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Recurrent Desmoid Tumor of the Gluteal Region about a Case and Review of the Literature

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Abstract **Case Report**

Desmoid tumors or aggressive fibromatosis are recurrent but non-metastatic fibrous infiltrative proliferations. They are close to low grade fibrosarcoma. They are rare tumors since they present less than 0.03% of all neoplasias. Surgical treatment remains the treatment of reference but local relapses occur in one third of cases within five years, depending on the location, age and the possibility of a satisfactory resection. Complementary radiotherapy reduces this risk. We report a case of a recurrent desmoid tumor in the right gluteal region with intra-abdominal extension in a 40-year-old patient.

Keywords: Desmoid tumor, aggressive, recurrence.

INTRODUCTION

Aggressive fibromatosis or desmoid tumor is a soft tissue tumor. It develops from supporting tissues and fasciae. It is a benign tumor under the microscope that does not give metastasis. However, although benign, aggressive fibromatosis is a very invasive tumor locally and whose complete removal is difficult. Its evolution is unpredictable; some forms relapse repeatedly and quickly, others evolve in thrusts, interspersed with prolonged phase of stability

OBSERVATIONS

The patient, a 40-year-old woman, was hospitalized for a tumor of the right gluteal region. The beginning dates back to 2010 by the appearance of a swelling in the right gluteal region gradually increasing in size with paresthesia in the territory of the sciatic nerve. A radiological check-up based on a standard radiograph of the pelvis and a CT scan of the pelvis revealed a gluteal and retro-peritoneal tumor with endo and exo-pelvic development. A biopsy was performed that was in favor of a desmoid tumor. At the time, the patient underwent surgical treatment with the most complete resection possible using a dual approach (posterior tongue of LANGENBECK (figure 1) and retro-peritoneal lumbotomy). In May 2014, the patient presented a recurrence of the tumor in the gluteal region for which it received a posterior recovery and resection of the tumor with an adjuvant hormone therapy with Tamoxifen. In May 2016, the patient was presented with a heaviness in the right lower limb. During the clinical check-up, the general condition was preserved with a thickness of the right gluteal region, and the patient had no motor or vascular deficit of the right lower limb. a standard radiograph (Figure 2) and a CT scan of the pelvis (Figure 3) favored a new recurrence of the desmoid tumor.

Since the symptomatology is not very worrying, and given the high potential of recurrence of the desmoid tumor, surgical abstention was recommended with regular monitoring of the patient.

DISCUSSION

Desmoid tumors, also known as aggressive fibromatosis, are part of deep fibromatosis, which are themselves integrated into the group of soft tissue tumors. The desmoid tumor is rare since it corresponds to 0.03% of all neoplasias [1], there is a female predominance with a sex ratio of 2/1 for an average age of 20 to 40 years [1]. The tumors desmoid extra abdominal can reach several sites, the most common: are the shoulders and the chest wall before it reaches the thigh and the cervical regions [1,2] from a clinical point of view, the tumor is in the form of a mass of miscegenation, poorly limited, slow growth, little or not sensitive, mobility discomfort or nerve compression may occur depending on the location and extension of the tumor [2]. The scanné, especially the MRI, represents the selected radiological examinations. The confirmation diagnosis is based on the anatomopathological study of the operative specimen.

Although never metastasizing, the desmoid tumor recurs frequently : 35% on average within five years of surgery. The transformation of aggressive fibromatosis into sarcoma is rare and remains controversial [1,3].

Iconography

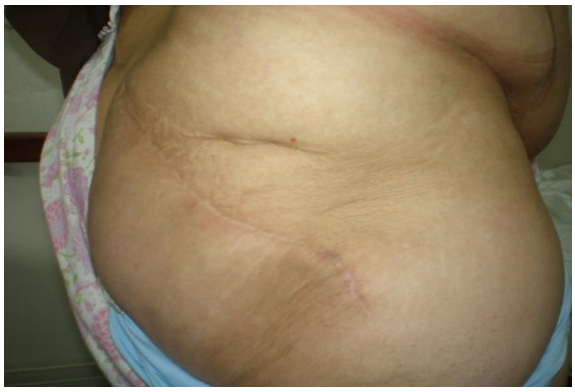

Image-1: Clinical image of the formé LANGENBECK posterior approach

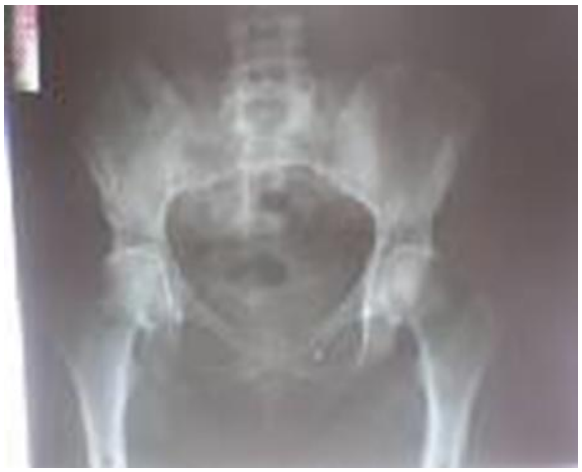

Image-2: Standard radiograph of the patient's pelvis does not show bone involvement

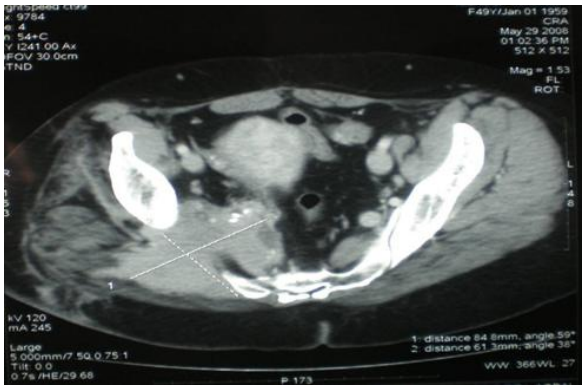

Image-3: Pelvic MRI shows the endo and exopelvic development of the desmoide tumor

Surgical treatment of desmoid tumors remains the basic treatment. In literature, to avoid relapses as much as possible, it is advisable to perform either a resection, a wide resection, or a radical resection of the affected muscles [3]. Adjuvant radiotherapy is especially indicated for resections with invaded margins or after relapse, thus allowing local control [4-6]. In case of inoperable or recurrent lesions, many treatments have been tested but with an effectiveness discussed [1, 3, 4-7]: hormone therapy based on sex hormones (tamoxifen, Gn-Rh agonists, progestins such as medroxyprogesterone acetate) nonsteroidal anti-inflammatory drugs such as indomethacin chemotherapy: the slow evolution of these tumors does not indicate a great chemosensitivity. The literature reports on the use of so-called "mild" chemotherapy with weekly vinblastine or vinorelbine and methotrexate.

CONCLUSION

The desmoid tumor is a rare condition whose diagnosis must be documented histologically, and management is based mainly on surgery supplemented by radiotherapy if the margins are unaffected. Medical treatment is recommended in case of progression or relapse.

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