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Radiology

Recurrent Desmoid Tumor of the Abdominal Wall

Abdelaziz Hebbezni[1*], Houda El Madkouri[1], Badr Boutakioute[1], Meriem Ouali Idrissi[1], Najat Cherif Idrissi Ganouni[1]

[1]Radiology Department, Cadi Ayyad University, University Hospital Mohammed VI of Marrakech, Morocco

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***Corresponding author:** Abdelaziz Hebbezni

Abstract **Case Report**

Desmoid tumors or aggressive fibromatosis are monoclonal fibroblast proliferations. They are characterized by an invasive infiltrating capacity, a very high local recurrence rate but without metastatic potential. Imaging plays an essential role in diagnosis and therapeutic management is multidisciplinary. We report the case of a 60-year-old patient presenting with an abdominal mass of the left lateral wall gradually increasing in size and whose diagnosis of desmoid tumor of the abdominal wall was retained.

Keywords: Desmoid tumors, diagnosis, abdominal mass, desmoid tumor.

INTRODUCTION

Desmoid tumors, also called aggressive fibromatosis, which are part of deep fibromatosis [1]. These are monomorphic proliferations of fibroblast tissues, infiltrating, non-metastasizing but recurrent and which develop from the muscle sheaths, fascia and aponeuroses [1, 2].

They are also desmoid tumors and represent less than 0.03% of all tumors and around 3% of fibrous tumors [3].

These tumors occur sporadically or as part of a familial adenomatous polyposis (FAP) where they represent one of the manifestations of the lesional association of Gardner syndrome [4].

The peak onset of desmoid tumors is between 20 and 40 years; 80% of these tumors occur before the age of 40. The most common site is the anterior abdominal wall, with an incidence of 50% [5].

The diagnosis is difficult and imaging makes it possible to evoke the diagnosis of these tumors, to orient the therapeutic attitude and the follow-up. The treatment is ideally based on surgery when possible and the clinical course is marked by frequent recurrences [6].

PATIENT AND OBSERVATION

We report the case of a 60-year-old patient, operated on in 2004 for an undocumented left lateral wall tumor, who consults for an abdominal mass gradually increasing in size, painless without transit disorders, all evolving in a context of apyrexia and general condition conservation.

On clinical examination, the patient was conscious, hemodynamically and respiratory stable.

Abdominal examination revealed an operative scar on the left flank, a painless supple abdomen. A swelling of the left lateral abdominal wall rounded, painless and mobile compared to the deep plane of 4cm long axis. The hernial orifices were free and the rectal examination revealed an empty rectal bulb. The biological assessment was unremarkable.

An abdominal ultrasound made objectified a rounded mass, homogeneous hypoechoic of regular contours at the expense of the muscles of the left lateral abdominal wall poorly vascularized by color Doppler measuring 45 mm long axis.

To better characterize this mass and study its relationships, further exploration by an abdominal CT scan with injection of PDC is carried out and showed a large parietal tissue mass deep in the left transverse muscle with a well-defined rounded shape, regular contours, spontaneously isodense neighboring muscles (Figure 1), intensely and heterogeneously enhanced in arterial phase (Figures 2) and homogenizing in portal phase (Figures 3 a and b) with washing in late phase (Figure 4 a and b), measuring 43x38 mm, suggesting a fibromatous tumor of the desmoid type.

The patient was operated, the procedure consisted of a complete resection of the tumor. The patient is put on antibiotics, analgesics, rehydration with change of dressing twice a day. The postoperative consequences were simple. His exit was authorized on the 3rd day.

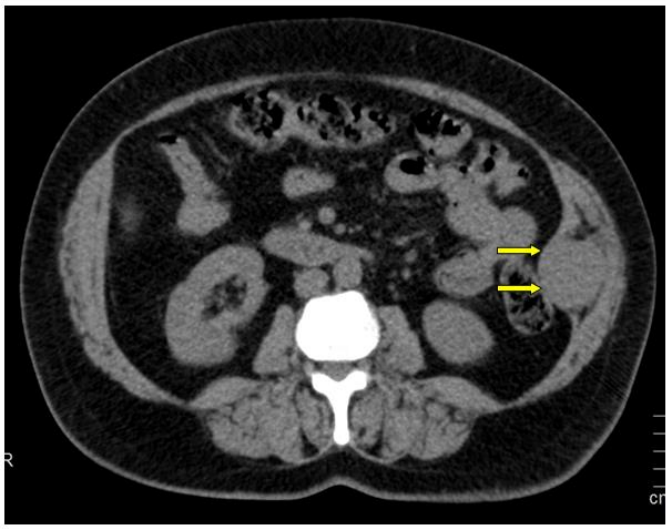


Figure 1: Abdominal CT scan without PDC injection: Axial section: objective a voluminous deep parietal tissue mass of the left transverse muscle with a well-defined rounded shape, regular contours, spontaneously isodense to the neighboring muscles (arrow)

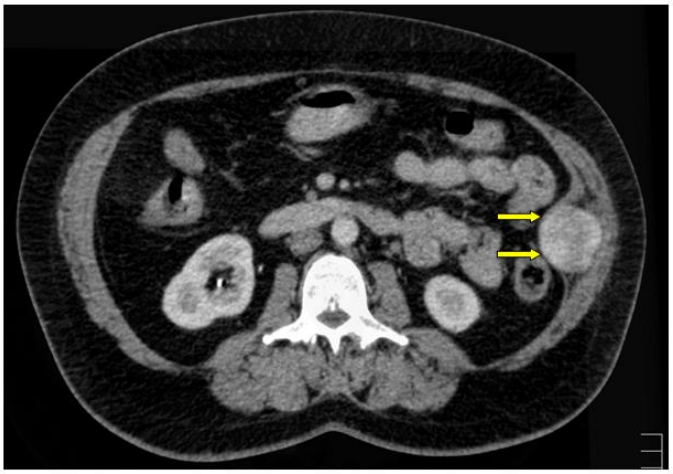
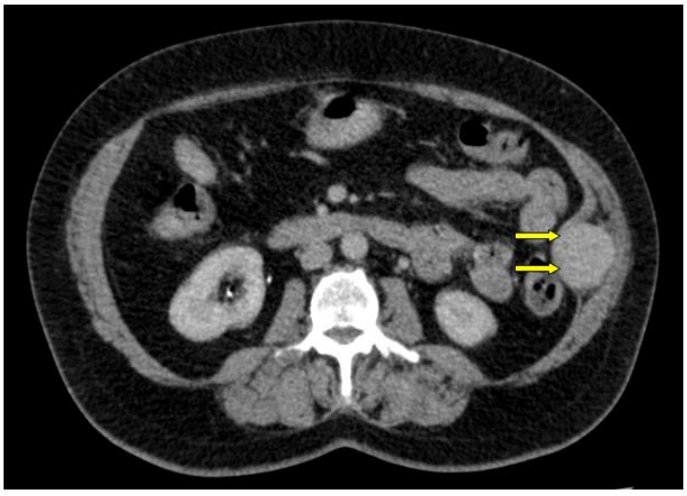


Figure 2: Abdominal CT scan with PDC injection; arterial phase (axial slice) which has objectified: heterogeneous enhancement of the mass (arrow)



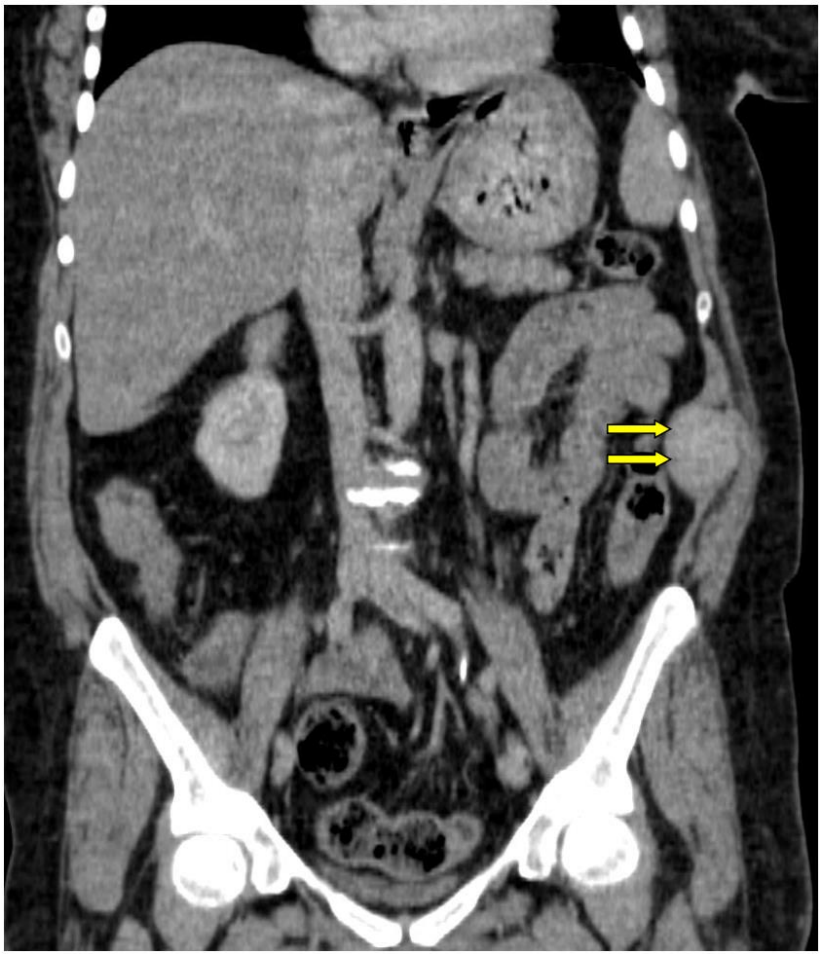
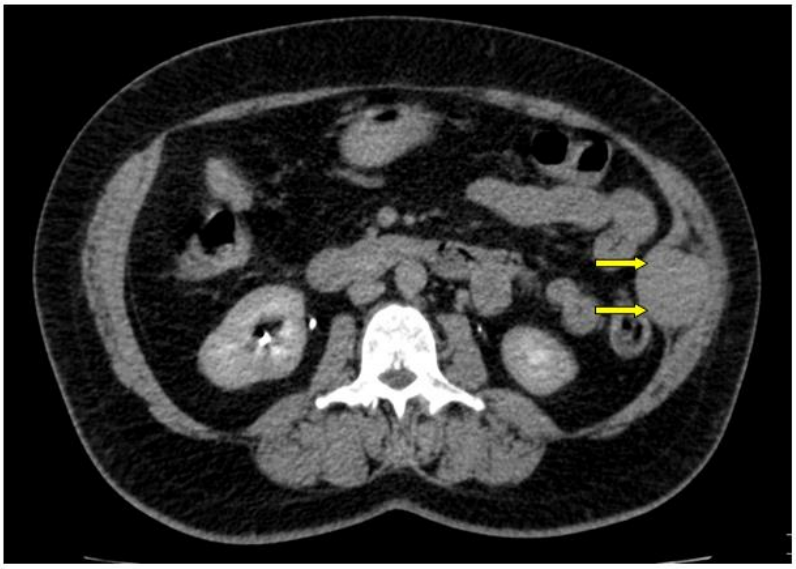


Figure 3 (a, b): Abdominal CT scan with PDC injection; portal phase (a: axial section, b: coronal section) portal phase homogenization (arrow)



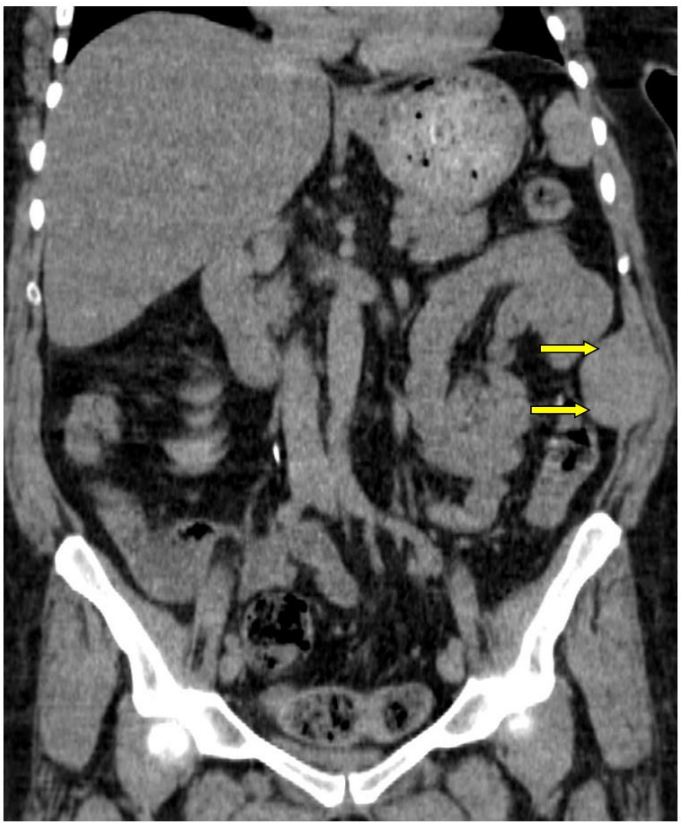


Figure 4 (a, b): Abdominal CT scan with PDC injection; late phase (a: axial section, b: coronal section) late washing (arrow)

DISCUSSION

Desmoid tumors are benign deep fibromatoses that arise from the fascia and muscular aponeuroses, especially the rectus and internal oblique muscle, and sometimes cross the midline. They have an infiltrating growth pattern and less often originate from the external oblique and from the transversalis muscle or fascia [2]. These are rare tumors occurring in 3.7 cases per million people each year. They have been correlated with female gender, PAF, and occasionally with abdominal trauma [1, 2]. The pathogenesis of desmoid tumors remains poorly understood, many hypotheses have been put [2]. Genetic factors in particular by deregulation of cell growth in the context of certain genetic syndromes such as familial adenomatous polyposis syndrome (FAP) or Gardner syndrome [2, 7]. Some arguments plead in favor of a probable hormone-dependence which explains the female predominance especially during periods of genital activity, and occasionally to abdominal trauma (physiological or surgical) [2, 6].

Clinically, PDDs are symptomatic in only 25-42% of cases [2]. The symptomatology remains nonspecific and dominated in the majority of cases by the presence of a deep, hard, poorly limited solitary mass of the abdominal wall [3] as is the case with our patient. The tumor size can reach about twenty centimeters.

Radiologically, imaging plays an essential role in the diagnosis, local extension assessment, preoperative planning and monitoring of desmoid tumors [6]. Scalability and locoregional extension, potentially very aggressive, are the most important criteria to evaluate to guide treatment (monitoring, surgery or medical treatment) and require a radiological reassessment initially shortly [8]. These tumors have no metastatic potential and it is therefore not necessary to do a remote extension assessment. On ultrasound, desmoid tumors appear as more or less well-defined tissue masses within a muscle or along an aponeurosis, of heterogeneous echostructure with hypoechoic components and posterior attenuation testifying to a component intra-lesional fibrous tissue within an iso- or hyperechoic stroma [2, 9]. The CT reveals a more or less well limited mass of tissue density, iso or discreetly hypodense compared to the neighboring muscle, enhancing homogeneously, sometimes heterogeneous in large tumorss with areas of necrosis, microhemorrhages and metaplasia [2, 6, 9]. MRI allows better tissue characterization, better study of relationships with adjacent structures (nerves, vessels, deep organs) and the differentiation of postoperative changes or after medical treatment of tumor recurrence. It is often an ovoid or infiltrating mass, with generally lobulated or sometimes irregular borders, presenting a homogeneous iso or hypointense in T1-weighted sequence and a variable signal often in hypersignal in T2 sequence, the contrast enhancement is intense and heterogeneous after

injection of gadolinium. The presence of hypointense bands, related to collagen bundles, on all the sequences is very characteristic [2, 6, 9].

The definitive diagnosis is based on anatomopathology [6]. The macroscopic appearance is very characteristic; it is most often a single non-encapsulated tumor adhering to neighboring tissues and at the periphery there are muscle bundles dissociated and included in the tumor [2]. Histologically, it is a proliferation of uniform spindle cells resembling myofibroblasts, in a stroma of abundant collagen and vascular network [2].

The therapeutic management of desmoid tumors is multidisciplinary [5], is essentially based on large surgical excision, when possible, despite the high risk of recurrence and the risk of peritoneal wound, large parietal defects, risk of eventration or infection and risk of ureteral and vascular wounds in deep forms [6]. Indeed, when the tumor is resectable, surgery must be performed. However, complete resection cannot be judged macroscopically because the tumor is not encapsulated [6]. In the event of an inextirpable tumor or a contraindication, medical treatment is based on hormone therapy and NSAIDs allowing stabilization or even more rarely regression of the size of the tumor. Chemotherapy cures or even external radiotherapy can be offered in aggressive forms, resistant to medical treatment or considered inextirpable as palliative or neo-adjuvant treatment [2, 5, 6].

CONCLUSION

Desmoid tumors remain rare entities, defined as aggressive fibromatosis due to their infiltrating nature with a high potential for recurrence. Imaging plays an essential role and makes it possible to make the diagnosis, guide therapeutic management and monitor progress. The treatment is ideally based on a large surgical excision followed by a parietal reconstruction using prosthetic reinforcement if necessary.

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