ultrasound is the first-line examination, it shows the malignant origin of the tumor mass. It displays a tissue mass of heterogeneous and vascularized Doppler echostructure, but sometimes it can not specify the testicular or extratesticular origin, where the MRI is useful because it allows a more precise analysis of these structures. MRI gives theaccurate measurements of the tumor in the three planes of space, as well as the locoregional extension. It is an essential aid for determining volumes targets for radiotherapy, and monitoring patients during a chemotherapy to evaluate its efficiency. [6]

The determination of serum tumor markers is essential to eliminate a germ cell tumor. Histological examination shows a tumor with small basophilic round cells,their architecture more or less lobulated, dense, uniform appearance. The cells are in relatively monomorphic effect, with rounded to oval nuclei, vesiculous membrane with a nuclear membrane, with homogenous finely granitized chromatin containing a small nucleolus. The cytoplasm is pale, poorly limited and abundant. [7]

The diagnosis is essentially cytogenetics and immunohistochemistry, especially due to the in situ hybridization by immunofluorescence. In Immunohistochemistry, tumor cells express CD99, which can be positive in other tumors namely: the lymphoma which expresses the lymphoid markers, that are CD45RB, CD3, CD20 and TdT; neuroblastoma that expresses markers neuroendocrine (synaptophysin, chromogranin), rhabdomyosarcoma or markers of skeletal muscle, namely, desmin, myogenin, myo-D1, and the myoglobin are positive, and synovial sarcoma that expresses pancytokeratines,EMA, BCL2 and calponin. Several immunohistochemical dyes are used to have a diagnosis definitive and eliminate other diagnoses. [8]

For our case the diagnosis was made thanks to immunohistochemistry taking into consideration the overexpression of CD99 and the negativity of other markers. Ewing's sarcoma has a translocation (11, 22) (q24, q12) involving the gene EWS (Ewing's sarcoma gene) and 5% to 10% translocation (21,22) (q22, q12).

This histogenetic analysis allows diagnosis in difficult cases, when Immunohistochemistry is unable to decide. [9] Although there is no consensus regarding the management of sarcomas of paratesticular Ewing, the treatment is like in cases of Ewing's soft tissue sarcomas.

Surgery is considered by the majority of authors and constitutes the main option of local control. Radiation therapy and chemotherapy are introduced into the therapeutic plan to improve locoregional control, and reduce metastatic diffusion. [10]

Our patient did not receive adjuvant treatment after the first resection by lack of means. He had a local and ganglionic relapse before consulting

his oncologist. The first chemotherapy was therefore indicated before moving on to surgery. The most commonly used chemotherapy regimen was VAC / IE, which showed its efficiency for localized and metastatic diseases. Adjuvant radiotherapy remains questionable but none of the studies that have used it has given details concerning the dose and the modality. [10]

Conclusion:

The paratesticular localization of Ewing's Sarcoma is exceptional, it is an aggressive tumor whose current management includes surgical resection and chemotherapy. A unified therapeutic protocol must be proposed for Future case to ensure better local control, and there for to improve the overall survival rates.

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