

Recurrent dermatofibrosarcoma protuberans treated with Mohs micrographic surgery

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SUMMARY

We present the case of a 68-year-old woman with a recurrent dermatofibrosarcoma protuberans on the neck and the upper back, surgically treated four times over a yearlong period. Neither radiotherapy nor chemotherapy have been effective. After the last recurrence the Mohs micrographic surgery has been applied. The tumor has been excised and cut in a considerable number of fragments, which have then been examined microscopically. A second procedure has been necessary to completely remove the tumoral mass. Four months later there have been no signs of local recurrence any more. The Mohs micrographic surgery is the treatment of choice for dermatofibrosarcoma protuberans, since classical surgical excision would have been, as our case reveals, difficult and vain.

KEY WORDS

dermatofibrosarcoma protuberans, treatment, micrographic surgery, Mohs

Introduction

The dermatofibrosarcoma protuberans is a locally aggressive and highly recurrent malignant neoplasm of dermal origin. Histologically, it is composed of spindle cells resembling more or less mature fibroblasts (1, 2).

The tumor occurs in middle-aged persons, most frequently on the trunk. Initially, it appears as a unique nodule that evolves later in a multinodular indurated plaque, usually flesh-colored, reddish or blue. The clinical course is characterized by a painless, progressive and irregular growth. In the advanced phase it may become painful and ulcerated. Metastases have also been described, but they are rare and may occur after the repeated recurrences (1, 2).

The differential diagnosis in the early phase includes a dermatofibroma and keloid. In the advanced phase the tumor's slow growth rate, a characteristic color and irregular margins suggest the diagnosis dermatofibrosarcoma (1).

In general, wide surgical excision is considered necessary to avoid reappearance of the tumor. Recurrence rate using undefined surgical margins is estimated from 49 to 53%. It drops to 10-20% when wide surgical margins (3 cm of clinically uninvolved tissue) reaching down to the superficial fascia, are used. Such procedure may lead however to a significant functional and aesthetical impairment (1). Chemotherapy is not indica-

ted being reserved for metastatic disease (1, 2). Radiotherapy has a limited role; it may however be useful, combined with surgery when margins are positive or even alone in cases of non-operable tumors (3–5).

Case report

A 68-year-old woman was affected by dermatofibrosarcoma protuberans extending from the posterior cervical region to the left upper part of the back (Fig. 1). Initially it appeared as an asymptomatic, flesh-colored, rapidly growing nodule, 2 cm in diameter.

The tumor was completely excised and the histopathology confirmed the clinical diagnosis. Three months later, the patient underwent a further surgical intervention because of a postoperative recurrence. Three months afterwards, as the tumor rapidly recurred, radiotherapy with photons of a linear accelerator (4 MeV) was applied. The complete dose amounted to 6000 cGy divided into 30 fractions, with single dose of 200 cGy per fraction. This treatment was not successful; thus the patient underwent a new surgical excision, as there was again evidence of tumor's recurrence. Three months after the last operation chemotherapy with Adriamycin (60 mg/m², 110 mg once weekly) and dacarbazine (220 mg/m², daily dose 400 mg, for five consecutive days, every fourth week) repeatedly for six cycles was introduced. There was no sign of tumor regression.

For this reason, three months after the chemotherapy, Mohs micrographic surgery has been performed. The tumor has been excised and divided into six large fragments. Due to their large size, each one has been cut into nine smaller specimens and examined microscopically, by multiple horizontal sections from the base to the epidermis in order to identify the persisting foci of tumoral microinvasion.

The tissue was fixed in 10% buffered formalin, conventionally processed and embedded in paraffin (wax). Sections 4 μm thick were stained with haematoxylin and eosin. Representative slides were studied by immunohistochemistry using an automated immunostainer with the avidin-biotin-peroxidase method. Antibodies used were CD34 and S-100 protein.

Histologically, the neoplasm was composed of densely packed, monomorphous spindle cells arranged in a storiform pattern. Small fibrosarcomatous-like areas were seen at the periphery of the lesion, where the tumor was often characterized by a fascicular growth pattern of spindle cells or by a honeycomb-like pattern (Fig. 2). Only scattered mitoses were observed. Immunostaining for CD34 resulted focally positive. The tumor was entirely negative for S-100 protein.

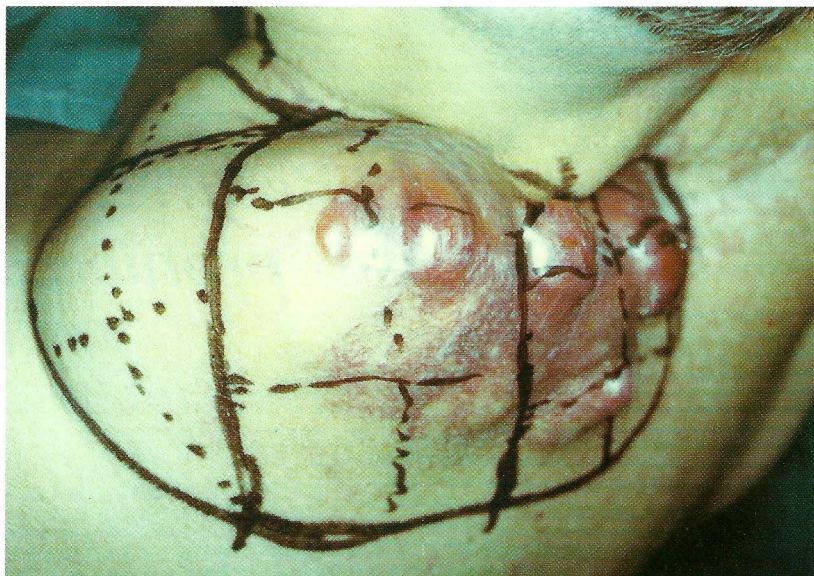
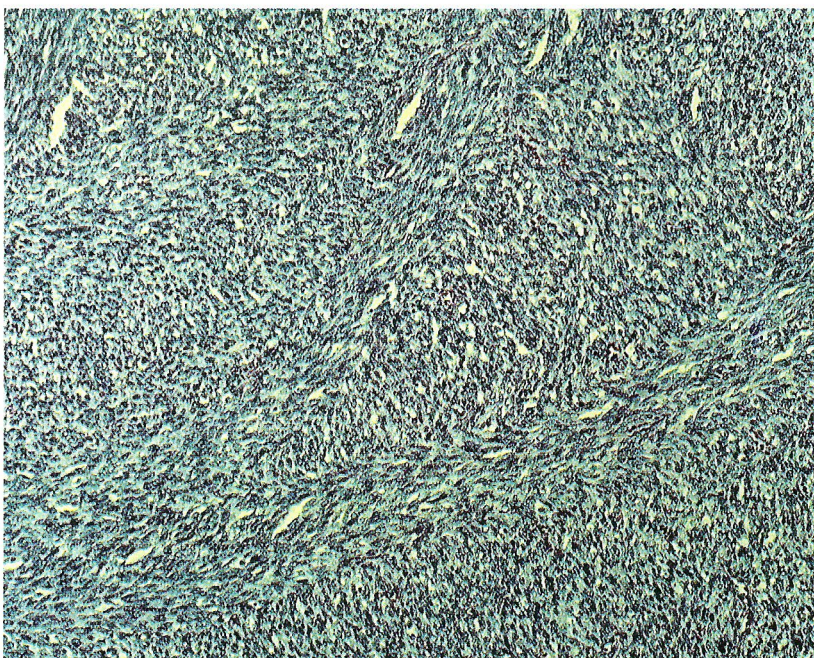


Figure 1. Recurrent dermatofibrosarcoma protuberans on the neck and the upper back: preoperative appearance.

Figure 2. Irregular strands of spindle cells with monomorphous nuclear pattern. No atypical mitoses are evident. H. E. x 100.



Two areas of deep tumoral involvement have been found (Fig. 3).

The second and ultimate excision, necessary to remove selectively these two areas, has been done. The surgical wound has been closed with a rotation-advancement flap. Four months later there is no evidence of local recurrence (Fig. 4).

Discussion

The Mohs micrographic surgery is an excisional technique with complete histological margins' control. This technique allows the complete removal of the neoplasm, examining all its margins, while conserving as much uninvolved tissue as possible (6, 7).

It is the most effective surgical treatment for the epithelial and mesenchymal neoplasms, characterized by contiguous growth, either primary or recurrent, having indistinct clinical margins and found in particular, high-risk areas. Particularly aggressive and extended tumors may need wide and destroying excisions for they may infiltrate other organs and vital tissues. Tumors, recurring after surgical excision or irradiation, may show unpredictable subclinical growth within and around scar tissue from previous treatments (8).

The advantages of this method are related to the high oncologic effectiveness and the maximal normal tissue saving.

The tumor is excised and cut into several pieces. The proper orientation of fragments is preserved by putting references on the wound's edge and by color-coding specimens' edges with different dyes (9, 10).

Figure 4. Result at four months postoperatively.

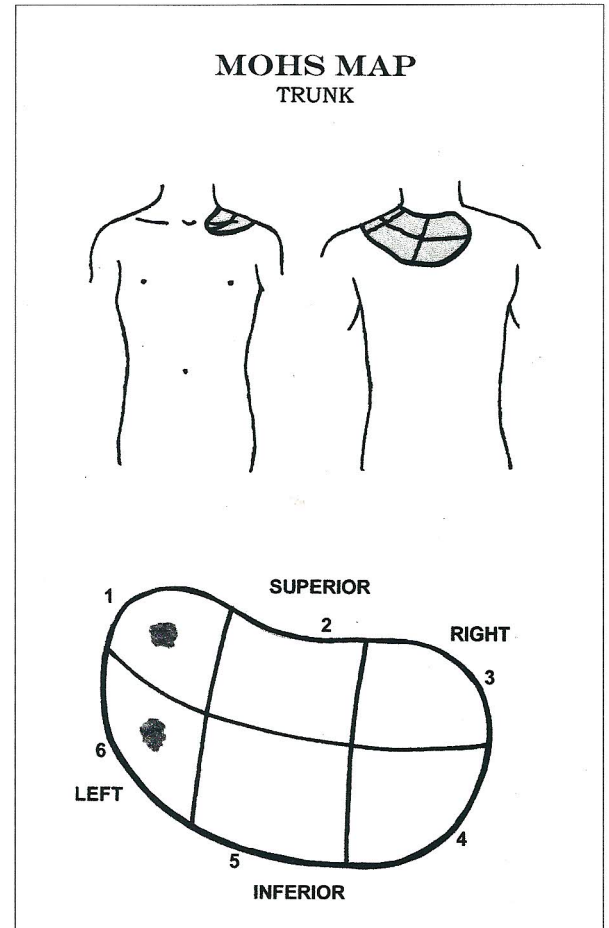


Figure 3. Mohs map: tumor divided into six large fragments and persistence of tumor in two areas.

After those specimens are placed upside down, the edges are teased on the same plane as the basement and a microscopic examination of the horizontal sections is performed. If there is evidence of persisting tumoral islets, they can be localized on the surgical wound and selectively removed. Thus, only the residual neoplastic areas are excised while healthy surrounding tissue is maximally spared. The procedure continues step by step until the entire tumor is excised and a three-dimensional view of the whole neoplasm with its microscopic extensions may be reconstructed (9, 10).

Conclusion

Although the follow-up period is quite short, it appears rather significant in view of the previous rapidly recurrent behavior of the tumor.

Data from the literature suggest that Mohs micrographic surgery is the treatment of choice for dermatofibrosarcoma protuberans. Therefore, it should always

be the treatment of choice in order to avoid recurrences and complications.

Our case confirms that Mohs micrographic surgery can reduce the recurrence rate that is rather high after classical surgical excision.

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