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Dermatofibrosarcoma protuberans: A clinical presentation and a surgical treatment

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Abstract
Dermatofibrosarcoma protuberans (DFSP) is a rare nosological entity from the group of soft tissue sarcomas. The etiology of the disease remains unclear. The neoplasm may start growing de novo over healthy skin or as a result of a local chronic trauma, radiation therapy or thermal scar. It affects more commonly middle aged patients with a clear male prevalence. Once developed, the tumor progresses slowly with an indolent growth pattern and might be wrongly taken as a benign neoplasm. However, DFSP has a high local recurrence level and intermediate metastatic activity reported. Surgical excision of the tumor is the most effective treatment method. In the following we present clinical case reports of patients with DFSP.

Keywords: Dermatofibrosarcoma protuberans, soft tissue sarcoma, indolent growth pattern

Introduction
Sarcomas in the maxillofacial region are a diverse group of mesenchymal neoplasms. They originate from cells that have undergone malignant transformation [1]. Aggressive biological behavior, a tendency for local invasion, and high metastatic potential are typical for sarcomas [2, 3]. Sarcomas usually have a nonspecific clinical presentation, as swelling, growing a painless mass, pain appear to be major symptoms [4, 5]. The classification of sarcomas is based on their tissue origin and is divided into bone sarcomas (BS) and soft tissue sarcomas (STS). Approximately 80% of all sarcomas are derived from soft tissue components [1, 6]. Dermatofibrosarcoma protuberans is a rare entity of soft tissue sarcomas of the head and neck.

DFSP is a superficial developing mesenchymal neoplasm that arises in the dermis and subcutaneous layers [7]. The disease is uncommon with an overall frequency of 4-5% of all sarcomas in the maxillofacial region and 5% of all soft tissue sarcomas [8, 9, 10]. The age distribution for DFSP ranges with the majority of middle aged patientswith an average age of 31 years. Males are more affected than females [11, 12]. DFSP affects mostly the extremities and the body [13]. In the maxillofacial region the sarcoma is usually seen on the scalp, the face and the neck are most [14]. Clinically, the tumor is presented as a poorly elevated, indurated or sclerotic plaque or nodules of skin with a dusky reddish-blue discoloration, which may resemble the morphea- form of basal cell carcinoma [7]. The neoplasm develops slowply over a long period of time until it becomes locally aggressive [12]. In the rapid phase processes of a vertical growth pattern begin leading to an elevated plaque with a protuberant appearance, which is fixed to the surrounding tissues [15]. In cases of DFSP with fibrosarcomatous changes a rapid growth from the very beginning is observed. In such cases the risk of metastic disease even with a wide surgical excision is 15% [16, 17]. In pure forms of DFSP without fibrosarcomatous changes, distant metastases are rare [17].

Clinical Case 1
A 30-year-old patient was admitted to our clinic of oral and maxillofacial surgery, with a history of a subcutaneous node in the scalp for more than 2 years. After medical examination by a general surgeon the lesion has been taken for a cyst and has been excised After medical examination by a general surgeon, an excision is recommended because of a suspected skin cyst, and the lesion is excised later. The histopathological examination of the material revealed the diagnose DFSP. On the clinical examination we observed an indurated plaque with a size of 30mm in greatest diameter, engaging the entire skin thickness and with

a centrally located crust (Fig. 1-A). The neoplasm clinically appears free from the underlying cortical plate. The planned excision boundaries are broadly about 3 cm wide from the periphery of the lesion (Fig. 1-B). Peripherally of the main lesion the skin appears altered and hyperemic with fuzzy boundaries. Several satellite lesions are visible at a distance from the main mass.

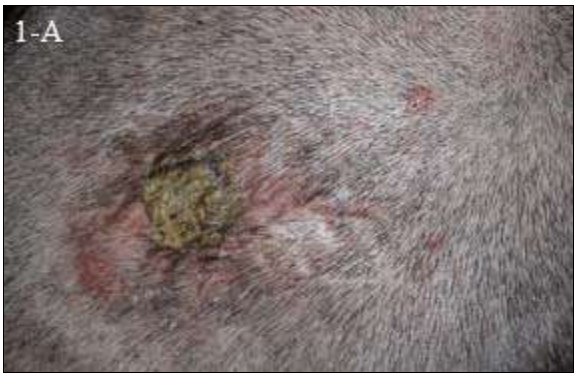

Fig 1A: An indurated plaque with a size of 30mm. A centrally located crust observed.

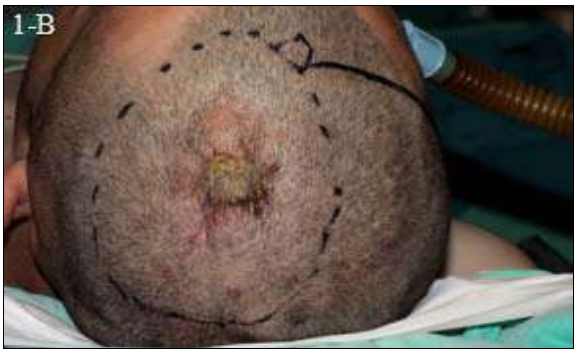

Fig 1-B: Preoperative; Resection lines are lined with excision boundaries broadly 3 cm

The excision also includes the subgaleal soft tissues, exposing the cortical plate of the underlying calvaria (Fig. 2-A). The tumor was excised with a clinically negative surgical margins (Fig. 2-B). The significant postoperative defect was reconstructed with adjacent large skin-fascial flaps; the peripheral wound defects were covered with a free skin grafts (Fig. 3). Long-term follow-up of seven years did not show any significal complications, local recurrence or metastasis.

Fig 2A: Excision of the tumor mass together with the Subgaleal soft tissues.

Fig 2B: The resected specimen with clinically negative surgical margins.

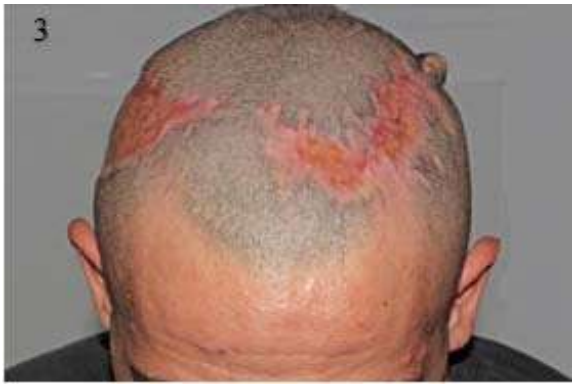

Fig 3: After the plastic reconstruction.

Clinical Case 2

A 44-year-old male patient diagnosed with DFSP on the right cheek skin is referred to the department of maxillofacial surgery for a clinical consultation. The patient reports surgical procedure 20 years on the same s ago. However, a clear diagnosis has not been established.

On the clinical presentation we observed a lesion, presented as a concave skin of the right cheek. On palpation infiltration in the adjacent soft tissues is found with a radial growth pattern. The neolasm spreads into the fibroadipose tissue resulting in lateral soft tissue deformity of the cheek, the upper eyelid and the alae nasi. This type of growth is a well-known characteristic of soft tissue sarcomas, as it results in tissue compression mimicking an encapsulated tumor process. After radical excision, a significant defect was formed, which is reconstructed with a one- step neck-based flap. Long-term follow-up of three years did not reveal any significant signs of recurrences nor metastatic activity.

Discussion

DFSP is an uncommon neoplasm of the soft tissue sarcoma group, which is classified as a low-grade tumor of malignancy [7]. The disease develops superficially from the subcutaneous soft tissues overlying the superficial fascia of the skin and may not affect the underlying structures for a long time [18]. A rapid infiltrative pattern of growth from the beginning can be seen in lesions on the scalp because of subperiosteal involvement [19]. During the active tissue infiltration phase, the lesion engages the surrounding structures and underlying fascial sheets [15] Symptoms such as pain and tenderness during the stage of active growth are

observed in 10-25% of cases [20, 21]. DFSP may develop de novo in healthy skin or in a region with local chronic trauma, scar or irradiated tissues [7, 12]. The tumor grows slowly which might be followed by misdiagnosis. The histopathological examination reveals an expansive and well-circumscribed gross appearance in the early stages [15]. Later, an infiltrative and asymmetric growth pattern with small drags extending beyond the underlying tumor mass may be seen [12]. The presence of satellite cells away from the major tumor mass is considered as a main reason for local recurrences, even when a wide 30 mm surgical excision of the tumor mass has been conducted [16, 17]. Achieving clear resection margins in the surgical treatment of DFSP is mandatory due to the presence of satellite tumor cells at a distance from the main tumor mass [12]. An inappropriate diagnosis may lead to inadequate treatment of DFSP, which is associated with an increased recurrence rate. Local recurrence rate varies in the literature, with a peak incidence of local recurrences reported in 20-55% of cases [15, 16]. Rapid growth in DFSP has been observed with fibrosarcomatous changes [16, 17].

The main treatment method of choice of DFSP is a surgical excision with achieving clear resection lines due to the tumor tendency of local recurrences [10, 12]. Often, negative resection lines cannot be achieved due to the tumor growth pattern [15, 12]. An incidence of recurrence of 5% were reported where a 5 cm excision was performed around the tumor [20]. The clinical and biological similarities of the neoplastic process to other benign or low-malignant neoplasms like keloid, basal cell carcinoma or scleroderma may lead to inadequate superficial or incomplete excision of the tumor mass, which is associated with recurrence rate of 43% reported of all cases and occurring within 3 years of the initial excision [15, 16]. In the maxillofacial region, a higher recurrence rate is reported. Achieving recommended clear resection margins in the region of the head and neck is not always possible because of the anatomical features and limitation [12]. In lesions with wide surgical margins of 2-3 cm, local recurrence rate is reported to be 18% of cases [16]. The Mohs-surgical approach is considered to reduce recurrences to rates of 2-7% [20]. The prognosis of DFSP is good in cases of operatively achieved, negative resection margins, with a 5-year survival reported in 100% of cases [21, 22, 23]. In case of positive resection margins, adjuvant radiotherapy is the method of choice [24, 25].

Conclusion

DFSP is a rare soft tissue sarcoma with low malignant potential. Its low incidence and clinical polymorphism lead to misinterpretation of the clinical signs and inappropriate diagnosis. The presence of satellite tumor cells peripheral to the major tumor mass is the reason for a high local recurrence rate. Its indolent course of growth mimics an innocent benign tumor process. However, if not treated radically, its biological behavior and histopathological features result in early recurrence. The conventional form of DFSP has e good prognosis when treated widely. In cases of DFSP with areas of fibrosarcomatous alterations the prognosis is, however, worse as a result of more aggressive course.

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