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Case Report

Paratesticular Embryonal Rhabdomyosarcoma on Ectopic Testis: A Case Report

Raouah Mehdi[1], Bouchabaka Yassine[1], El Matlini Abdelali[1], Saadoune Mohamed[1], Laatitioui Sana[1], Igarramen Tariq[1], Bounid Oumayma[1], Darfaoui Mouna[1], Lalya Issam[1], El Omrani Abdelahamid[1], Khouchani Mouna[1]
[1]Department of Radiation Oncology, Mohammed VI University Hospital, Marrakech, Morocco

*Corresponding author: **Raouah Mehdi**
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Abstracts: Para testicular rhabdomyosarcoma is a rare malignant tumour, which usually presents as a painless mass in the scrotum or groin. A case of paratesticular embryonal rhabdomyosarcoma in a 16-year-old male, with a history of left testicular ectopia treated with orchiopexy, is being reported here who presented with chronic scrotal swelling. Paratesticular rhabdomyosarcoma is a rare non germ cell tumour of scrotal sac in children and young adult/teens which can invade testis at presentation. Embryonal variant is the most common type. 40% cases can have metastasis to retroperitoneal lymph node. Diagnosis can be done on high degree of clinical suspicion coupled with biopsy and immunohistochemistry. Multimodality approach of treatment is often beneficial for patients.
Keywords: Embryonal, Paratesticular, Rhabdomyosarcoma, testicular ectopia.

INTRODUCTION

Testicular and paratesticular tumors in children are rare. Seven to 10% of primary genitourinary tumors locate in paratesticular region. Scrotal rhabdomyosarcomas generally originating from paratesticular tissue are most frequently seen in childhood and young adulthood (Stewart, L. H. *et al* 1991). Paratesticular rhabdomyosarcomas typically present as a unilateral, painless scrotal swelling or mass above the testis. Lymph node metastasis develops in approximately one-third of cases with paratesticular lesions (Shapiro, E., & Strother, D. 1992). Hematogenous metastases to lungs, liver, bone and bone marrow is present in 20% of patients at initial presentation (Demir, A. *et al* 2004). In some conditions, testicular and/or paratesticular masses were given different diagnoses such as inguinal hernia, hydrocele (Zaslau, S. *et al* 2005), especially if the scrotal ultrasonography was not performed. We report a boy with paratesticular embryonal rhabdomyosarcoma on a ectopic testis, who underwent orchiopexy and a surgical resection of the mass. He survives without recurrences for nine months after management with multidisciplinary approach.

CASE PRESENTATION

A 16-year-old boy with a medical history of left testicular ectopia treated in 2014 with orchiopexy. Recently he presented a 2- month history of testicular swelling and a painless mass in the left scrotum. In the scrotal ultrasound the testicles were normal size measuring 44x30x27mm on the right, i.e. a volume of 18ml and reduced on the left with a high testicle located in the scrotum measuring 34x23x18mm, i.e. a volume of 7ml. The ultrasound also reported the presence of a left intrascrotal tissue mass inside and below the left testis, with fairly well defined contours measuring 40x25mm in major axes, it is discreetly heterogeneous dotted with fine calcifications, it is vascularized in the color flow Doppler. It is associated with a moderate dilation of the venous vascular structures along the spermatic cord and intascrotally with veins measuring 2.1mm (Fig. 1). A thoraco-abdomino-pelvic Computed tomography (CT) did not reveal any secondary localization.

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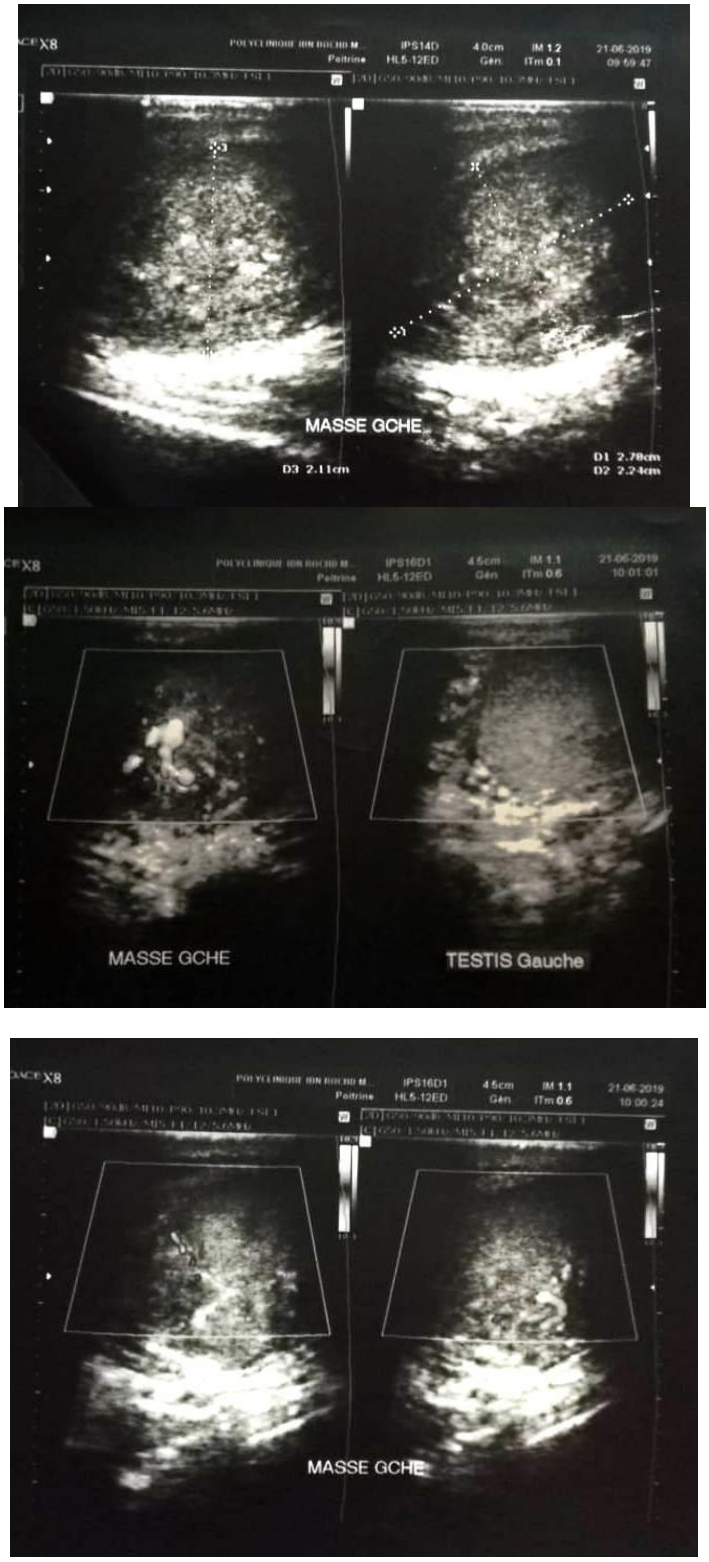

Figure 1: left intrascrotal tissue mass inside and below the left testis, with fairly well defined contours measuring 40x25mm in major axes, it is discreetly heterogeneous dotted with fine calcifications

The patient underwent a surgical resection of the mass, without orchiectomy. On histopatho-logical examination, the removed solid mass was completely encapsulated and measured nearly 5×4×3 cm. It is a tumor proliferation organized in sheets and areas and bundles crisscrossed in places without formation of glands, papillae or horny cysts. it is made up of cells with elongated spindle-shaped nuclei presenting moderate atypia with some mitosis. the stroma is quite abundant and partially fibrous and dotted with mono and polynucleate inflammatory elements with thin-walled vessels without calcifications or foci of necrosis. this tumor proliferation infiltrates and dissociates the sections of epididymal ducts in the vicinity and with a

regular border without vascular emboli or presence of individualized intact testicular pulp. The tumor cells were positive for anti-desmin antibody while negative for Anti-Myogenine antibody on immunohistochemistry. The patient was followed up postoperatively with chemotherapy. Vincristine, doxorubicin and cyclophos-phamide (VAC) regimen was used. The patient is disease-free in follow-up nine months after completion of chemotherapy.

DISCUSSION

Childhood Rhabdomyosarcoma is the most common soft tissue sarcoma with an annual incidence of four to seven cases per million. Paratesticular RMS accounts for 7% of all RMS and is the most common soft tissue sarcoma in childhood in this subsite. It presents with painless scrotal mass and arises from spermatic cord, epididymis and testicular envelopes. It becomes easier to identify for patient as well as doctors because of its superficial location (Sultan, I. *et al* 2009; & Kumar, R. *et al* 2013). They are amenable for complete surgical resection. Overall five year survival rate is more than 80%. Complete physical examination of patient and history are the most important way to detect it in initial stage. MRI is the first and best imaging modality which provides three dimensional images for planning of surgical extent and radiation. CT scan of chest, abdomen and pelvis is done to rule out possibility of metastasis. Bone scan of entire skeleton is undertaken to see possibility of tumour mets. PET-CT is done in case of ambiguous findings on CT and MRI as well as to see response of treatment in follow up stage. Bone marrow aspiration is also done for evaluation of bone involvement (bone marrow involvement is reported in less than 2% cases). It has been observed that approximately 20% newly diagnosed patients will have one or two sites of metastasis. Hence, above modalities of investigations are essential. Normally tumour is diagnosed histopathologically on specimen collected after percutaneous or incisional or open excisional biopsy. Tissue for biopsy is sampled by three routes- percutaneous (with diagnostic yield of 90%), incisional (diagnostic yield of 100%) and excisional biopsy(with curative intent).

Three different types of RMS have been categorized based on Histo morphology of tumour cells under microscope- Embryonal (65-70%, seen in two third childhood RMS with botryoid and spindle variant), Alveolar type (20-25%) and undifferentiated/Pleomorphic type (5-10% with classic pleomorphic, round cell and spindle cell pattern). Rhabdomyoblasts is the characteristic cell but is not essential for diagnosis (Parham, D. M., & Ellison, D. A. 2006). In Embryonal RMS, small cells with hyperchromatic nuclei, minimal cytoplasm as well as cells with rims of eosinophilic cytoplasm and spindle cells with cytoplasmic tails and variable cross striations in myxoid or collagenous stroma is seen. Tumour markers like alpha feto protein, beta HCG and carcinoembryonic antigen all are within normal range in RMS. RMS can be included in differential diagnosis of fibrosarcoma, leiomyosarcoma and liposarcoma—however these entities are more common in adults. Other common entities which can be put in differential diagnosis are-infantile fibromatosis, neuromuscular hamartoma of soft tissue and rhabdomyoma among benign entities and malignant mesothelioma, melanotic neuroectoodermal tumour and desmoplastic small round cell tumour among malignant tumours in this location. All can be diagnosed morphologically and with help of IHC markers.

Multimodality approach with surgery, chemotherapy and radiation has improved prognosis in this tumour. Factors potentially affecting prognosis are local invasiveness, tumour size, morphological appearance of tumour, nodal involvement, surgical resection and age of the patient. Risk stratification into Low-A, Low- B, Intermediate, High and Intergroup Rhabdomyosarcoma group are basically used to assign appropriate treatment to patient (Qualman, S. *et al* 2008). Genitourinary location and less than 5 cm size of tumour are favourable parameters to determine prognosis. Retroperitoneal lymph node dissection has been used as staging procedure. Lmphangioscintigraphy for evaluation of nodal status prior to surgery is under study for its usefulness. On IHC, Tumour cells are immunoreactive for Desmin, Vimentin, Actin and Myogenin and negative for Pan CK, S100 and CD99. Immunoreactivity for Myogenin virtually clinches diagnosis. However, in rare cases over-expression of insulin like growth factor-2 needs to be established for diagnosis on RT-PCR. Reciprocal translocation between PAX and FKHR creates a hybrid oncogene which results in an "overdosage" of a "growth promoting gene" responsible for insulin-like growth factor Type II (IGF-II), that is located on chromosome 11 (Barr, F. G. *et al* 2006). Over expression of IGF-II leads to unrestrained growth and proliferation of muscle in RMS.

RMS can spread locally, regionally (lymph nodes) and distantly (through blood). Most common sites of distant metastasis are lung, bone and bone marrow. Brain, liver and spleen are uncommon sites for distant metastasis. Metastatic disease with bone marrow involvement and aggressive behavior is more common in adult RMS. Prognosis depends on age of patient, site, size, morphology, residual tumour left after surgery and spread to other parts of body. Child above ten years of age will have high chances of nodal metastasis. However, chances of distant metastasis are one in five. Preferred route is through blood stream.

Paratesticular localization of RMS is rare and develops from mesenchymal tissue of the spermatic cord, epididymis and testicular envelope. Bimodal peaks of incidence have been observed- one at four

years of age and another peak at 18 y of age. Tumour mainly presents as hard painless inguinoscrotal mass and very rarely invades scrotal skin. Rarely mass may evolve near external inguinal ring away from scrotal contents.

RMS is chemo sensitive and most common protocol is VAC-Actinomycin-D, Vincristine and Cyclophosphamide. Radical orchidectomy, hemiscrotatectomy and high inguinal cord dissection with inguinal lymph node dissection is main surgical treatment. Radiotherapy is also helpful to treat residual microscopic foci of tumour.

CONCLUSION

Paratesticular RMS is a rare tumour with multimodality approach in diagnosis as well as treatment. Localized form has good prognosis and metastatic disease has poor prognosis. Retroperitoneal lymph node assessment for microscopic foci by Lmphangioscintigraphy is under evaluation. Risk stratification has been done to achieve maximum benefits. Strict follow up for long term is the rule in all cases.

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