Dermatofibrosarcoma Protuberans of the Face: A Case Report

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Abstract

Background: Dermatofibrosarcoma protuberans (DFSP) is a rare locally aggressive tumor with high rate of local recurrence but low metastatic potential. Its occurrence in the head and neck is a rare finding but it is associated with increased morbidity and mortality.

Methods: A 53-year-old Iranian man referred to a complaint about an increase in the size of the subcutaneous mass on the right cheek that had been present since he was 17 years old. On clinical examination, the lesion was not painful and had no erythema. In examining the patient's CT scan without contrast, a mass-like area with soft tissue density was seen. During the surgery, the flap was lifted after a lateral rhinotomy incision. The biopsy sample was sent for frozen section, and the pathology result was a Spindle cell tumor. In immunohistochemistry (IHC), two factors, including CD34 and VIMENTIN, were positive and Dermatofibrosarcoma protuberans (DFSP) was reported.

Conclusions: The most important prognostic factor for surgery of Dermatofibrosarcoma protuberans is obtaining tumor free surgical margins. As a result, accurate diagnosis and correct management of this uncommon and locally invasive malignancy is very important, especially in head and neck surgery.

Keywords: DFSP, Dermatofibrosarcoma Protuberans, Head and Neck, Cancer

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, low to intermediate-grade malignant tumor, a type of soft tissue sarcoma originating from the dermal layer of the skin, especially located in the maxillofacial region [1]. It is a locally aggressive but rarely metastatic skin tumor, which tends to recur after excision. Although historically it has been attributed as fibroblastic in origin, recent immunohistochemical evidence suggests that it may arise from dendritic cells in the skin [2].

It is presented as a slow-growing indurated plaque on which nodules develop over time. The lesion arises in the dermis but can invade subcutaneous tissue, fascia, muscle, and even bone. COL1A1-PDGFB translocation is specific to dermatofibrosarcoma protuberans, and the presence of this fusion contributes to diagnosis in certain cases [3].

This tumor has been reported to involve many body surfaces, mainly the trunk, followed by the extremities, and less commonly in the head and neck [4]. This article describes a case of dermatofibrosarcoma of the face in a 53-year-old male patient who was referred to the Otorhinolaryngology Department of Baqiyatullah University of Medical Science.

Case report

A 53-year-old Iranian man referred to a complaint about an increase in the size of the subcutaneous mass on the right cheek that had been present since he was 17 years old. The mass, which extended from the nasofacial groove to near the medial punctum, had a gradual and slow growth. On clinical examination, the lesion was not painful and had no erythema. The

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consistency of the mass was firm and sticks to the skin (Figure 1).

In the medical history of the patient, he had no history of any previous illness or use of any special medicine and no history of sensitivity. There was no history of cancer in the family history. In a habit review, he had no history of alcohol or tobacco use. In examining the patient's CT scan without contrast, a mass-like area with soft tissue density was seen. The approximate size of the lesion was 25 x 30 mm, which was located in the right infraorbital area and extended to the nasofacial groove and cheek. Also, soft tissue swelling without encroaching on the adjacent bone was noted (Figure 2).

Before the surgery, informed consent was obtained from the patient and wide local excision was planned for treatment. During the surgery, the flap was lifted after a lateral rhinotomy incision with extension of the nasolabial fold in the lower part of the mass and extension into the path of the lower eyelid to the upper part of the mass. The biopsy sample was sent for frozen section, and the pathology result was a spindle cell tumor. Also, a part of the skin attached to the mass was removed by the tumor. The length of the mass was more than what was detected in the CT scan, and it was about 7 cm long. The tumor was relatively adherent to the surrounding tissue and did not have a clear capsule, but it was completely resected and sent for pathology examination (Figure 3). The defect was repaired with an advancement flap in two layers (Figure 4A).

In immunohistochemistry [IHC], two factors, including CD34 and VIMENTIN, were positive and (DFSP) was reported. After the surgery, a consultation was done with the radio-oncology service and a decision was made for radiotherapy, and 33 sessions of radiotherapy were performed on the patient on a daily basis.

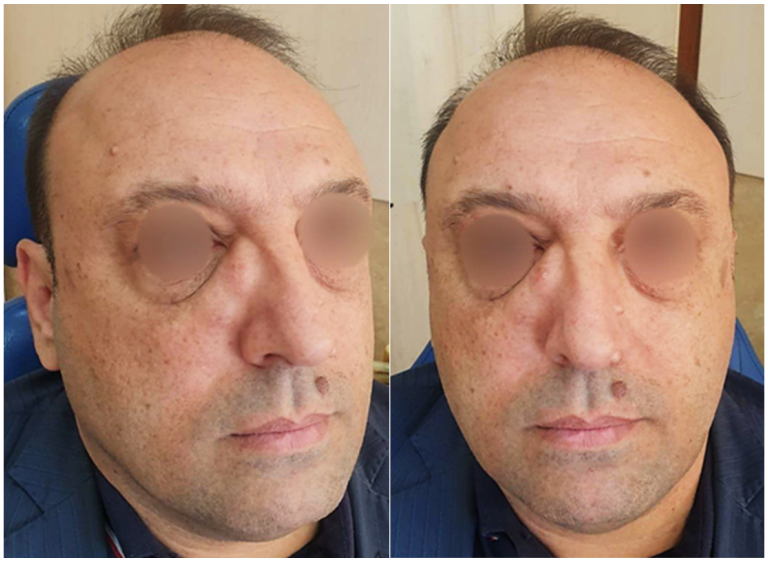

Fig. 1: Pretreatment

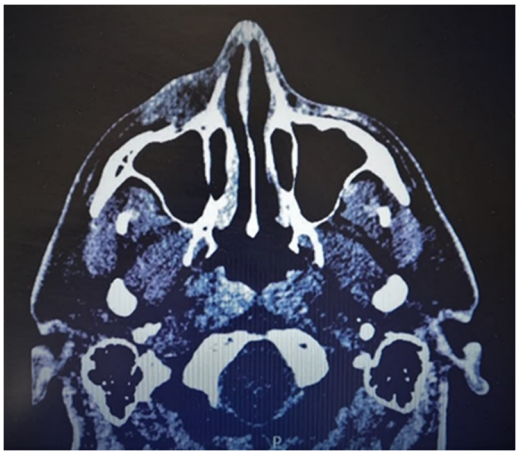

Fig. 2: CT scan, axial view

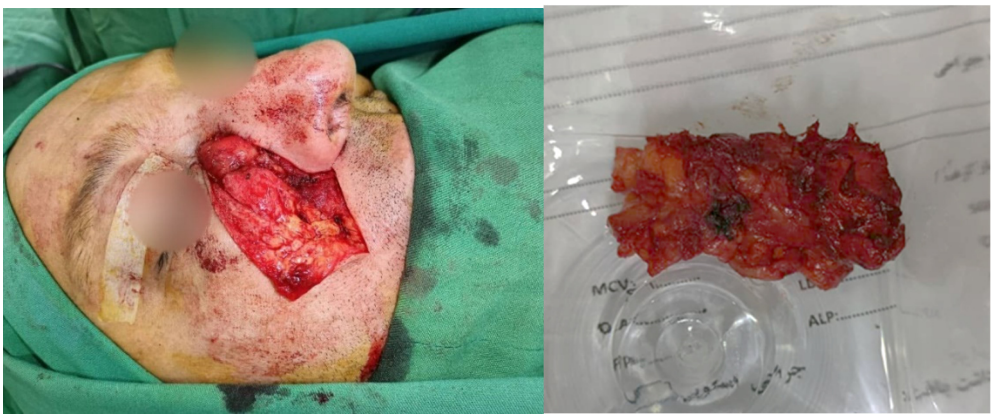

Fig. 3: At the surgery, after the flap was lifted. The tumor was completely resected and sent for pathology examination

Fig. 4: A. Immediately after surgery. B. After about a year of surgery

Discussion

DFSP is a rare soft tissue malignancy occurring most often on the trunk and less commonly in the head and neck. DFSP accounted for approximately 1.8% of all soft tissue sarcomas encountered. Persons aged 24 to 50 years have the highest likelihood of developing this tumor, which occurs in 80% of this group. Also, DFSP in the head and neck occurs more frequently in males. Overall, local recurrence rates have been reported as high as 60%, although the rate of distant metastasis is extremely low [5].

USPSTF arise as pink or violet-red plaques, and the surrounding skin may be telangiectatic. These lesions typically are fixed to the dermis but move freely over deeper-lying tissue, and they do not exhibit a nodular growth pattern until late in their course. Fixation to more deeply seated structures is often observed in advanced and/or recurrent cases of DFSP [6].

DFSP is a rare monoclonal cutaneous soft tissue sarcoma that was first described by Taylor in 1890 [1]. Histologically, the tumor is highly cellular and composed of monomorphic, fusiform cells with elongated nuclei showing little or no pleomorphism or hyperchromasia. The cells have little cytoplasm, which usually appear amphophilic to eosinophilic. They are arranged in irregular interwoven fascicles forming a storiform pattern. Lack of multinucleated giant cells and the presence of entrapped isolated fat cells are other diagnostic features [7].

The extensive excision of DFSP brings about an aesthetic challenge for surgeons. The treatment for DFSP is wide local excision with histologically negative margins [8]. There is a high local recurrence rate after inadequate excision [9]. But in this case, after about a year of surgery, the lesion has not had recurrence and the site of that has been healed favorably (Figure 4B).

Conclusions

DFSP is a locally aggressive tumor with a high rate of local recurrence but low metastatic potential, and it is a rare skin malignancy. This entity is more likely to recur when there is 1) a close margin of excision, 2) head/neck disease, or 3) fibrosarcomatous transformation. Wide local excision is the gold standard of primary treatment, while radiotherapy is recommended for recurrent disease. The most important factor for determining the prognosis of DFSP is obtaining tumor-free surgical margins. As such, accurate diagnosis and correct management of this uncommon and locally invasive malignancy are very important, especially in head and neck surgery.

Consent

The patient provided written consent for treatment and informed consent for the use of the data for scientific purposes.

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Conflict of Interests

The authors declared no conflict of interests.

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