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Intra-Abdominal Desmoid Tumor: A Case Report

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Abstract **Case Report**

A 35 years old female patient, with no particular pathological history, presented for three months with an abdominal mass that was progressively increasing in volume, painful without vomiting or transit disorder. The clinical examination revealed a mass in the right iliac fossa, painful to palpation, round and mobile without inflammatory signs, measuring 50*40mm, with no palpable enlarged lymph nodes. The abdominal CT scan showed an anterior abdominal mass lateralized to the right side, localized in the right internal oblique muscle, measuring 73*54*86 mm, associated with right external iliac lymph nodes measuring up to 8 mm with a low peritoneal effusion of the pouch of Douglas. A biopsy was performed and the histological and immunohistochemical study was positive for desmoid tumor. The patient was treated by wide tumor resection, and put on medical treatment: Tamoxifen-based hormone therapy (20mg/day) with regular monitoring by abdominal CT scan every six months. The evolution was favorable, with no recurrence after 23 months of follow up.

Keywords: Desmoid tumor, Management, Outcome.

INTRODUCTION

Desmoid-type fibromatosis (DTF) is a rare disease with an annual incidence of 2 to 4 patients per million [1, 2]. Abdominal wall DTF is often affecting young female patients, aged between 10 and 40 years old [3]. DTF is defined as benign neoplasms of the connective tissue with musculoaponeurotic origins [4]. Recurrence rates are reported at 40 to 80% after surgery alone [5]. In the present paper, we report a case of an abdominal desmoid tumor well controlled by medical treatment.

CASE REPORT

A 35 years old female patient, with no particular pathological history, presented for three months with an abdominal mass that was progressively increasing in volume, painful without vomiting or transit disorder.

The clinical examination revealed a mass in the right iliac fossa, painful to palpation, round and mobile without inflammatory signs, measuring 50*40 mm, with no palpable adenopathies.

The abdominal CT scan showed an anterior abdominal mass lateralized to the right side, localized in the right internal oblique muscle, measuring 73*54*86 mm, associated with right external iliac lymph nodes measuring up to 8 mm with a low peritoneal effusion of the pouch of Douglas.

A biopsy was performed and the histological and immunohistochemical study was positive for desmoid tumor.

The patient was treated by wide tumor resection with tumor-free margins and put under Tamoxifen-based hormone therapy (20mg/day) with regular monitoring by abdominal CT scan every six months. After 24 months of follow-up, no recurrence was reported.

DISCUSSION

Desmoid tumors are characterized by their low incidence, locally aggressive presentation, and high rates of local relapse. Although they don't metastasize, their infiltrative growth and tendency to recur even after macroscopic complete excision presents a real challenge in desmoid tumors. They are un-encapsulated fibrous tumors consisting of spindle- shaped cells [6]. They arise from the muscles or aponeurotic tissues and are classified as sporadic or hereditary when they occur in patients with familial adenomatous polyposis (FAP) [6, 7].

The incidence of these tumors has been estimated at 2 to 4 cases/10 [7], they occur in about 10% of patients with FAP, particularly those with Gardner syndrome [8]. Interestingly, the anatomical location of desmoid tumors in FAP patients differs quite markedly from the location of sporadic tumors, being much more likely to be intraabdominal or in the anterior abdominal wall than in extra-abdominal sites [9].

The symptoms of desmoid tumors depend on their location and ranges from incidental small stable lesion to rapidly-growing, huge abdominal masse [10]. These tumors may be asymptomatic and, in this case, they are found incidentally at routine clinical examination or remain asymptomatic until their growth and infiltration cause visceral, neural or vascular compression. The most common sign, as in our case, is the onset of a relatively firm mass or lump, causing mild pain [10].

For imaging findings, on ultrasound, DT appears as homogeneously hypoechoic masses. They may have a similar appearance to muscle, they may be lobulated and may show vascularity on color Doppler interrogation [11]. On CT scan, most desmoid tumors appear as a well-circumscribed masse, although in some cases they may appear more aggressive with ill-defined margins [12]. They are relatively homogeneous or focally hyperattenuating when compared to soft tissue on the non-contrast scan and demonstrate enhancement following administration of intravenous contrast. When it comes to soft tissue tumors, MRI is the more sensitive imaging modality to characterize local tumor extension. Their appearance is accounted for by their dense cellularity [13]. Typical signal characteristics include a T1 and T2 low signal intensity with homogeneous, inhomogeneous, or no significant enhancement after gadolinium injection on T1 C+ sequences [13].

Management of patients with desmoid tumors remain a big challenge for oncologists. Although the significant improvements of different treatments, many issues remain controversial, mainly regarding early detection, the role, type and timing of surgery, and the value of non-operative therapies.

Complete surgery with wide resection and sufficient margins remains the only curative treatment and considered to be a key determinant for disease prognosis. However, it can be very challenging even for experimented surgeons, especially for large and locally advanced tumors. When well executed, the surgical resection alone confers a very fluctuent rate of local control ranging between 40 and 80% at 5 years [14].

Radiotherapy (RT) is another treatment modality for desmoid tumors but its efficiency is controversial. It has a long tradition in the treatment of desmoids since 1928, when James Ewing, who was the first to propose RT as a treatment option, described that desmoid lesions respond 'slowly but satisfactorily' to RT [15]. These observations were confirmed by several studies that demonstrated high local control rates after RT [16, 17]. Based on retrospective studies and a recent metanalysis, radiation therapy (RT) improved local control in the primary and postoperative settings [18, 19]. therefore, the best local control is currently ensured by a combined approach.

Radiation therapy is mainly indicated for primarily unresectable or recurrent tumors and non-in-Sano resected tumors, its dose range between 50 and 60 Gy [17], with no benefit has been observed using doses higher than 60 Gy [19].

In other hand, there are no evidence-based or widely accepted guidelines for the use of systemic therapies in the management of unresectable desmoids tumors [20]. Although, they are being increasingly integrated into a multidisciplinary approach for selected patients. They may be classified into chemotherapy, Tyrosine Kinase Inhibitors (TKIs), nonsteroidal anti-inflammatory drugs (NSAIDs), and hormonal therapy [20].

Consensus-based guidelines for treatment of desmoid tumors from the National Comprehensive Cancer Network (NCCN) suggest systemic therapy as an option for unresectable desmoid tumors and those for which surgery would be unacceptably morbid, but provide limited guidance for the choice of agent [10].

Antihormonal therapies such as tamoxifen can be used alone or in association with NSAIDs as first-line treatment. There are some suggestions that higher doses (up to 120 mg/ day) in combination with NSAIDs are more effective than tamoxifen alone [21].

Chemotherapy can be the treatment of choice in case of unresectable, aggressive, rapidly growing, and/or life- threatening tumors [22]. Many chemotherapy protocols have been used mainly pegylated liposomal doxorubicin (PLD), Doxorubicin, combinations of doxorubicin plus Dacarbazine, or Methotrexate plus Vimblastine. Some institutions also use methotrexate alone, although there is almost no published experience utilizing this approach [22].

Tyrosine kinase inhibitors (imatinib and sorafenib) have demonstrated activity in the treatment of Desmoid tumors due to PDGFRs expression in tumor stroma. In a retrospective study published by Gounder *et al.*, Sorafenib was administered at 400 mg oral daily. There was a 30% reduction of the tumor size in 92% of the patients [23]. Penel *et al.*, evaluated the efficacy of Imatinib in patients with unresectable and progressive symptomatic Desmoid tumors, and showed that 83% of patients had stable disease; 3% had a complete response rate and 9% had a partial response rate [24].

CONCLUSION

Surgery is still the primary treatment modality for patients with resectable desmoid tumors. Adjuvant radiotherapy is required for incomplete resection and combined with surgery they result in a better local control. The use of systemic therapies still controversial. More prospective randomized trials are needed to establish the indications of each treatment modality.

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