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Case Report

Recurrent Vulvar Dermatofibrosarcoma Protuberans: A Real Management Dilemma

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Abstract

In 1924, Jean Darier and Marcel Ferrand described for the first time dermatofibrosarcoma as a true clinical and histological entity. It is a fibrous tumor of the skin with high local malignancy, progressive and with a high potential for recurrence. We report an unusual location of recurrent dermatofibrosarcoma protuberans of the vulva in a 58-years-old patient. The clinical examination showed a large, rounded mass of firm consistency. The surgical resection was complete, passing to 5 cm laterally with economy of normal tissue leaving the minimum of functional and aesthetic sequelae. Histological examination of the operating piece confirmed the diagnosis. This observation illustrates the voluminous nature of the mass, the local aggressiveness of the tumor and the potential difficulties of wide excision laterally and in depth of the lesion in an important anatomical region. This excision results in large losses of substances whose cover is a real challenge for the surgeon.

Keywords: Dermatofibrosarcoma; Vulva; Recurrence; Treatment; Surgery.

INTRODUCTION

Jean Darier and Marcel Ferrand described for the first time dermatofibrosarcoma as a true clinical and histological entity in 1924, when they initially defined it as "progressive and recurring dermatofibroma" [1]. The protuberant dermatofibrosarcoma (DFSP) is a rare, low-grade, soft tissue sarcoma arising in the dermis, which is distinguished by its slow growth, its local aggressiveness, its high ability of recurrence and the rarity of distant metastatic spread [1,2]. The treatment consists of a radical surgery for adequate margins. The prognosis depends essentially on the quality of the initial surgical excision. We report the case of recurrent vulvar dermatofibrosarcoma treated by iterative surgery. Our aim was to highlight the local aggressiveness of this type of DFSP and point out its management challenge, usually requiring a wide and aggressive surgery.

CASE REPORT

A 58-year-old postmenopausal women, G5P5, has been treated twice with local excision for a recurrent DFSP of the vulva, presented a local recurrence. Patient stated that mass had been present for at least three years, and had grown slowly with more recent bothersome growth. Physical examination revealed a large, fungating mobile left groin mass measuring 15 x 10 cm involving the mons pubis and distorting the left labia majora, rounded and ulcerated in many areas. On palpation, the lesion had an elastic but firm consistency, it was adherent to the skin and mobile in relation to the deep plane (Figure 1 A/B). The rest of the physical examination was unremarkable. Preoperative magnetic resonance imaging was performed and revealed a multi-lobulated vulva/left groin mass measuring 10 x 10 x 14,3 cm. A slightly enlarged bilateral inguinal nodule were also present, but no other lymphadenopathy or evidence of metastatic disease was noted (Figure 2 A/B). The surgical resection was complete, passing to 5 cm laterally of normal tissue and removing the first anatomical barrier in depth, saving normal tissue and preserving the important structures of the perineal region (Figure 3/A). Post-operatively, the patient had an uncomplicated course (Representative photo of thirty six-month follow

up are provided in Figure 3/B). Her final pathology revealed a tumor process organized in bundles and areas without glandular structure, made of spindle-shaped cells with elongated sinuous nuclei moderately increased in size, irregular with some mitotic figures (1 to 3 mitoses /10 high-powered fields (HPF)).

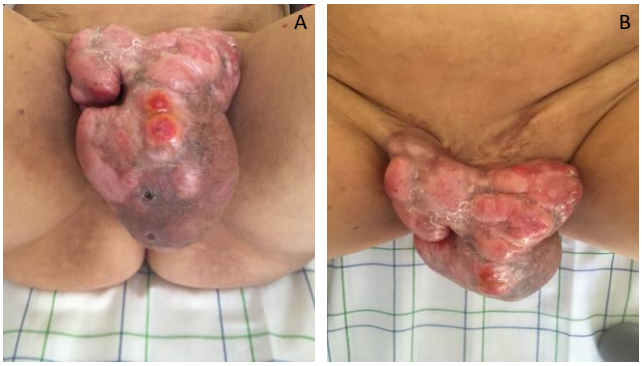

Figure 1 : Clinical features of large vulvar tumor.
A : Clinical appearance of the tumor involving the upper labia majora.
B : Superior view of the tumor extending to mons pubis.

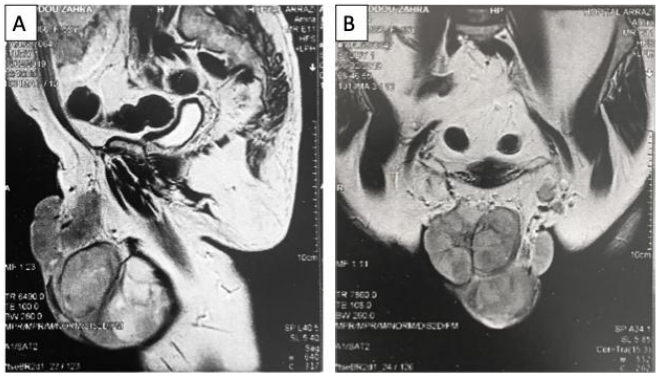

Figure 2 : Representative T1 MRI pelvis image of the tumor.
A : Parasagittal section B : Axial section

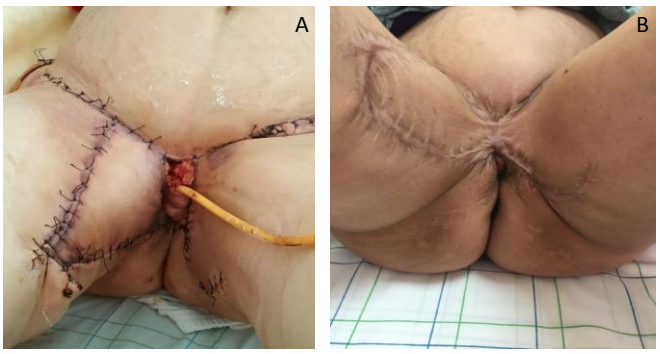

Figure 3 : A : Representative photo of immediate post op
B : Representative photo of thirty six-month follow up

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a rare low grade soft tissue malignancy representing 0.1% of all cancers, 2 to 6% of all soft tissue sarcomas [1]. The clinical diagnosis is not easy by its unusual clinical appearance. However, the recurrence of the tumor is a guide, but the histopathology confirms the diagnosis. In fact, the dermatofibrosarcoma affects preferentially young to middle-aged adults [3]. The DFSP can grow anywhere on the integument but certain localizations are more frequent: on the trunk or proximal extremities [1,3]. Vulvar localization remains exceptional with around sixty cases reported in the literature [4].

Clinically, DFSP is revealed as an asymptomatic plaque, purplish or brownish red in color, on which gradually appears a nodular lesion confined to dermal or superficial subcutaneous tissue but usually mobile on the deep planes [4].

In all cases, the diagnosis is based on a set of arguments consisting of the evolution of the tumor, the strict dermal site of proliferation and the histological aspect of the tumor (spindle cytological configuration, swirling architecture, hypervascular stroma and perinervous flows). Immunohistochemistry, currently routine, often targets positive labeling for the CD34 antigen and vimentin, but these markers are not specific [4]. As for the paraclinical assessment, it is part of the extension tests. It is essentially a radiological assessment. Computed tomography (CT) or magnetic resonance imaging (MRI) can provide information on the extent of the tumor [5].

In the case of a recurrent vulvar dermatofibrosarcoma protuberans, the practitioner is confronted with a certain number of essential constraints in the management [6]. The first constraint is a radical local resection with negative margins of 3 to 5 cm of normal tissue, making it possible to remove the tumor [6]. The second constraint concerns the vulvar reconstruction following large resection [6]. Finally, there is the choice between an exclusive surgical treatment, an exclusive chemotherapy or the chemotherapy-surgery combination in order to minimize the risk of recurrence [6]. Several studies have been conducted to evalute the efficacy of chemotherapy in the management of these tumors, especially in the context of recurrence [7]. For surgical treatment, teams using the Mohs technique have shown that margins of 3 cm are sufficient to remove all of the tumor cells [8]. Indeed, the frequency of recurrences depends on the margins of the excision: 70% for margins of 1cm, 40% for margins of 2cm, 10 to 20% for margins of 3cm, 5% for margins of 4cm and 1.75% of cases for margins of 5cm [9].

From a molecular cytogenetic perspective, DFSP shows two types of karyotype anomalies with a supernumerary ring chromosome type r (17;22) or a translocation t(17;22)(q22;q13), which produces a protein allowing the fusion of the collagen gene of type I alpha1 (COL1A1) with the gene of the chain β of the growth factor PDGFβ (Platelet-derived growth factor of group B) [3]. This rearrangement leads to a disruption of the expression of PDGFβ with the continuous activation of the PDGF receptor tyrosine kinase which stimulates the growth of DFSP [3]. Cells transformed with the COL1A1–PDGFβ fusion gene as well as cell lines derived from patients with DFSP are growth-inhibited by the tyrosine kinase inhibitor imatinib [3]. Currently, imatinib is the targeted therapy indicated as first-line treatment in the case of inoperable, metastatic dermatofibrosarcoma or in the case of a recurrent form after several surgical excisions [3]. A starting dose of 400 mg once a day is sufficient and better tolerated for patients with translocation t(17;22) [3]. As for radiotherapy, most of the literature defines Darier-Ferrand dermatofibrosarcoma as a radio-resistant tumor [10]. However, Others, have argued that radiation therapy decreases the rate of local recurrences [11]. According to Haas and col, Local tumor control was 82% with adjuvant radiotherapy for insufficient or invaded excision margins, with a follow-up of 1 to 22 years [11].

Vulvar reconstruction following large resection represent challenging cases for surgeon, it may require myocutaneous flap coverage. Traditional options for perineal reconstruction include both transposition flap such as a V-Y advancement flap, and myocutaneous flaps such as the vertical rectus abdominis (VRAM) [12]. In our case, given the substantial size of the defect a V-Y advancement flap was chosen which allowed for satisfactory coverage. Monitoring flap viability by assessing adequate blood flow and preventing infection with good hygiene are the most crucial aspects of the immediate post-operative care for these patients [12].

CONSLUSION

DFSP is an uncommon skin sarcoma. Vulvar localization is rare. The diagnosis of certainty is histological and immunohistochemical. The treatment consists on a wide surgery of the lesions with margins of excision of 3 to 5 cm, on which the prognosis mainly depends. Vulvar reconstruction following large resection is a real challenge for the management of recurrences.

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