*ejpmr, 2020,7(2), 65-68*

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Review Article

ISSN 2394-3211

EJPMR

# METASTATIC RECURRENCE OF PARA TESTICULAR RHABDOMYOSARCOMA: ABOUT A CASE WITH REVIEW OF THE LITERATURE

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**Article Received on 01/12/2019** **Article Revised on 21/12/2019** **Article Accepted on 11/01/2020**

## ABSTRACT

Para-testicular rhabdomyosarcoma (RMS) is a rare mesenchymal tumor that can develop from the spermatic cord, epididymis or testicular vaginal. It constitutes 7% of cases of rhabdmyosarome The diagnosis requires histological confirmation. The treatment must be multimodal in view of the aggressiveness of these tumors, based on surgery, chemotherapy and radiotherapy. We report the observation of a paratesticular rhabdomyosarcoma case in metastatic recurrence. In the light of these clinical data we will discuss the diagnostic and therapeutic modalities.

**KEYWORDS:** Para-testicular rhabdomyosarcoma (RMS) is a rare mesenchymal modalities.

## INTRODUCTION

Paratesticular rhabdomyosarcoma is a rare mesenchymal malignant tumor in children. It represents around 0.5% of all childhood and adolescent cancers.[1] The diagnosis is made in front of a large painless scrotal bursa and retained by the anatomopathological study of the orchidectomy piece.[2] About 25% of patients have distant metastases at the time of diagnosis.[3] Early diagnosis and treatment is essential to improve the prognosis.

## OBSERVATION

Child Z.Z. 12-year-old from a non-consanguineous marriage. Without any particular pathological history.

The symptomatology goes back to 2 years, by the appearance of a large painless hard left bursa, gradually increasing in volume, evolving in an apyretic context and deterioration of the general state.

Clinical examination revealed a painless left scrotal mass on palpation which extended towards the path of the spermatic cord, without inflammatory signs; with a tissue-like appearance on transillumination.

Scrotal ultrasound shows a left testicle crushed by a heterogeneous epididymal tissue mass vascularized with doppler measuring 4.5 * 2 cm. Computed tomography (CT) thoraco-abdomino-pelvic confirmed the existence of a mass occupying the entire lower part of the bursa, pushing the testicle up and forward without lesions at a distance. Testicular tumor markers (alpha-fetoprotein, beta-hCG, lactic dehydrogenase) were negative.

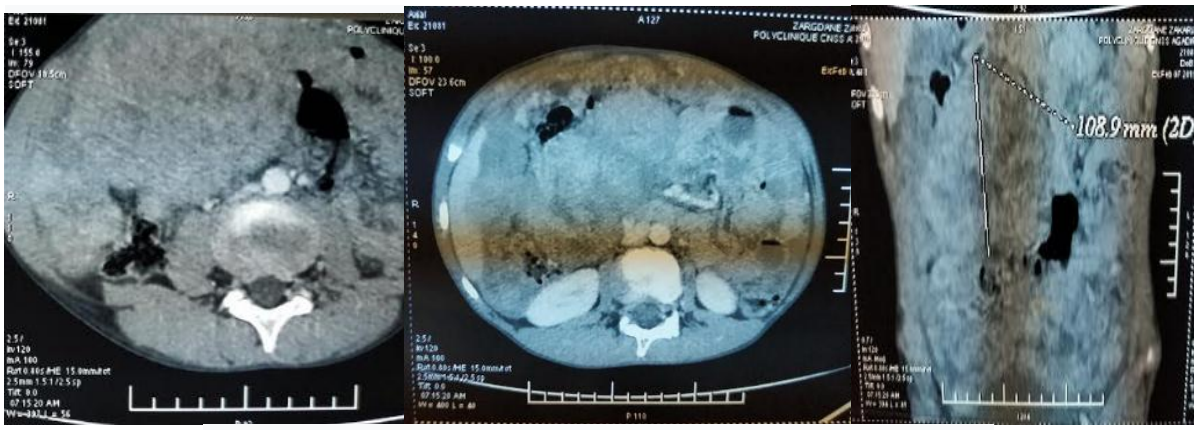
An orchiepididymectomy was performed inguinally after clamping the spermatic cord; revealing to the anatomopathological study: A firm, encapsulated lobulated paratesticular tumor mass well limited to the macroscopic study; with the microscopic aspect of a sarcomatous process with superimposed monomorphic cellular component expressing anti-desmin and anti-myogenin under study immunohistochemical, allowing to retain the appearance of an embryonic paratesticular rhabdomyosarcoma.

Chemotherapy according to the rhabdomyosarcoma protocol (RMS) 2005 subgroup A (Oncovin 1.1 mg / week for 4 weeks, Actinomycin 1095 mg 2 / month) was administered for 3 cycles at 1 month intervals. This treatment was well tolerated by the patient.

The evolution was marked by the installation of abdominal pain with CT monitoring TAP: Round intraperitoneal tissue formation opposite the hepatic dome measuring 25 * 42 * 46 mm with a voluminous suspected hepatic formation measuring 92 * 104mm, with the presence of nodules of peritoneal carcinosis and bilateral cervical lymphadenopathy measuring 14 * 9 mm for the largest (Figure 1).

An exploratory Laparoscopy with biopsy was carried out objectifying to the pathological study a malignant tumor proliferation with round cells expressing anti-desmin and anti myogenin retaining the metastatic recurrence of an embryonic rhabdomyosarcoma.

The child received 4 IVADO protocol courses (ifosfamide, vincristine, actinimycin, oncovin) with control TAP CT (Figure 2) regression estimated at 70% of abdominal mass and carcinosis nodules. regular monitoring by TAP is planned.




**A** **B** **C**

**Figure 1: TDM TAP: axial and fronatal sections: Round tissue formation opposite the hepatic dome measuring 25 * 42 * 46 mm, bulky formation under suspect hepatic measuring 92 * 104mm (c), presence of nodules of peritoneal carcinosis (a).**

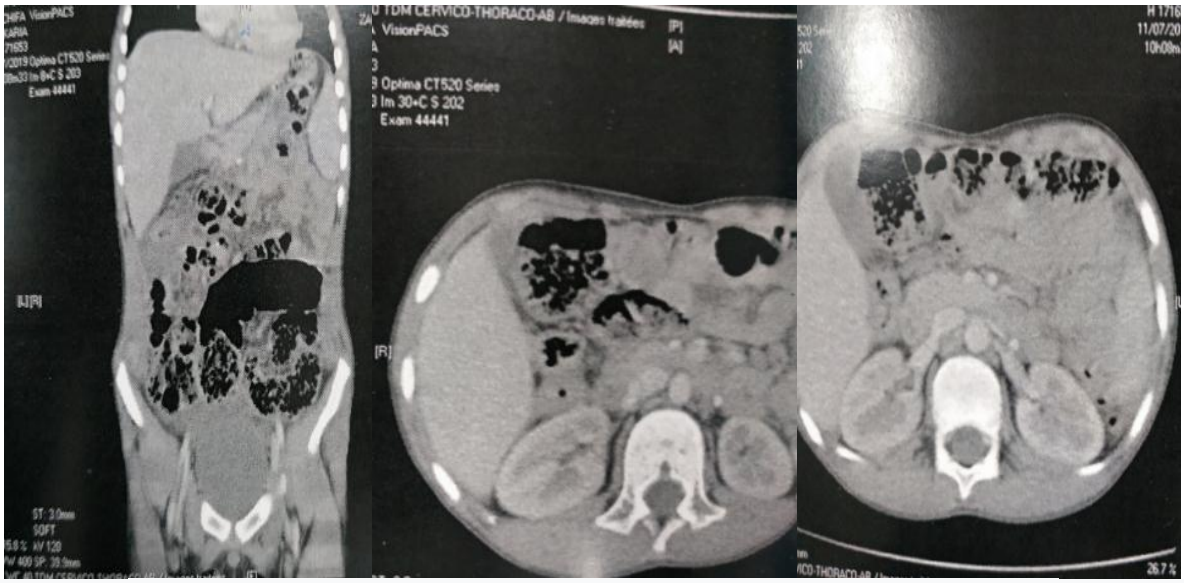



**Figure 2: Regression estimated at 70% of abdominal mass and carcinosis nodules.**

## DISCUSSION

Paratesticular rabdomyosarcomas are rare malignant tumors which represent 7% of all localizations[4,5,6]; electively affecting children and adolescents with Two peak ages 4 years and 16 years.[4] There are 3 histological types: embryonic most frequently encountered with good prognosis if localized, pleomorphic and alveolar.[7]

Clinically, paratesticular RMS is manifested by a hard, painless scrotal mass, rapid devolution, opaque to transilumination. Local extension is very early and dissemination at a distance is by lymphatic and blood route.[4] The presence of inguinal lymph nodes on clinical examination favors scrotal invasion.[8]

Bilateral scrotal ultrasound objective a mass of tissue density, heterogeneous, intra scrotal, developed at the expense of testicular envelopes, hyper vascularized with doppler.[9]

Thoraco-abdomino-pelvic computed tomography (TAP) with injection of contrast agent is recommended to assess the regional and remote extension of paratestecular RMS; by specifying the invasion of the pelvic and lumboaortic glands and of possible hepatic and pulmonary metastases; while Pelvic Magnetic Resonance Imaging (MRI) is recommended in case of doubtful diagnosis allowing to visualize a hypointense mass in T1, and heterogeneous in T2; separated from the testicle.[6,10,11]

Tumor markers, including alpha-fetoprotein, beta-human chorionic gonadotropin and carcinoembryonic antigens are systematically recommended before any treatment. They are generally normal in RMS. This was the case with our patient.

The diagnosis of certainty is based on a histological examination of the inguinal orchidectomy part.[12] Histologically; The characteristic cells are rhabdomyoblasts; of varying sizes and shapes; if absent, the immunohistochemical complement is essential for the diagnosis objectifying a diffuse positivity to pancytokeratin (AE1 / AE3), to anti desmin and a focal positivity to myogenin.[13]

The classic TNM classification is used, but also others, such as the classification of the "Intergroup Rhabdomyosarcoma Study" or IRS which takes operability into account.[14]:
***Group I***: localized tumor, complete microscopic ablation, confined to the muscle or the original organ without lymph node invasion.
***Group II:*** a- total gross removal but persistence of microscopic tumor tissue; b- regional disease (going beyond the original muscle or organ) but completely resected or extension to the nodes that have been completely resected.
***Group III***: incomplete ablation with macroscopic tumor persistence.
***Group IV***: distant metastases at diagnosis.

The therapeutic modalities of paratesticular RMS are based on surgery, radiotherapy and chemotherapy.[15] RMS are extremely chemosensitive tumors; Most children receive preoperative chemotherapy, followed by a reassessment at the end of treatment, to specify the type of excision surgery, while in paratesticular forms, surgical treatment precedes chemotherapy.[8] provides local control of the tumor; The approach is inguinal; and lumboaortic or retroperitoneal lymph node dissection is of no interest in first intention.[16,17]

Multi-drug therapy is indicated to provide local and remote control by preventing the appearance of metastases. The most used chemotherapy protocols consist of vincristine, actinomycin D and cyclophosphamide, etoposide and ifosfamide, several cures should be spread over 5 days and spaced 2 to 4 weeks apart.[6]

Radiation therapy is only recommended in the event of an incomplete response to chemotherapy and in the event of incomplete surgery, at a dose of 50 to 60 Gy, in conventional fractionation, spread over 5 to 6 weeks.[18]

The therapeutic decision depends on the prognostic group according to the IRS classification. In so-called localized group I: adjuvant chemotherapy is systematic. Radiation therapy after retro-peritoneal lymph node dissection is recommended; in advanced forms (group II and III). While in metastatic forms (group IV); the therapeutic strategy must be aggressive, multidrug therapy without lymph node dissection with metastasis removal if it is extirpable, is supplemented by radiotherapy directed at the retroperitoneal lymph nodes and metastases.[6,19]

The prognosis of paratesticular rhabdomyosarcoma depends on several factors which are essentially the tumor stage, the histological type; the response to treatment, the quality of the excision, as well as the patient's age.[6,20] The localized nature of the tumor, the embryonic type and the young age of less than 10 years are considered to be elements of good prognosis.[20,21] The therapeutic development has transformed the prognosis of these tumors, and has increased overall survival; the 5-year survival rate varies from 80[16,21] to 88.5%[19] in localized forms, to 22.2%[19] in metastatic forms. RMS operations must be monitored regularly because the patient is not immune to recurrence.[22]

For a long time, local recurrences have been so frequent that they have left the problem of distant dissemination in the background. This is the case of our patient who presents with a metastatic recurrence.

## CONCLUSION

Early diagnosis and precise extension assessment are necessary; in front of the paratesticular RMS; in order to adapt the therapeutic strategy. The advent of therapeutic modalities has improved the survival of this aggressive tumor; but monitoring must be regular in order to detect local or distant recurrence early.

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