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Case Report

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DESMOID FIBROMA OF SOFT TISSUES OF EXTRA-ABDOMINAL SITE: ABOUT A CASE AND REVIEW OF THE LITERATURE

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ABSRACT

Extra-abdominal desmoid fibromatosis (EADF) is a rare benign tumor with a high risk of local recurrence, which distinguishes it from other benign tumors.This tumor mainly affects women in the third and fourth decades. The pathogenesis of these extra-abdominal tumors remains unknown. MRI (Magnetic resonance imaging) is most useful for the diagnosis and follow-up of a desmoid tumor. Anatomopathological examination confirms the diagnosis of this tumor, which raises a problem of differential diagnosis with low-grade fibrosarcomas. Surgery remains the main part of the therapeutic protocol despite the appearance of new means such as radiotherapy and hormone therapy. We report a case of extra abdominal desmoid tumor treated essentially by surgery with adjuvant radiotherapy without any sign of recurrence.

KEYWORDS: Desmoid Tumours, Fibrosarcomas, Recidivism, Surgery, Radiotherapy, Hormone Therapy.

1. INTRODUCTION

Desmoides tumors or aggressive fibromatosis are soft tissue tumors.[1] These are fibroblastic infiltrating tissue proliferations that do not metastasize but tend to recur. It is not a reaction proliferation but a low-grade neoplasia, whose management must be identical to that of low-grade fibrosarcomas.[1] Several factors have been implicated in the pathogenesis of this tumor type. We report in this context the case of a 48-year-old woman who had a desmoid tumor in the left gluteal region. as well as the review of the literature in order to specify the characteristics of the tumor and to codify the behavior to be maintained.

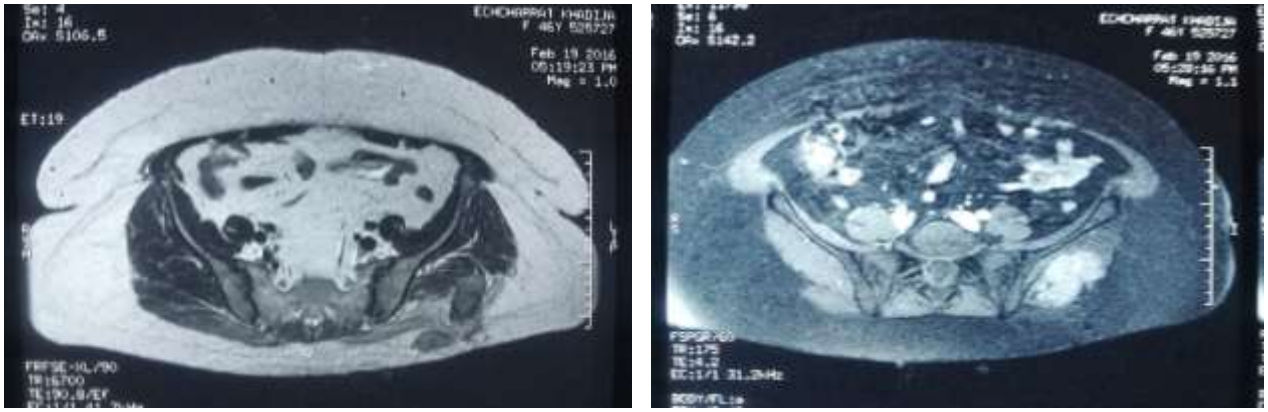
2. THE CASE REPORT

A 48-year-old woman, mother of 3 children, with no pathological history, who consulted for a mass of the left gluteal region, evolution is rapidly progressive over 1 year and becomes painful, the general examination is without particularity. Local examination revealed a tumefaction 7 x 6 cm poorly limited at the superior left outer edge of the left buttock, roughly oval, longitudinal, the skin opposite was normal, the examination of the inguinal and supraclavicular ganglion areas was normal.. Standard radiography did not show any evidence of bone involvement. The diagnosis of liposarcoma or rhabdomyosarcoma has been discussed.

The MRI pelvis and gluteal region (Figure 1). A biopsy of the mass was performed and concluded the diagnosis of deep fibromatosis desmoide type (Figure 2).

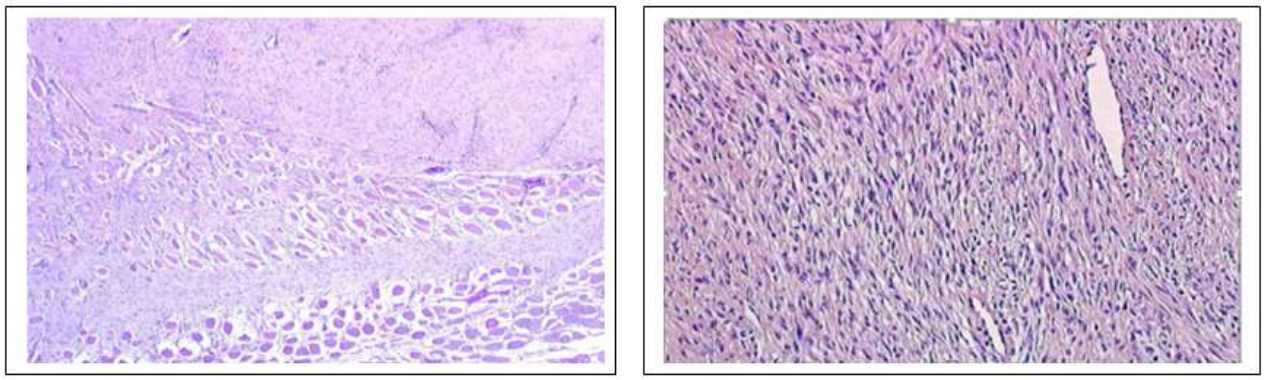
the patient has benefited from large tumor excision, histopathological examination confirmed the diagnosis but the internal limit of the excision was reached and Surgical revision was difficult given the risk of functional complication, we therefore opted to complete the therapeutic management by an adjuvant external radiotherapy on the tumor bed after a good healing of the exeresis wound (Figure 3) at a dose of 50 Gy: 2Gy per fraction in 25 Fractions; 5 jours sur 7, with a very good tolerance.

After a 2-year follow-up, there was no functional impairment, the scar was of good quality and there was no clinically and radiologically detectable recurrence.



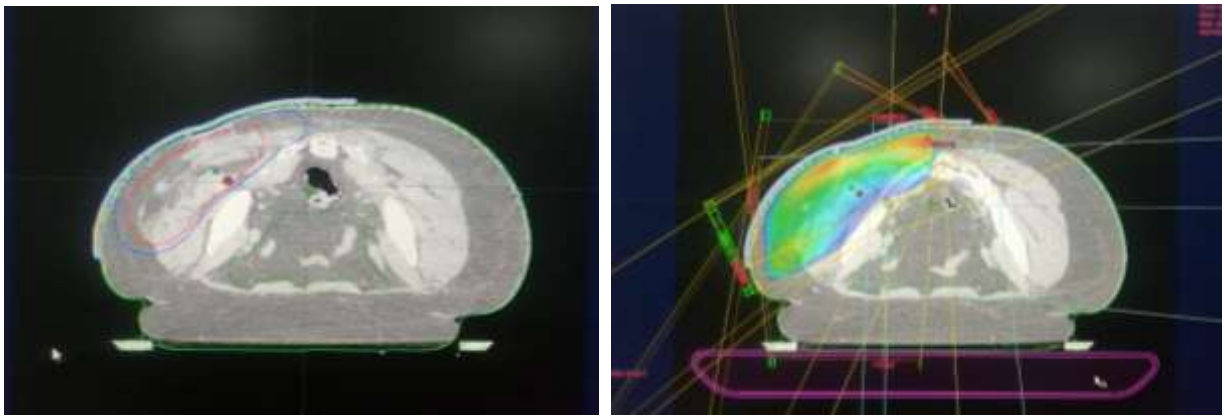
(A) **(B)**

Figure 1: MRI: Cross section: tumor process of the gluteal region left infiltrating the muscles large and medium gluteal, (A): T1 isosignal injury to the muscle with hypersignal zones, (B): Fat-sat Gadolinium: intense contrast enhacement, infiltration of subcutaneous fat.



(A) **(B)**

Figure 2: Biopsy of Desmoid tumor of the left gluteal region.: histological section: (A) proliferation of fusiform cells (myofibroblasts), poorly bounded at the periphery and entangled with adjacent striated muscle fibers. (B) High magnification: proliferation of myofibroblasts with atypical nuclei forming bundles in a Fibrous stroma,



(A) **(B)**

Figure 3: CT simulation of radiotherapy (in procubitus position): (A): The delineation of the target volumes: CTV (clinical target volume) which corresponds to the tumor bed, PTV (predictive target volume) and organs at risk. (B): Dosimetry: homogeneous distribution of the dose on the target volume, 95% isodose which covers PTV well.

3. DISCUSSION

Extra-abdominal desmoid fibromatosis (EADF) have significant malignant potential because of local invasion and frequent recurrence. They represent 0.03%[2] of tumors, all types of cancers combined. Most of the major series published in the literature are multicenter studies. These tumors can occur at all ages[3], in both sexes, but especially in young women between 20 and 40 years old.[4] The etiology of EADF is unclear. Several factors were mentioned, but no cause-and-effect relationship

could be scientifically highlighted. Among the supposed factors, various authors incriminate an inaugural traumatism in 19 to 49% of cases.[5,6] These traumas include: a genetic predisposition; simple bruises; fractures; the pregnancy; surgical history.[5-7] As in our case, the majority of EADF reported in the literature sit at the level of the limb root, but also at the level of the belts and more rarely at the level of the thorax and the neck.[8,9] They are generally unique, but multi-focal locations have been described.[8,10] Cases have been observed in the literature where two desmoid tumors very close anatomically may be due to an extension gradually by following the connective flows.[11] Magnetic resonance imaging (MRI) has been confirmed as the exam of choice, it allowed to appreciate the limits of the tumor, its relations with the vasculo-nervous bundles. Diagnosis could be evoked in the presence of a poorly limited mass with cicatricial components with a signal inferior to fat in T2.[12,13] The diagnosis of certainty is anatomopathological and the problem is to eliminate a low-grade fibrosarcoma of malignancy. The predominance of collagen fibers over fibroblasts, the rarity or absence of mitoses and the absence of cellular atypias favor desmoid tumors[14] As in our case. Ultrastructural examination shows the presence of myofibroblasts, which are the most common, more rarely fibroblasts and mesenchymal cells.[15]

Surgical treatment and / or radiotherapy are widely discussed in the literature, Surgery remains the main part of the therapeutic protocol despite the appearance of new means such as radiotherapy and hormone therapy. Surgical excision should be wide to avoid the risk of relapse. This risk of global relapse is 30% at five years and 33% at ten years.[16,17,18,19] Indeed, wide excision is not always possible because of the importance of tumor size, invasion of vasculo-nervous bundles, difficult locations (shoulder, buttock, thigh root) and the poorly limited appearance of the tumor. Desmoid tumors are not very radiosensitive and require high energy radiation with total doses greater than 50 Grays.[20] In case of invaded margins (As in our case) or relapse, adjuvant external radiotherapy with doses of 50 to 55 Gy can improve local control in 80% of cases, as well as for macroscopically inoperable tumors[16,17], The risk of radiotherapy is the malignant transformation[21], especially in fibrosarcoma, even if this possibility is rare.

The detection of hormone receptors in the tumor cells has led to therapeutic trials based on tamoxifen-type antiestrogens, GnRH agonists (gonadotropin-releasing hormone) and progestins, these products used alone or in combination have shown objective response in 52% of cases.[17]

Nonsteroidal anti-inflammatory drugs such as sulindac or indomethacin, used alone or in combination, have made it possible to obtain stabilization or even regression with very low toxicity.[16] Chemotherapy is not very effective, most of it comes within the framework of research protocols. The location at the shoulder, of the gluteal region, gives huge tumors affecting several muscles and whose resection can affect the function of the limb.[8,22]

Despite the insufficiency and the difficulties of the surgical treatment, the wide resection remains the recommended treatment of first intention by the majority of the authors. The important thing for this type of benign tumor, although recurrent, is to find a compromise between a carcinological excision and a surgery preserving the function.

The question of the therapeutic choice thus meets two criteria, namely the minimal surgical excision without risk of recurrence and the maximum surgical excision limiting the risks of functional sequelae. These tumors present themselves, as usually reported, in the form of a muscular mass, more or less deep, of variable volume ranging from a few centimeters to several tens of centimeters. They are adherent to the deep plane, never superficial, but contrary to our case, generally painless. The seat near a joint can cause a limitation of mobility. This typical clinical aspect and localization, can sometimes make mention the diagnosis before the complementary examinations. On the other hand, in the case of children, these tumors may have a symptom of borrowing that is often misleading.[23]

Recently, Leithner et al.[24] conducted a comparative analysis to evaluate the prognosis of microscopic margins of removal of EADF treated only by surgery and noted that the recurrence rate was much higher when it was a marginal or intra-lesional excision unlike a large extra marginal excision passed in sano (respectively 72% versus 27%), Based on this analysis, the majority of authors, except for Reitamo[20] and Plukker et al.[22] for whom the quality of initial resection has no influence on recurrences, recommend a wide and radical excision of EADF, The margin of excision is not codified; it depends mainly on the technical possibilities.[9] Other factors that have been reported as a major prognostic factor for the prognosis of EADF: the existence of anterior recurrence of the tumor desmoide, the size of the tumor, the location (the limbs and the trunk are factors of poor prognosis), the female sex and finally the age, Since in our case, it was a 48-year-old postmenopausal woman who was seen for a tumor in the left gluteal region, with invaded margins. (difficult surgical revision), this patient has a high risk of recurrence and we preferred to supplement with adjuvant treatment by external radiotherapy and not to cover with a flap before making sure of the absence of recurrence (delay of 3 years). However, G. Gallucci et al.[25] reported a case of excision and reconstruction in one time in a young patient (23 years old) who did not have a factor of poor prognosis and given the long life expectancy.

4. CONCLUSION

EADF are rare benign tumors that are peculiar by the high risk of local recurrence. Concerning the case of our

patient, in the current state of the medical knowledge, and after study of the literature, we can affirm that there is neither a statistical link nor any scientific proof that could explain the occurrence of a EADF. The extra marginal marginal resection remains the reference treatment, with a significant cure rate in the case of healthy margin resection, and adjuvant radiotherapy seems the reference treatment in case of invaded margins, which is the case of our patient,with a low recidivism rate.

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