Case Report: Adult Retroperitoneal Rhabdomyosarcoma


ABSTRACT

Rhabdomyosarcoma (RMS) is the most common malignant mesenchymal tumor in children and adolescents. In this article we report a rare case of RMS considering the age of 37 years and its retroperitoneal location. The symptomatology was characterized by abdominal pain and alteration of the general condition. The diagnosis was confirmed after a scan-guided biopsy followed by an immunohistochemical study. Its management must be discussed in order to propose the appropriate therapy which was a chemotherapy in our patient with an unfavorable evolution.

Keywords: Adults, rhabdomyosarcoma, retroperitoneal, therapeutic.

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I. INTRODUCTION

Retroperitoneal rhabdomyosarcoma in adults is a rare tumor not only in terms of histological type but also in terms of location. The diagnosis is usually made belatedly and is most often revealed by abdominal pain or mass. Ultrasound and abdominal scanner are the most commonly used means of imagery for the identification of the mass.

The rarity of the tumor requires a multidisciplinary diagnostic and therapeutic management in a dedicated center in order to improve the prognosis which relies on several criteria (surgical, anatomo-pathological, ...).

Carcinological surgery is the core of the medical care. Radiotherapy and chemotherapy are still indicated for neo-adjuvant treatment.

II. MEDICAL OBSERVATION

The subject was an adult patient of 50 years old, a chronic smoker of 50 packs/year who was in the process of weaning, and who had been experiencing pain in the left hypochondrium for 2 years, associated with uncalculated weight loss followed by abdominal distension, without any urinary signs, in particular hematuria.

An abdominal ultrasound and a scanner were conducted, revealing an inter-spleno-rena1 mass measuring 245 x 2020 x 300 mm, which could originate from retroperitoneal liposarcomatous or GIST.

A fibroscopy and colonoscopy which showed no abnormalities were indicated to rule out GIST.

A biopsy of the mass scanned-guided was performed with a morphological and immunohistochemical aspect in favor of an embryonal rhabdomyosarcoma grade II according to the FNCLCC.

The presence of a proliferation of spindle cells as well as cells with rhabdomyoblastic differentiation was observed at the anapath, at the IHC: focal expression of desmin and myogenin; CKit, AML, CD34, CKAЕ1/Е3, Mdm2 were negative. The search for Dog1 was found to be negative in order to eliminate a GIST.

An extension workup consisting of a thoracic-abdominal-pelvic CT scan and a bone scintigraphy did not reveal any secondary locations with a normal biological workup (CBC, g-GT, renal workup).

III. DISCUSSION

Retroperitoneal sarcomas are very rare tumors with an incidence of 0.5 to 1 case per 100,000 inhabitants [1]. Retroperitoneal rhabdomyosarcoma is an even less recurrent entity of retroperitoneal sarcomas after leomyosarcoma, liposarcoma, and malignant histiocytosarcoma [2]. The different anatomo-pathological forms are classified according to FNCLCC (Fédération Nationale des Centres de Lutte Contre le Cancer) [2].

IV. DIAGNOSIS

As with all retroperitoneal tumors, retroperitoneal rhabdomyosarcoma manifests itself delayed by signs of compression of the surrounding organs responsible for pain, abdominal mass, abdominal discomfort related to the volume of the tumor mass, or alteration of the general condition [3]. The main symptom in our patient was pain.

The diagnosis of retroperitoneal sarcoma is evoked on abdominal-pelvic CT scan with contrast injection, which usually reveals a tissue mass and also allows analysis of the tumor relationships, orientation of the nature and guidance of a biopsy. MRI is a good alternative in case of allergy to contrast medium or impaired renal function [4]. The imaging in our patient suspected the diagnosis of Rhabdomyosarcoma or GIST.

V. ANAPATH

In order to validate the diagnosis, a biopsy with anatomical-pathological and immunohistochemical study is performed under ultrasound or scanographic guidance [5]. The latest studies have demonstrated a negligible risk of tumor seeding in the biopsy pathway, however, the biopsy must be performed by a posterior or lateral approach and never by a transperitoneal approach. Similarly, biopsies by laparotomy or open laparoscopy should be avoided [6].

Excisional surgery with anapath + IHC study can be proposed if tumor is operable but without knowledge of the ideal technique for the histological type.

RMS presents as a proliferation of poorly differentiated round or spindle-shaped cells with a striated muscle differentiation line. Immunohistochemistry is of great help, looking for labelling of rhabdomyoblasts by myosin, actin, desmin, myoglobin and Myo-D [7].

In IRS IV, frequencies of the various tumor subtypes were embryonal, 70%; alveolar, 20%; undifferentiated, 4%; and others, 6% [8] with poor prognosis for adult patient.

Reference [9] demonstrated that the embryonal and pleomorphic subtypes had a better prognosis compared to the alveolar subtype although the results were not statistically significant; however, an Indian study concluded that there was no prognostic difference between these different histological subgroups [10].

Our patient underwent a scanographic-guided biopsy by lateral approach with anapath + immunohistochemistry: absence of expression of Dog 1, morphological aspect and IHC in favour of an embryonic Rhabdomyosarcoma grade II according to the FNCLCC.
VI. ASSESSMENT OF EXTENSION

The distant extension concerns mainly the lung, bone marrow, liver and bone. The workup of extension is done by thoracoabdomino-pelvic CT, bone scan and bone marrow aspiration and biopsy in patients with RMS [11]. Bone marrow involvement is present in 2% of patients with isolated RMS [11].

Our patient underwent a thoracic CT scan (since the abdominal-pelvic CT scan had already been performed) and a bone scan, which did not reveal any secondary sites.

VII. CLASSIFICATION

Several classifications are involved in the treatment of the patient. A first classification system based essentially on the location of the tumor and the tumor status will allow to define 4 stages.

A staging system based on the quality of the initial surgical resection, suggested by the IRS group, is still insufficient since only the surgical aspect is taken into account.

The "Fifth Intergroup Rhabdomyosarcoma Study" (IRS-V) has led to the elaboration of a classification that takes into account several prognostic factors, in particular the location of the tumor, the histological subtype, the distant tumor extension and the quality of the surgical resection...; this classification is essentially used in the choice of the chemotherapy protocol.

VIII. TREATMENT

A. Surgery

Key element in the management of embryonic RMS. Together with radiotherapy, it ensures local control. The quality of the surgery depends mainly on the margins (complete resection, microscopic residue or macroscopic residue) [12].

It can be performed:
1) Straightforward: must be complete with healthy margins respecting the shape and function of the organ
2) Surgical revision: in case of incomplete resection or surgical biopsy, surgical revision may be indicated if the surgeon is convinced of the possibility of a R0 resection [13].

Following chemotherapy: different indications; decrease of tumor volume, better local control for tumors with partial response to chemotherapy, decrease of the dose of adjuvant radiotherapy [14].

B. Radiotherapy

Indispensable component in the multimodal treatment of RMS; in combination with surgery, it allows better local control with, DMFS and OS. The recommended dose is 50 Gy initially followed by a 10 to 20 Gy boost to the tumor bed [15].

Indicated in case of non-operable embryonal RMS, microscopic residue or in case of lymphatic invasion.

For R0 patients, radiotherapy in embryonal RMS improves progression-free survival but has no impact on overall survival, unlike in alveolar RMS where radiotherapy is indicated even in cases of complete surgery [15].

It can be used as a neoadjuvant to surgery to avoid intraperitoneal dissemination during surgery [16].

C. Chemotherapy

Vincristine, dactinomycin, ifosfamide or cyclophosphamide represent the drugs mainly used for rhabdomyosarcoma.

Although the response to the protocols is less in adults, the protocols used in children can give good results in adults, especially on recurrence-free Survival metastasis [17].

MSKCC and MDA found an impact of pediatric chemotherapy protocols on overall survival for embryonal and alveolar rhabdomyosarcoma, as well as the Ferrari study...
which confirms this hypothesis, while for pleomorphic the interest of chemotherapy cannot be identified [18].

In contrast, the [19] concluded that there was no benefit from chemotherapy.

IX. CONCLUSION

Retroperitoneal rheumatoid arthritis is a rare tumor in adults. It comprises different histological types with different prognosis. The management of RMS in adults must be discussed given the polymorphism of this pathology. Multimodal treatment remains the reference treatment, but RMS remains an aggressive tumor with a high mortality rate.

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CONFLICT OF INTEREST

Authors declare that they do not have any conflict of interest.

REFERENCES