



Review

Transformed dermatofibrosarcoma protuberans: A series of nine cases and literature review

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Abstract

The fibrosarcomatous variant of dermatofibrosarcoma protuberans (DFSP) represents an uncommon form of DFSP which has a higher risk of local recurrence and distant metastases than ordinary DFSP.

The aim of our study is to investigate clinicopathologic characteristics, treatment modalities and prognostic factors of nine cases of transformed DFSP admitted in Salah Azaïez Institute between 2002 and 2009.

They were five men and four women. Median age at diagnosis was 52 years (35–87 years). The lesions were located on the abdominal wall (three cases), the upper limb (two cases), the back (two cases), the lower limb (one case) and the chest wall (one case). Tumor size ranged from 25 mm to 150 mm. After diagnosis, six patients were treated by wide local excision with margins ≥ 2 cm, two patients had local excision without defined margins and one patient had incomplete local excision. Three patients underwent radiotherapy because of either cramped or unknown limits. Local recurrence was diagnosed in 5 cases and distant metastasis occurred in one patient.

Fibrosarcomatous DFSP exhibits more aggressive behavior than DFSP. Their similar clinical presentation requires histopathological differentiation for prognosis. Treatment is based on wide local excision, radiation and targeted therapy.

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Keywords: Dermatofibrosarcoma protuberans; Fibrosarcoma; Surgery

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1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive fibrohistiocytic tumor of intermediate malignancy with a great potential of recurrences and a small risk of distant metastasis. The fibrosarcomatous variant of DFSP represents an uncommon form of DFSP in which the prognosis is closely linked to the fibrosarcomatous component.

The aim of our study is to investigate clinicopathologic characteristics, treatment modalities and prognostic factors of 9 cases of transformed DFSP.

2. Methods

We have compiled nine patients from Salah Azaiez Institut of Tunisia during the period from 2002 to 2009. Those patients had histologically proven fibrosarcomatous dermatofibrosarcoma protuberans (FS-DFSP).

Treatment modalities and outcomes were analyzed retrospectively.

3. Results

We analyzed the clinicopathologic features of a series of nine patients presenting FS-DFSP (Table 1). Six patients have been treated initially in other hospitals and three were seen first in our department.

They were five men and four women, and their age at diagnosis ranged from 35 to 87 years (mean, 52). All patients had a history of a slowly growing painless mass.

The lesions were located on the abdominal wall (three cases), the upper limb (two cases), the back (two cases), the lower limb (one case) and the chest wall (one case). Tumor size ranged from 25 mm to 150 mm (mean, 86 mm).

In five patients FS-DFSP occurred as a recurrence of classic dermatofibrosarcoma, and the number of recurrences varies from 1 to 6. The remaining patients presented this type of lesion for the first time.

After diagnosis, six patients were treated by wide local excision with margins ≥ 2 cm, two patients had local excision without defined margins and one patient had incomplete local excision. Three patients underwent radiotherapy because of either cramped or unknown limits.

Follow-up ranged from one to 144 months. Recurrences occurred in five cases at a median of 2.4 years. All these patients were treated by wide local excision, and one of them had local radiotherapy. In one case, lung metastasis was diagnosed and treated by chemotherapy. The patient was lost of view in poor condition.

4. Discussion

The DFSP affects predominately adults between 20 and 50 years. A slight male predominance has been reported (Rutgers et al., 1992), as shown in our study; 5 men and 4 women. There is no difference in anatomic location between DFSP and fibrosarcomatous DFSP. Similar to our series, it is seen mainly on the trunk (42–72%) and the proximal extremities (16–30%) (Lemm et al., 2009).

The fibrosarcomatous transformation of DFSP is exceptional. In 1951, Penner reported a case of metastasizing DFSP containing areas that were histologically indistinguishable from fibrosarcoma (Penner, 1951).

As FS-DFSP is clinically indistinguishable from ordinary DFSP, its diagnosis is established histologically. It is characterized by more spindle cells, greater number of nuclei and increased mitotic rate. Immunohistochemically, a strong CD34 positivity is an important feature for the diagnosis of DFSP. However, in the sarcomatous cases, the CD34 reactivity can be weak and this can be explained by the loss of expression of CD34 during the tumor differentiation (Weiss and Goldblum, 2001).

The standard therapeutic approach used for the treatment of the DFSP is wide and deep local excision including the underlying fascia with margins ≥ 2 cm (Voth et al., 2011). These margins have been respected in 6 patients in our series. Some authors suggest the use of Moh's micrographic surgery in order to achieve negative resection margins and simultaneously preserve the uninvolved tissue from resection (Llombart et al., 2011).

The extent of the initial resection is the most significant prognostic factor for relapse. In our study, five patients presented local recurrence. In four cases, margins were ≥ 2 cm in anatomopathologic examination. Ding and al suggested a high recurrence rate (89%) in cases of fibrosarcomatous transformation, but the adequacy of surgical excisions was not made clear (Ding et al., 1989). Mentzel et al. and Pizarro et al. documented a recurrence rate of 59%, but a minority of the patients received wide local excision and treatment details were not provided (Mentzel et al., 1998). One of the largest studies by Goldblum et al. found no prognostic differences between conventional DFSP and DFSP containing sarcoma treated by wide local excision (Goldblum et al., 2000). These desperate results are due to small sample sizes and inadequate treated patients included. In a retrospective review of 122 patients with DFSP at the University of Texas MD Anderson Cancer Center in Houston, fibrosarcomatous change was significantly associated with local recurrence and patients with a higher percentage of FS change (25%) carried a

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