Unusual presentation of dermatofibrosarcoma protuberans: 
a large, high-grade, skull eroding tumor – Case report

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Introduction

Soft tissue and bone sarcomas in the head and neck are rare tumors, with an incidence around 0.7 cases/100,000 inhabitants. These cases are distributed among at least 10 main histologies. Dermatofibrosarcoma protuberans represents 1% of these lesions\(^1,2\). It is a slow-growing, locally aggressive fibrous tumor, which has a pronounced tendency to local recurrence\(^1\). It rarely metastatizes to regional lymph nodes or distant sites. Recognition of this tumor is important because of the excellent prognosis after adequate surgical excision, once ensured adequate margins of 3 to 5 cm.\(^3,4\)

Abstract

Dermatofibrosarcoma protuberans is a rare neoplasm, usually presenting as a low-grade, non-invasive, slow-growing tumor, with a pronounced tendency to local recurrence. We present a rare case of a high-grade, histologically undifferentiated lesion, which eroded underlying bony structures. A wide local excision with careful pathologic analysis of margins was performed with immediate reconstruction of skull bone and musculo-cutaneous flap transposition in order to cover the skin defect. A one-stage surgical procedure, such as presented in this report, is the treatment of choice recommended by the authors.

Key-word

Tumor cytology, dermatofibrosarcoma protuberans, fine needle aspiration, recurrence, surgical resection.

Resumo

Relato de caso de uma apresentação rara de dermatofibrossarcoma protuberans: um grande tumor, de alto grau, erodindo o crânio
Case Report

A 72 year-old man presented to the Emergency Room of Hospital da Clinicas of São Paulo University Medical School with a history of a 4-years’ progressive growing lesion located in the temporo-parietal region. The mass was non-tender, non-transluminescent and non-pulsatile. CT scan showed a left parietal heterogeneous lesion (Figure 1). 3D reconstructions demonstrated erosion of the underlying skull bone (Figures 2 and 3). Fine needle aspiration was proceded in order to obtain tumor cells for microscopic analysis, which suggested the diagnosis of a dermatofibrosarcoma of undifferentiated characteristics.

Microscopically, there were fascicles of spindle shaped cells (fibroblasts) arranged in storiform pattern. Surgery was proceded with total resection of the tumoral mass and craniectomy, followed by immediate reconstructive cranioplasty. Two muscular flaps were transposed to the fronto-temporal area in order to cover skin defect. Skin grafting was done over donor area. The patient had no further symptoms and was discharged in 3rd postoperative day. There was no evidence of regrowth after seven months of follow-up.

Discussion

Epidemiology

Hoffmann was the first author to describe dermatofibrosarcoma protuberans. Other terms used to describe this neoplasm are hypertrophic morphea, progressive and recurrent dermatofibroma, fibrosarcoma of skin and sarcomatous tumor resembling keloid. Age at onset of the disease ranges from 6-65 years, with maximum incidence in 2nd and 3rd decades. The majority of patients are under 40 years of age at the time of diagnosis. The tumor has marked predilection for the trunk, although no anatomical region is spared except for the hands. Head and neck regions are rarely involved. Some authors, however, reported...
higher incidence of the tumor in head and neck region and lower limbs, implicating trauma as the probable cause.

**Treatment**

The treatment of choice for dermatofibrosarcoma protuberans is wide surgical excision (ensuring adequate margins of 3 to 5 cm). Most of the authors recommend, as best therapeutic choice, a single staged radical excision of scalp and underlying bone with immediate cranioplasty and free musculo-cutaneous flap coverage of skin defect.

In fact, the ability to provide single stage reconstructions of large scalp defects and underlying cranium represents a considerable advance of contemporary reconstructive plastic surgery.

**Recurrence**

Local recurrence rates of dermatofibrosarcoma protuberans following simple excisions may reach 50%. Local recurrence is thought to occur due to local infiltrative capacity of tumoral cells. In fact, the dense connective tissue at the periphery of the tumor may give a false appearance of encapsulation when the real tumor, actually, extends beyond the perceived margins, as demonstrated by intraoperative microscopic analysis of tumoral bed biopsies.

**Metastasis**

In some cases, this kind of tumor remains unchanged over long periods of time. It is possible that any manipulation in the form of inadequate excision (such as cutting the tumor and simultaneously opening vascular channels) may be the cause of vascular or lymphatic tumoral spread.

Distant hematogenic metastases are seen in 4-6% of cases, while metastasis through lymphatic route may reach 10%. Nevertheless, metastasis is not a justification for elective prophylactic lymph node resection in all the cases. It is important, however, that lymph nodes be carefully examined during a long period of follow-up. When regional lymph node metastasis is discovered, in the absence of systemic metastasis, radical regional lymph node dissection may be attempted.

**Conclusions**

Distinguishing dermatofibrosarcoma protuberans from other spindle cell tumors and fibrohistiocytic lesions may pose significant challenges to the pathologist. However, in the appropriate clinical setting and applying strict diagnostic criteria, fine needle aspiration cytology may a reliable tool in establishing the initial diagnosis of this neoplasm and orientate an appropriate surgical approach to the lesion.

Although most of cases of dermatofibrosarcoma protuberans are low-grade, the surgeon must be aware of the possibility of a high-grade undifferentiated neoplasm, which may even erode the underlying bony structures as in the reported case.

**References**


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