The silent "Cheeky" mystery: A case of indolent yet invasive dermatofibrosarcoma protuberans

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare fibrohistiocytic tumor with low-to-intermediate malignant potential. Here we described a neglected, painless and slow-growing mass on the right cheek in a 46-year-old man.

CASE PRESENTATION

A 46-year-old man, with chronic Hepatitis B infection, presented with a slow but progressively enlarging, painless mass over his right cheek for 3 years. There were no complaints of speech, chewing and pain. The patient reported no prior history of trauma or dental infection. On examination, a firm, erythematous-to-brown lobulated nodule measuring 8x5cm was noted on the right cheek, with prominent peripheral telangiectasia (Figure 1a). There were no palpable lymph nodes. Computed tomography revealed a subcutaneous mass extending from right cheek to the lower eyelid, with erosion of the right maxillary sinus. There was no regional lymphadenopathy. However, subsequent magnetic resonance imaging shows no infiltration to surrounding tissue.

Histopathological examination of the swelling demonstrated a poorly circumscribed, spindle cell proliferation arranged in a storiform pattern in the dermis, with presence of grenz zone. The spindle cells were uniform, with ovoid to elongated nuclei, fine chromatin, and scant cytoplasm. Immunohistochemical stains showed diffuse positivity for CD34, confirming the diagnosis of DFSP(Figure 2a, b).

The patient underwent wide local excision of the right cheek and partial maxillectomy, followed by reconstruction with titanium mesh, a left radial forearm free flap, and split-thickness skin graft (Figure 1b, c). Given the tumour's close proximity to the anterior maxillary wall, this structure was excised en bloc with the lesion to ensure adequate margins. A titanium mesh was placed to restore the contour of the right cheek. The orbital floor remained intact. Suspension of the lower eyelid was achieved with a palmaris longus tendon sling. In addition, a neck dissection was performed to expose suitable recipient vessels for microvascular free-flap reconstruction. Post operative histopathological study showed clear margin. Review at 18-month postoperatively showed that he was well without any clinical recurrence (Figure 1d).

DISCUSSION

DFSP is a locally aggressive soft tissue neoplasm characterized by indolent growth with high tendency for local recurrence due to its infiltrative nature.[1] Population-based studies and national cancer registry data confirm that DFSP remains a rare cutaneous malignancy, with incidence rates consistently below 1 case per 100,000 person-years.[2] Epidemiologic data from Europe and North America demonstrate relatively stable incidence over time, although a modest increase has been observed in more recent analyses. For example, national registry studies from Denmark[3] and the United States[4,5] have reported incidence rates ranging from approximately 0.4 to 0.5 per 100,000 person-years, while a recent SEER database analysis indicated a slightly higher age-standardized incidence of 0.62 per 100,000 person-years.[6] Similarly, registry data from England showed a small, non-significant upward trend between 2013 and 2019.[7] Although population-based studies and national cancer registries from Europe and North America have provided valuable insights into the epidemiology of dermatofibrosarcoma protuberans, comparable data remain scarce in many regions, including our local setting. The absence of regional or national incidence statistics limits the ability to assess disease burden, demographic patterns, and temporal trends within the local population. This lack of data likely reflects the rarity of DFSP, under-recognition, and the absence of dedicated sarcoma or skin cancer registries, underscoring the importance of individual case reports and institutional series in contributing to the existing literature and improving clinical awareness.

DFSP generally shows no strong sex-related predilection, occurring in men and women at comparable rates, though one series by Asuquo et al.[8] observed a modest male majority. The tumour is most frequently diagnosed in adults in their 20s through 50s.[9] Previous literature has also described cases arising within areas of prior injury or surgical scarring.[10] In contrast, our patient did not report any history of trauma or operative intervention at the site.

DFSP frequently erupted on the trunk (50-60%) and proximal extremities (20-30%). Head and neck involvement is less commonly reported, occurring in approximately 10-15% of all DFSP.[1] DFSP on the cheeks such in our case are extremely rare and tends to have a higher recurrence rate at 30 to 50%, attributed to the complex anatomy and the tumor's propensity to invade deeper structures, such as periosteum, skeletal muscle, and, in rarely, bone.[1]

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Fig. 1: a) Lobulated nodules on right cheek measuring 8x5cm; b) Wide local excision of right cheek, and right anterior wall maxillectomy followed by reconstruction with titanium mesh; c) Post left radial forearm free flap and split-thickness and skin graft; d) The outcome after 18 months

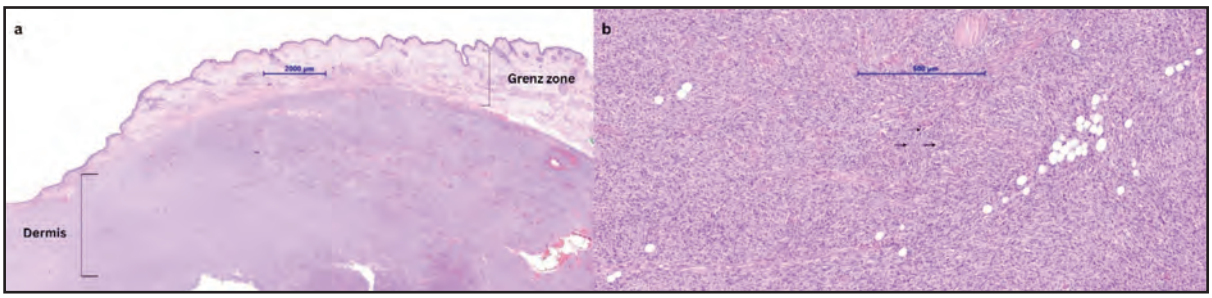

Fig. 2: a) (H&E) A circumscribed, dense spindle cell proliferation in the dermis, demonstrating a clear grenz zone, demonstrating storiform growth pattern. b) Under high power, the spindle cells are large, with ovoid nucleus, mitoses are present with clear chromatin, and high N:C ratio (black arrows)

Clinically, in its early phase, it often presents as an indurated, skin-coloured to erythematous or brownish-yellow plaque with irregular borders, typically ranging from 2 to 5 cm in size. However, these lesions may be larger and display nodular or multilobulated surfaces.[11] With progressive growth, it infiltrates deeply into the dermis and subcutaneous layers, giving rise to multiple indurated nodules that become fixed to adjacent anatomical structures, including adipose tissue, fascia, muscle, periosteum, and, in advanced cases, bone.[12] Three non-protuberant variants have been described-morphea-like, atrophoderma-like and angioma-like forms.[13] The presentation in our patient, a large plaque with multiple nodules, represents the most clinical morphology in adults.

Diagnosis of early facial DFSPs often challenging as shown in our case. It may present as long-standing painless indurated plaques or nodules, frequently misdiagnosed as benign lesions.[11] The average delay to diagnosis in such cases ranges from 2 months to 41 years, reflecting the tumor's indolent progression and clinical ambiguity.[13] The differential diagnoses include dermatofibroma, schwannoma, cutaneous neurofibroma, solitary fibrous tumor, intradermal spindle cell lipoma, spindle cell squamous cell carcinoma, and desmoplastic or spindle cell melanoma.[14]

Definitive diagnosis requires a multidisciplinary approach combining clinical, histopathological, immunohistochemistry, and radiological assessments. Histologically, DFSP is typified by a storiform proliferation of uniform spindle cells infiltrating the dermis and subcutis. Immunohistochemically, the tumor cells exhibit diffuse CD34 and vimentin positivity while lacking HMB45, desmin, smooth muscle actin (SMA), and S100.[1,15]

Once the diagnosis of DFSP has been histologically established, the next step is to evaluate the extent of local disease, exclude distant spread, and assess patient suitability for subsequent treatment. The priority of initial staging is determining involvement of deeper structures, such as the fascia or beyond, for which contrast-enhanced soft-tissue MRI remains the preferred modality.[11]

Currently, there is no universally accepted staging system for DFSP.

In 2020, a modified staging scheme was proposed in the United States, adapted from the 2015[16] guideline recommendations. This system stratifies DFSP into five stages: Stage I, comprising non-protuberant lesions such as atrophic, macular, or sclerotic plaques; Stage II, representing protuberant primary tumours and subdivided into Stage IIA, limited to superficial tissues without fascial invasion, and Stage IIB, in which the tumour infiltrates or extends beyond the underlying fascia; Stage III, defined by regional lymph node involvement; and Stage IV, reserved for cases with distant metastatic spread. This proposed classification was applied in the 2024 guideline as it corresponds logically with conventional TNM-style staging frameworks.[11] Nevertheless, it should be noted that external validation of this system has not yet been reported. According to the 2020 DFSP staging proposal, the tumour in our patient is best categorised as Stage IIB. This corresponds to a protuberant primary tumour with extension beneath and beyond the superficial fascia, evidenced by involvement of the anterior maxillary wall, with no clinical or pathological evidence of lymph node involvement or distant metastatic disease.

DFSP demonstrates marked local aggressiveness, and recurrence remains a significant concern depending on the therapeutic approach. Published recurrence rates vary considerably, ranging from 0% to 40%, although more contemporary series report lower figures. One cohort with a median follow-up of 59 months demonstrated recurrence-free survival rates of 86% at five years and 76% at ten years.[14] In another large study involving 200 DFSP cases and 34 patients with fibrosarcomatous transformation, none of the conventional DFSP patients developed distant metastases. In contrast, metastatic spread occurred in 23.6% of those with transformed DFSP, most frequently affecting the lungs, soft tissues, and skeletal system.[17] The fibrosarcomatous variant is the most aggressive variant, characterised histologically by high-grade transformation and clinically by significantly greater risks of recurrence and distant metastasis relative to classic DFSP.[12] This transformation is typically indistinguishable from conventional DFSP on clinical examination, making tissue diagnosis essential for detection. Recurrence risk in DFSP is influenced by several tumour-related characteristics, including histologic subtype, degree of cellularity, lesion size, anatomical location within the head and neck region, and an elevated mitotic index.[18]

Achieving adequate surgical margins on the face poses significant challenges due to aesthetic and functional constraints. Although wide local excision with margins 2-5 cm remains the mainstay of treatment, clear margins are often difficult to achieve in cosmetically sensitive areas. Reported recurrence rates are significantly higher when excision margins are less than 3 cm (approximately 46%) compared to 7% when margins of 3–5 cm are achieved.[13] Micrographically controlled surgery has been shown to provide superior margin control and lower recurrence rates.[11] The cure rate reported ranges from 0.6-6.6%.[13] When Mohs micrographic surgery is unavailable, the recommended treatment approach is wide local excision with comprehensive histopathological assessment of all surgical margins to ensure complete tumor clearance.[13] The choice of reconstructive technique is influenced by multiple factors, including the location and size of the tumor. Complex or extensive defects such as our patient require distant or free flaps to achieve both functional restoration and satisfactory cosmetic outcomes and function while minimizing morbidity.[19]

Interestingly, more than 90% of DFSP demonstrated a translocation t(17;22)(q22;q13), resulting in COL1A1-PDGFβ fusion gene. This leads to constitutive activation of the platelet-derived growth factor receptor beta (PDGFRβ) signaling pathway, promoting autocrine-driven tumor proliferation.[11] The detection of this fusion through fluorescence in situ hybridization (FISH) or RT-PCR is diagnostic and has guided the introduction of targeted therapy with tyrosine kinase inhibitors such as imatinib. It is particularly useful for unresectable, recurrent, or metastatic cases.[11] Radiotherapy may be considered when additional surgery is not feasible or in cases involving fibrosarcomatous transformation.[1] DFSP on the face and scalp, especially those greater than 5cm, typically recur within 3 years of surgery, while distant metastasis remains rare (<5%).[11] Although our patient remains disease-free at 18-month post-surgery, long-term clinical surveillance is required.

The clinical course in our patient mirrors previously reported cases, with a prolonged period of indolent plaque development before nodular progression. Similar delays in diagnosis of approximately 3–5 years have been documented in the literature, underscoring the deceptively benign appearance of early DFSP. Notably, involvement of the cheek with extension to the anterior maxillary wall remains uncommon, and few published cases describe the need for maxillary resection and microsurgical reconstruction, highlighting the complexity of management in this anatomical region.

CONCLUSION

This case highlights the importance of early recognition and prompt management of slow-growing facial masses. A timely management includes a crucial diagnostic biopsy may lead to an early yet less extensive surgery. This will greatly reduce the reconstructive complexity and thence minimize postoperative morbidity.

AUTHOR CONTRIBUTIONS' STATEMENT

IPLT was responsible for the study design, data collection, manuscript writing, AHMZ and KSC participated in data collection. IPLT, AHMZ, KSC, and MMT was involved in the discussion, manuscript editing, and language proofreading. All authors read and approved the final manuscript.

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