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(CASE REPORT)

Giant intra-abdominal and retroperitoneal desmoid tumor

Othman CHAMA [1, 2, *], Rhyan OUADDANE ALAMI [1, 2], Amine BOUCHTIB [1, 2], Mustapha Ahsaini [1, 2], Soufiane Mellas [1, 2], Jalal Eddine El Ammari [1, 2], Mohammed Fadl Tazi [1, 2], Mohammed Jamal El Fassi [1, 2] and Moulay Hasan Farih [1, 2]

[1] *Department of Urology, Hassan II University Hospital, Fez, Morocco.*
[2] *Faculty of Medicine, Pharmacy and Dentistry of Fez, Sidi Mohammed Ben Abdellah University, Morocco.*

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Abstract

Desmoid tumors are extremely rare. They represent 3% of all soft tissue tumors. Giant desmoid tumors are rarely reported in the literature and are a challenge in their therapeutic management.

The radiological evaluation of extension allowed to delimit its extension in depth and its limits of exeresis. In our case, the tumor was surgically resected with simple postoperative care.

This case illustrates the difficulties of managing this entity given its large size.

Keywords: Giant tumor; Desmoid; Rare pathology; Surgery; Retroperitoneal tumor

1. Case report

Patient 34 years old, single, chronic smoker for 14 years, never operated, hospitalized in our training for PEC of an intra abdominal mass evolving for 6 months, all evolving in a context of weight loss.

The interrogation did not objectify any pain, no fever or hematuria apart from the increase of the abdominal volume and the weight loss quantified at 20 Kg.

The clinical examination found an abdominal mass on palpation, taking the whole abdomen, no lumbar contact or collateral venous circulation.

On the biological level, a normocytic hypochromic anemia estimated at 8.5 mg/dl was observed, no renal insufficiency with a slight inflammatory syndrome.

The patient underwent a CT scan which showed a large solid tumor mass (22*17*15cm), well limited and partly merging with the left kidney, no secondary lesions at a distance.

The patient underwent a biopsy of the lesion which showed a non-epithelial tumor proliferation, not very proliferative, evoking a desmoid tumor first.

he benefited from a surgical removal of the mass and nephrectomy by xipho pubic approach requiring a transfusion by 5 CG in total.

* Corresponding author: Othman CHAMA

The postoperative follow-up was simple, resumption of transit 4 days later.

He was discharged on the 8th day of the postoperative period.

Informed consent: the patient provided his consent.

2. Discussion

Desmoid tumor is defined by the World Health Organization as a clonal fibroblastic proliferation that occurs in deep soft tissue and is characterized by an inability to metastasize although it may be multifocal in the same limb or part of the body [1,2]. It is most common in women between the ages of 15 and 60, with a peak between the ages of 30 and 40 [3,4].

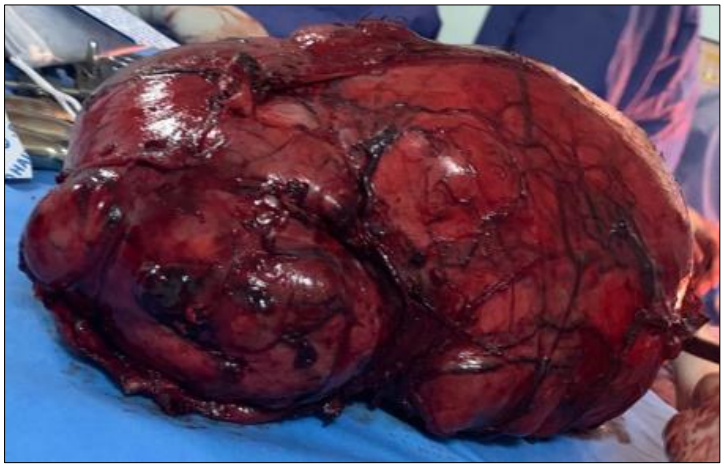

Figure 1 Post operative part

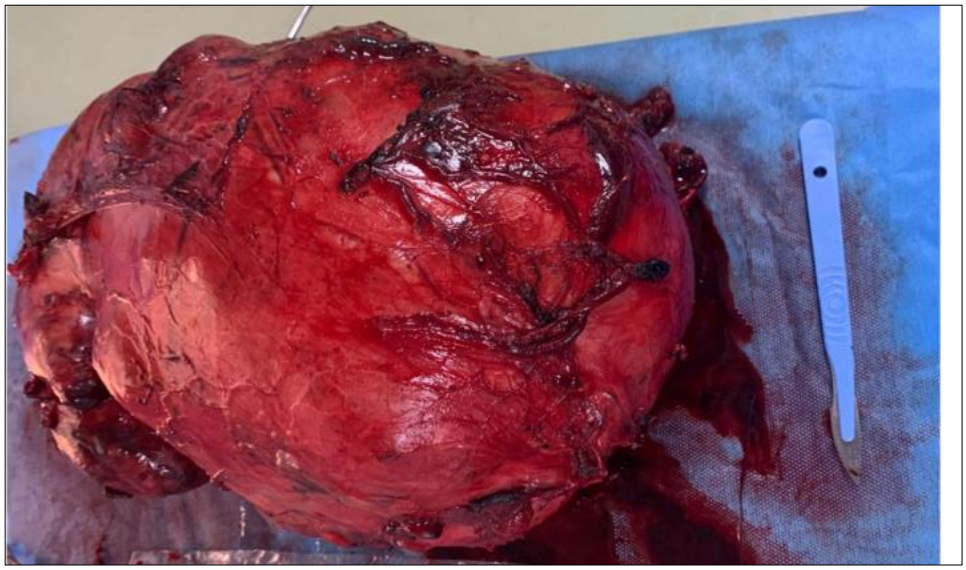

Figure 2 Post operative part

Two important categories of desmoid tumors are recognized: sporadic tumors, which account for 85-90% of cases and are associated with a mutation in the coding pathway for betacatenin CTNNB1, and tumors associated with the APC gene mutation of familial adenomatous polyposis (FAP), mainly affecting the abdomen [3,5]. The clinical picture can present several aspects depending on the location and can be very aggressive with increased growth and mass effect as in our patient's case [6]. MRI is the reference examination for diagnosis, staging and follow-up. It allows to study the tumor extension and to determine the surgical resection plan to be achieved. On T1-weighted images, the tumor appears

hypointense or isointense in relation to the muscle and hyperintense on T2 with enhancement after gadolinium injection and persistence of hypointense bounds [2,3]. The diagnosis of certainty is histological on percutaneous or operative biopsy showing a proliferation of uniform spindle cells resembling myofibroblasts, in a stroma of abundant collagen and vascular network.Surgical treatment remains a challenge for surgeons especially for giant tumors. Due to the infiltrative growth pattern, the extent of resection required to achieve negative margins could often result in significant functional and cosmetic alterations [4]. This resection confers a good local control rate of about 80% at 5 years [2].

Chemotherapy remains an option in cases of unresectable or advanced symptomatic disease [3-5]. Targeted therapies (Imatinib) confer a high rate of disease stabilization in 60-80% of cases [4], hormonal therapy (Tamoxifen) by its anti-estrogenic action is associated with clinical benefit in about 30% of cases [5] and non-steroidal anti-inflammatory drugs (NSAIDs) by their ability to regulate the ß-catenin pathway by inhibiting cyclooxygenase-2 or prostaglandins [4]. Locoregional treatment with radiotherapy or cryotherapy may reduce the risk of recurrence after incomplete surgical resection [2,3,5,7].

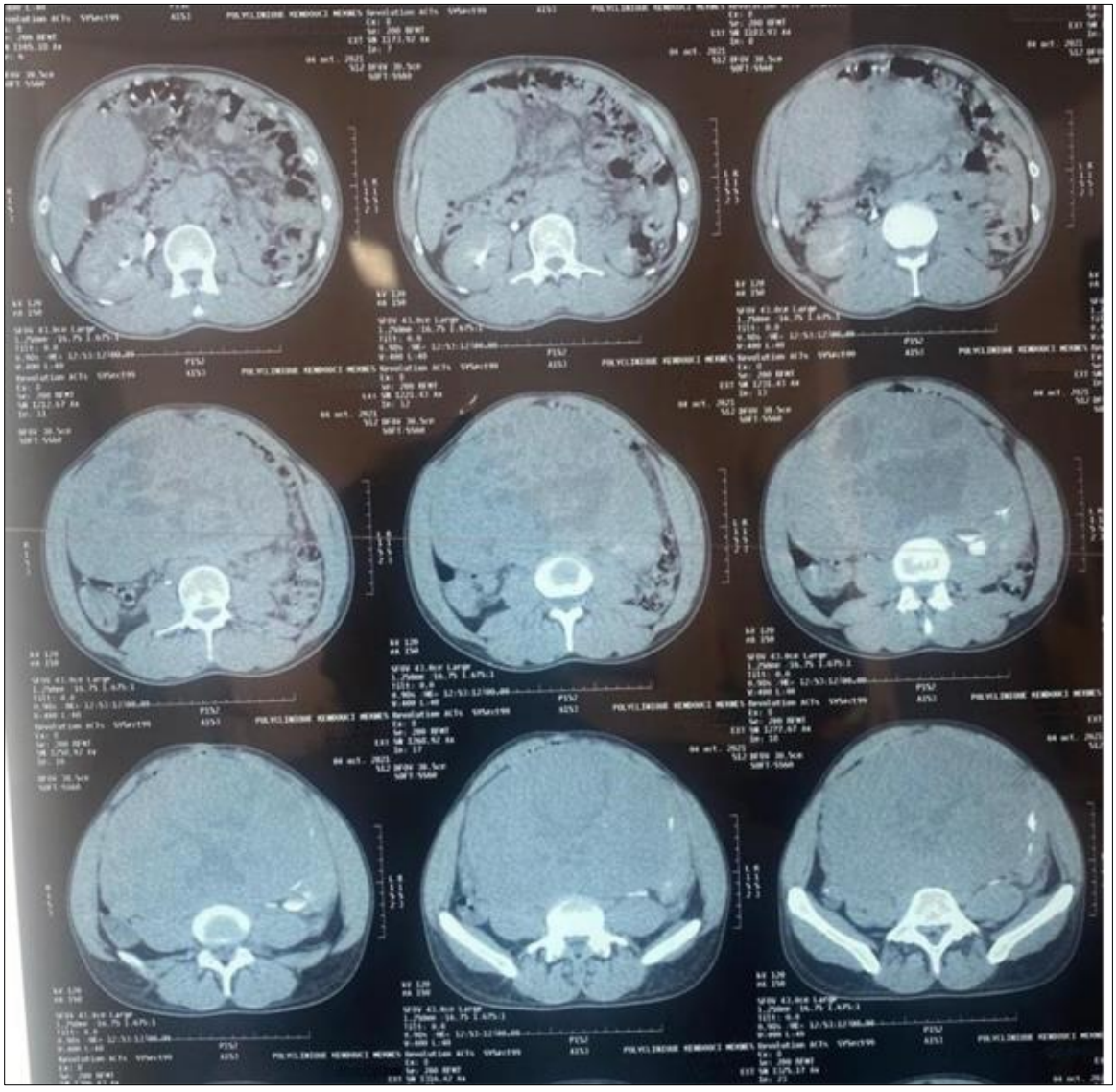

Figure 3 Scannographic section on late time « mass which depends on the left kidney »

The initial strategy is to recommend active surveillance for patients with desmoid tumors for a period of 1 to 2 years by clinical examination and monthly MRI during the first months and then at an interval of 3 to 6 months, looking for tumor progression or increase in symptoms that will justify active treatment [3,8]. Some situations make surgery unavoidable such as complications (occlusions, perforation, hemorrhage) or major aesthetic problems, as in the case of our patient. Surveillance after surgical resection is essentially clinical and radiological by MRI or CT scan [9].

The local recurrence rates depend mainly on the resection margins.

3. Conclusion

Giant desmoid tumors are aggressive fibromatoses with a high potential for recurrence. It should be evoked in front of any abdominal parietal tissue mass and confirmed histologically. Management is currently based on a strategy of initial active surveillance and then, depending on the signs of complications or progression, removal of the mass may be proposed.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

no conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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