

DESMOID TUMORS OF THE ABDOMINAL WALL AND THE THERAPEUTIC MANAGEMENT: CASE REPORT.

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ABSTRACT

Desmoid tumors are fibrous proliferations with an aggressive and infiltrating character which tend to recur locally without giving any metastasis. They are rare, representing less than 0.03% of all tumors. Mostly, they are sporadic, but in 2% of the cases they could come in a genetic context “The Gardner Syndrome”. Desmoid tumors are localized, appearing in a decreasing order of frequency in the structures of the abdominal wall; the peripheral muscle fascia or more rarely in the intra-abdominal contents (mesentery, retroperitoneum).

We report here the case of a 32 years old female patient who had a desmoid tumor of the abdominal wall discovered during pregnancy. From this clinical case, we will focus on the difficulties of their therapeutic management, recalling through a literature review: the clinical, paraclinical and therapeutic features of those tumors.

Keywords: abdominal wall- Aggressive fibromatosis- desmoid tumor- infiltrated margins.

INTRODUCTION

Desmoid tumors or Aggressive Fibromatosis are soft tissue tumors caused by the proliferation of fibroblastic cells. They are neoplasms with a low grade of malignancy that never give metastasis but tend frequently to recur locally [1-4]. The circumstances of discovery are multiple and their clinical manifestations are not specific. The therapeutic management should be similar to the one for the low-grade fibrosarcoma [1, 2]. Their evolution is unpredictable making them dreaded tumors [5, 6]. This character affects their management which remains the real problem: What treatment must be followed initially for an optimal care? What should we do in case of surgery with positive margins or in case of recurrence?

OBSERVATION

We report the case of a 32 years old female patient, ASA 1 (American Society of Anesthesiologists Score), WHO 0 (World Health Organization performance status), Body Mass Index (BMI) 27, without any significant pathological history. The patient presents a painless induration of the right side of the abdomen initially appeared during her second pregnancy and constantly increasing volume. The abdominal examination found a mass of 60x30mm localized halfway between the umbilicus and the right antero-superior iliac spine, highly suspecting a parietal origin. The results of the Biology screening tests were normal. The abdominal computed tomography (CT) scan objectified a 70x20mm subcutaneous process having an intimate contact with the rectus abdominis muscle (**Figure 1**).



Figure 1 A, B: Computed tomography showing the abdominal wall tumor

The patient had an “en bloc” resection taking the whole process with the skin, the sub- cutaneous tissue and the superficial fascia of the rectus abdominis with 2 cm of safety margins that are macroscopically healthy. The patient did not need any parietal reconstruction and the closure process was made by simple sutures. The pathological result showed a desmoid fibromatosis with invaded margins (R1). A complementary surgery and/or adjuvant radiotherapy were discussed with our patient who opted for just a clinical and radiological surveillance. A gastro-intestinal endoscopy and a total colonoscopy were made as part of the exploration of a possible Familial

Adenomatous Polyposis (FAP) associated, but they revealed no abnormalities.

A control CT scan was performed 6 months after the surgery and showed just remnant scars without any parietal process (**Figure 2**). The surveillance policy was maintained for another year by performing every 3 months ultrasounds that revealed no local recurrences or any abnormalities.



Figure 2 : Computed tomography 6 month after the surgy showing remnant scar without recurrences.

DISCUSSION

As already mentioned, desmoid tumors are rare neoplasms representing less than 0.03% of all tumors [7]. They are either sporadic or part of a genetic syndrome. They are localized in a decreasing order of frequency at the abdominal wall (49%), extra-abdominal (43%) or intra-abdominal 8% (mesentery, retroperitoneum) [8]. According to Reitamo et al, they can be divided into 4 groups: Juvenile tumors; women’s procreative period tumors; tumors of menopause and tumors of the elderly period (without discrimination between genders).

Several factors have been implicated in their genesis [1, 3, 9]

Incidence of genetic factors [1, 2, 7, 10, 13] in patients with FAP [7, 11, 14] or carriers of the Gardner Syndrome [1, 15] increases compared to the general population. Hormonal factors [1, 2, 7, 15] and specifically the estrogen stimulation (procreation period, pregnancy or oral contraception) also increase their incidence. Surgery for FAP or any kind of surgery in general can promote the occurrence of desmoid tumors on the old scars or the port sites [7, 16].

Their clinical manifestations changes according to their localizations, but over all they are painless masses, slowly evolving, causing a discomfort requiring finally a consultation. The angio CT scan is the main stay imaging test. It makes the diagnosis by showing a mass without clear limits with the same muscle density. The enhancement after the contrast administration is heterogeneous. The angio CT scan can also eliminates the fibro sarcoma by showing the absence of a highly developed neo -vascularisation [1, 2, 4, 7].

The gastro-intestinal endoscopy and the colonoscopy are systematic to eliminate an associated FAP and to have the right label: a sporadic tumor or a genetic context [11, 12]. For the post-operative surveillance, the Magnetic resonance imaging (MRI) is the best imaging test to explore the recurrences. It shows a heterogeneous mass, poorly limited, iso-signal to the muscles in T1 and hyperintense in T2 sequences and heterogeneously enhancing after the contrast administration [1, 2].

The biopsy is not recommended due to the boosting effect on the tumor growth. Although it remains indicated in case of unresectable tumor to confirm the diagnosis and initiate a treatment. The pathological result commonly shows a uniform proliferation of fibroblastic cells separated by collagen tissue without any necrosis or haemorrhage which differentiate them from the fibrosarcoma. Microscopically, the tumor proliferation extends up to 2-3cm in the peripheral tissue of the actually palpable mass making the R0 resection very difficult. The immunohistochemistry shows a positive reaction on the muscle cells markers like the vimentin, the desmin, the S100 protein and the alpha-actin. The desmoid tumors can sometimes express positively the c-KIT marker rising more difficulties to determinate the right diagnosis.

There are other markers like CD117 antibodies and CD34 that are used to differentiate between desmoid tumors and other ethiology such as the gastrointestinal stromal tumors (GIST).

The oncological R0 resection +/- associated with a parietal reconstruction remains the mainstay of the treatment. A radical resection with margins that are macroscopically and histologically tumor free is the goal. This condition does not eliminate the recurrences which represent 27% even in R0 resection and 54% in R1/R2 resection [1]. A macroscopically healthy margin (R0) does not eliminate invaded margins (R1) at the pathological result due to the infiltrant character without any

individualised capsule of the tumor [1, 17]. A partial resection with the aim of a cyto-reduction should be avoided because of the accelerating effect on the tumor growth.

When the radical resection comes with the cost of a major parietal defect, the reconstruction requires a multidisciplinary collaboration between the general and plastic surgeons. The objective is to protect the intra-abdominal contents, to ensure the parietal function while trying to maintain a satisfactory appearance.

Several therapeutic options are available such as: the controlled healing of the wound; the muscle grafting or the cures by using prosthetic material. The radiotherapy allows a local control in case of unresectable tumors, invaded margins or recurrences [1]. Even after a R0 resection, it can reduce the recurrences from 28% to 6% [12]. Because of the influence of the estrogenic hormone as a possible etiological factor in the genesis of desmoid tumors, the prescription of a hormonotherapy based on the anti-estrogens (Tamoxifen), the progestins or the Gonadotropin-releasing hormone (GnRH) agonists have shown in several studies a shrinking effect on the tumor's volume[1,8,18].

Generally, desmoid tumors have a low chemosensitivity, but the chemotherapy remains the last resort in cases of highly aggressive desmoid tumors, unresectable ones or resistant to the conventional medical treatment [19]. Several molecules are tested but the most commonly used are the anthracyclines which can give a stabilization of the tumor's volume over a long period or even a regression. 20 to 30% of the desmoid tumors express CD117 or c-Kit at the immunohistochemistry result [19, 20, 21]. In these particular cases, the prescription of Imatinib showed good results with a long stabilization period [22]. Recently, there were studies that evaluated the effect of Sorafenib on the desmoid tumors with promising results. Gounder et al establishes a retrospective review on 26 patients with desmoid tumors (clinical/radiological progression, recurrence) which were placed under Sorafenib 400mg/day as a first-line therapy (11patients/26) or after a median of 2 lines of prior systemic therapy (15patients/26) [23]. 70% of the patients reported a rapid clinical benefit: partial or complete resolution of the symptoms (pain, swelling...). At a median of 6 months of treatment, the Response Evaluation Criteria In Solid Tumors (RECIST) included 6/24 (25%) patients with partial

response, 17/24 (70%) with stable disease, and 1 with progression. There was also a radiological benefit which was 30% decrease in T2 signal intensity in patient evaluated by MRI 12/13 (92%) patients [23]. Other medical treatments have also shown a good response, such as non steroidal anti-inflammatory drugs (NSAIDs), the Tumor Necrosis Factors (TNF), the Colchicine and the interferon by their anti-COX2 action and by inhibiting the growth factors [7, 8, 15,13].

The therapeutic abstinence with periodic clinical and radiological follow-ups is a significant and a defendable therapeutic option especially for asymptomatic patients with small desmoid tumors. In fact, there is a study about 27 non-operated patients who were initially put on simple surveillance: more than half of them (16 patients) have not progressed; a spontaneous regression was noticed in 5 of them and an increase in tumor volume was seen in 6 patients [24].

Other cases of spontaneous regression (although rare) have been reported in some series (especially in women after menopause or after oophorectomy). These results confirm the possibility of establishing "The wait and see" policy [25].

Our patient had a radical resection for curative purposes with macroscopically tumor free margins which returned invaded (R1) in the pathological result. "The wait and see" policy combined to a rigorous surveillance based on clinical and radiological examination remains a tenable option according to literature data in case of refusing a complementary surgery or radiotherapy. Our case has an 18 months follow-up without any recurrences.

CONCLUSION

Desmoid tumors are neoplasms, benign but locally aggressive. They mainly develop at the expense of musculo-aponevrotic structures of the abdominal wall. Their evolutionary pattern is unpredictable. The management is not codified and remains question of debate between the scientific communities. "The wait and see" policy seems to be a defendable option for no symptomatic patients, no FAP associated context or the refusal of any invasive treatments.

ABBREVIATIONS

ASA: American Society of Anesthesiologists Score.
WHO: WHO performance status.
BMI: Body Mass Index.
CT: Computed tomography.
FAP: Familial Adenomatous Polyposis.
MRI: Magnetic resonance imaging.
GIST: Gastrointestinal stromal tumor.
GnRH: Gonadotropin-releasing hormone.
RECIST: Response Evaluation Criteria in Solid Tumors.
TNF: Tumor Necrosis Factor.
NSAIDs: Non Steroidal Anti-Inflammatory Drugs.
Anti-cox2: Anti Cyclo-oxygenase 2.

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