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Case Report Open Access

"It Started as a Spot"... Dermatofibrosarcoma Protuberans

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Abstract

We present a case of Dermatofibrosarcoma Protuberans (DFSP); a locally invasive soft tissue sarcoma that arises in the dermis of the skin. DFSP often starts as an asymptomatic, slow growing, pigmented, indurated/nodular area of skin. At this stage DFSP can be confused with a dermatofibroma, keloid scar, epidermal cysts and melanoma amongst others. Phases of rapid growth and bleeding occur, which prompt patient presentation. Wide local excision is the treatment of choice with close follow up due to the risk of local recurrence, however, metastasis is rare.

Keywords: Dermatofibrosarcoma protuberans; Dermatofibroma; Metastasis; Malignancy; Parotidectomy

Case Report

A 50 year old male presented with a left sided facial swelling that had been slowly growing for 20 years, with rapid growth, bleeding and ulcerated over the previous 6 months (Figure 1). On presentation the mass measured 10 × 15 centimeters, was ulcerated and intermittently bleeding. The patient reported limited pain and there was no weakness of the left side facial nerve. Initial clinical diagnosis was a primary parotid gland malignancy. Ultrasound scan and Magnetic Resonance Imaging (MRI) showed an extensively lobulated mass within the superficial lobe of the left parotid gland, with no pathological neck lymphadenopathy. Incisional biopsy of the mass showed extensive myxoid tissue.

The patient underwent a total parotidectomy, sacrificing the buccal and zygomatic branches of the facial nerve. There was no facial bone involvement and following resection, the defect was reconstructed with a left side radial forearm free flap, with skin paddle measuring 10 × 12 centimetres (Figure 2). The patient made an uneventful recovery and was discharged on day 5 post operatively. To date, 4 years post operatively there have been no signs of recurrence. Surveillance includes 6 monthly MRI scan on the recommendations of the Head and Neck Multi-Disciplinary Team (MDT).

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare low-grade malignant tumour which is locally aggressive in nature. It most commonly presents on the trunk (50%) and extremities (20-30%) with 10-15% of cases involving the head and neck [1,2]. Although only accounting for 0.1% of malignant neoplasms it does constitute up to 7% of all head and neck sarcomas [3]. Overall incidence of the disease has been estimated at between 0.8 and 5 per million per year with the most common age of diagnosis between 30 and 50.2. The incidence in the Afro-Caribbean population is almost double that of the Caucasian (6.5 and 3.9 per million respectively) [4]. Some cases of congenital DFSP have been noted although no hereditary predisposition has been found.

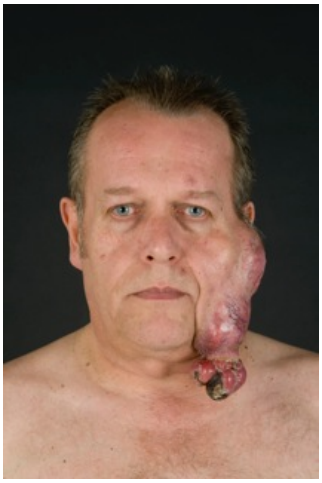

Figure 1: DFSP left side of face.

DFSP typically presents as an asymptomatic skin coloured plaque which may have been present for some time without change. It is usually a superficial lesion tethered to the overlying skin but not the underlying structures and therefore remains mobile on palpation. In more advanced cases it can become fixed to underlying muscle or fascia although this can occur relatively early in scalp lesions. Pain and tenderness are not usually associated with patient presentation [5]. It has been suggested that DFSP has a predilection for occurrence in skin which has been traumatized or irradiated as well as that which has been tattooed or contains scar tissue [6].

DFSP is commonly misdiagnosed clinically due to its asymptomatic and slow growing nature. Differential diagnoses include dermatofibroma, neurofibroma, epidermal cyst, fibrosarcoma, sarcoidosis, nodular fasciitis, nodular melanoma and scar tissue amongst others [7]. Diagnosis is most commonly made by incisional

biopsy and histological analysis. FNA does not generally yield a definitive diagnosis and is therefore avoided [2].

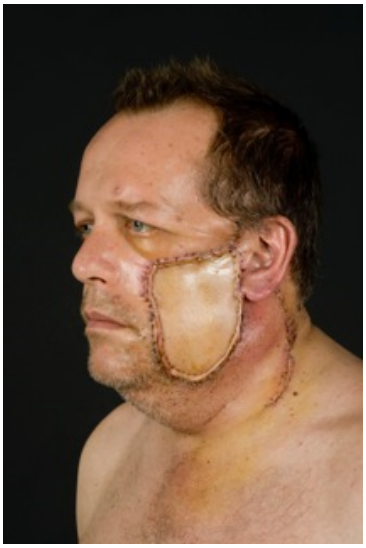

Figure 2: Day 5 post resection of DFSP and reconstruction with radial forearm free flap.

Treatment of DFSP is wide local excision with awareness that the finger like projections of the tumour can reach over 1 cm beyond the clinical margins in 70% of cases [2]. More recently Mohs micrographic surgery has been advocated as an excellent alternative [2].

Although DFSP is radiosensitive, radiotherapy is generally reserved for recurrent and large lesions. It has however been shown to decrease rate of tumour recurrence when used in conjunction with surgical excision [8]. Traditional chemotherapy is not considered a treatment option unless all other treatments have been exhausted.

Paradisi et al., found local recurrence of DFSP following WLE to be 13% in 5 years, necessitating close follow up of these patients, in the appropriate MDT setting [9]. The rate of metastasis of DFSP is estimated at between 0.5 and 5% [2].

DFSP is a rare tumour of the skin which commonly mimics other benign lesions and therefore can often be misdiagnosed at initial presentation. It continues to be treated primarily with surgery and has a cure rate of 93% with WLE and up to 99% with Mohs technique [2]. Due to the extent of the tumour beyond clinically obvious margins the surgical defect can be large and there can be a high associated morbidity in the head and neck. It is for this reason and the ability of the disease (although low) to metastasize that we present this case in order to allow for early diagnosis and treatment in order to improve prognosis for patients.

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