CASE REPORT

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Dermatofibrosarcoma Protuberans of Parotid Region: A Case Report

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive fibroblastic neoplasm that commonly occurs in the trunk and extremities but rarely in the head and neck region. We report an atypical presentation of DFSP with fibrosarcomatous transformation of the parotid region. A 34-year-old gentleman presented with a huge left parotid swelling that progressively increased in size over the past seven years. He claimed that a biopsy of the swelling was done five years prior in another hospital and he was told it was benign. He defaulted to follow-up as the lesion was painless initially but it started to be painful and bleed intermittently one month before the presentation. Total parotidectomy was done with the removal of adjacent enlarged lymph nodes but histopathological evaluation of the samples showed fibrosarcomatous DFSP with positive margins. Unfortunately, the patient defaulted to follow-up again post-op and presented again after one year with extensive systemic metastasis. The dilemmas in managing this case and suggestions for countermeasures together with references according to the relevant literature are discussed.

Keywords: *Dermatofibrosarcoma protuberans; fibrosarcomatous; head and neck; metastasis; parotid*

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is classified by the 2020 WHO Classification of Soft Tissue Tumours as an intermediate locally aggressive fibroblastic tumour (Sbaraglia *et al.*, 2021). DFSP is rare with an estimated incidence of 0.8 to 5 per million (Bogucki *et al.*, 2012). It represents <1% of all tumours and 7% of all soft tissue sarcoma. It occasionally occurs in the head and neck region (10% to 16%), and extremities (16% to 30%), with the most common sites at the trunk (42% to 72%) (Lemm *et al.*, 2009). DFSP is known to have a high rate of local recurrence post-excision and a low potential for metastasis (Huis In't Veld *et al.*, 2019). Histologically, it is represented by the proliferation of spindle cells with a storiform pattern and

positive staining for CD34. DFSP may present in about 85% to 90% of cases as a low-grade tumour, however, the remaining may progress into fibrosarcomatous transformation which is more aggressive and has a higher incidence of distant metastasis at 10% to 15% (Angouridakis *et al.*, 2011).

CASE REPORT

A 34-year-old gentleman with no known medical illness presented with a huge mass over the left parotid region. It initially started as a painless peanut-sized lesion and progressively increased in size over the past seven years. He claimed that a biopsy of the swelling was done five years prior in another hospital and he was told it was benign. He defaulted to follow-up as the lesion was painless initially but it started to be painful and bleed intermittently one month before the presentation.

Clinical examination revealed a huge mass over the left parotid area, which was firm, non-mobile, and non-tender, measuring 20 cm × 24 cm × 20 cm , and with an area of ulceroproliferative lesion at its center with a tendency to bleed when scratched. The skin appeared thin with features of telangiectasis (Fig. 1) and is fixed to the underlying mass. The lobule of the left pinna was attached and stretched by the mass. There was no trismus, and no facial nerve involvement was noted. Regional lymph nodes were not palpable. Other clinical examinations were normal.

Trucut biopsy showed a mesenchymal tumour with spindle cells, low-to-intermediate grade. Computed tomography (CT) showed a huge well-defined heterogenous enhancing mass at the left neck with areas of hypodensity likely representing necrotic components. There was poor demarcation with the parotid gland but a good plane was seen with the underlying muscles (Fig. 2).

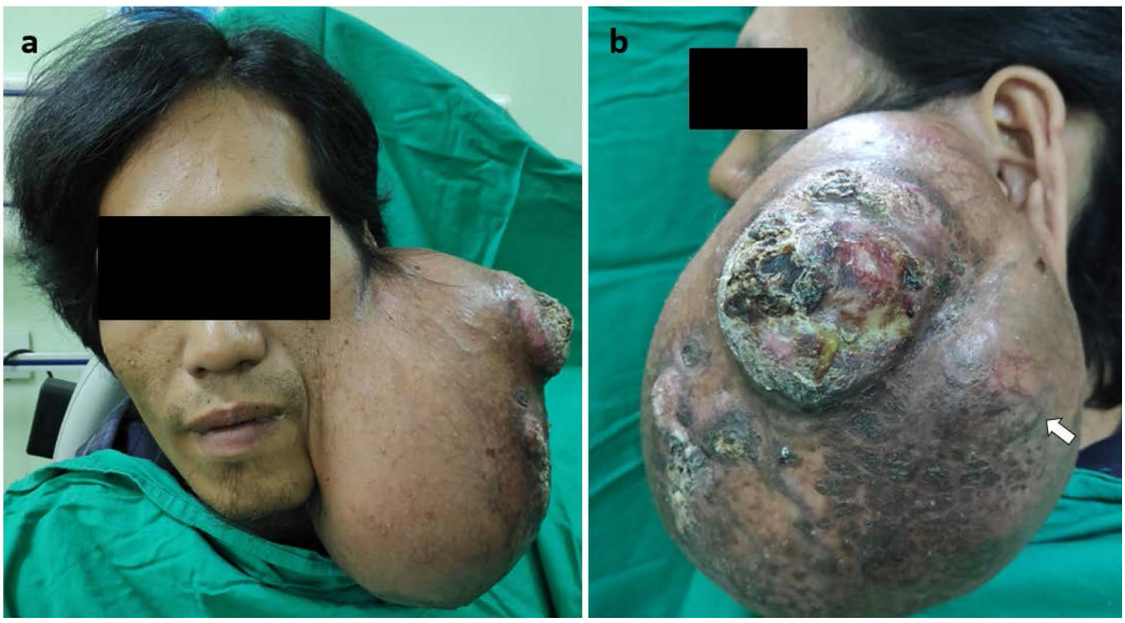

Fig. 1 Left parotid swelling measuring 20 cm × 24 cm × 20 cm in size: (A) with ulceroproliferative lesion at the centre; and (B) note the thinned skin with telangiectatic changes (arrow).

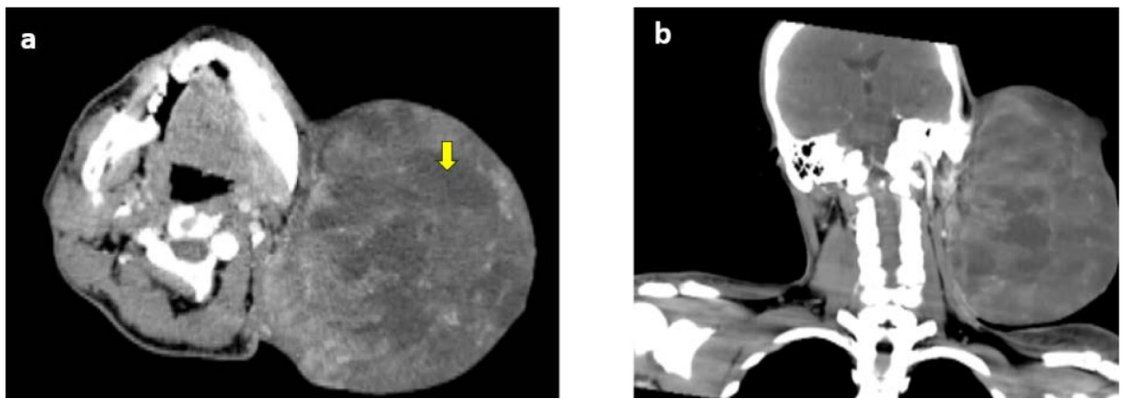

Fig. 2 Heterogenous enhancing mass measuring 15.8 cm × 13.3 cm × 18.8 cm in (A) axial and (B) coronal view of contrasted CT neck. Areas of hypodensity (arrow) likely represent necrotic components.

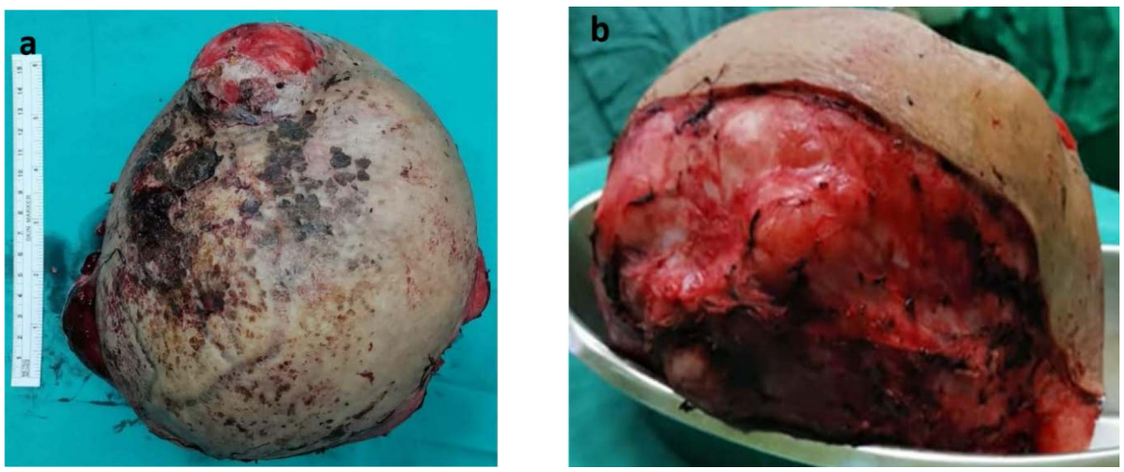

Fig. 3 (A) Tumour excised en bloc weighing 2.2 kg; (B) tumour easily separated from the underlying layer with a smooth whitish rubbery surface.

Left total parotidectomy was done and a pale rubbery mass measuring 18 cm × 20 cm, weighing 2.2 kg was excised en bloc along with the overlying skin (Fig. 3).

It was superficial to the facial nerve and all branches of the nerve were identified and preserved. Multiple subcentimetre lymph nodes were noted at level V and were excised. The wound was closed with primary closure and a drain was inserted. Postoperatively, the patient recovered well with grade 2 left marginal mandibular branch facial nerve palsy that gradually improved.

Histopathological evaluation of intraoperative samples showed spindle-shaped cells arranged in compact fascicles and storiform patterns which were rather uniform (Fig. 4A), having hyperchromatic nuclei and scanty cytoplasm with ill-defined cell borders (Fig. 4B). Mitotic count is 10-20/10 HPF. Immunohistochemical markers were positive for CD34 (Fig. 4C) and negative for S100 and EMA in favour of fibrosarcomatous DFSP. The level V lymph nodes were negative for malignancy. Less than 1 mm resection margins were noted and the patient was planned for urgent CT staging and adjuvant radiotherapy.

Unfortunately, the patient defaulted to follow-up post-op and presented again after one year with a new bleeding lesion at the right inguinal region and metastases to the lung, bone, omentum, and intra-abdominal

lymph nodes (Figs. 5A and 5B). Biopsy from the right inguinal lesion confirmed the diagnosis of metastatic fibromatous DFSP and the patient was started on hemostatic radiotherapy 30Gy/6 fraction and planned for a trial of imatinib by the oncology team with palliative intent. Unfortunately, the patient passed away before the treatment was initiated.

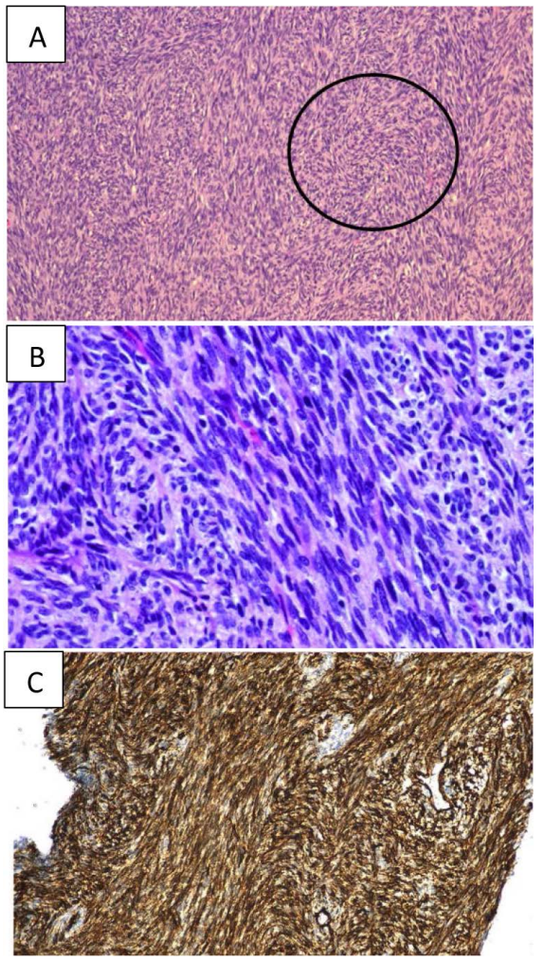

Fig. 4 Microscopic view of the specimen. (A) 10× magnification: Spindle-shaped cells arranged in compact fascicles and storiform pattern (circle); (B) 40× magnification: Lesional cells having hyperchromatic nuclei and scanty cytoplasm with ill-defined cell borders; (C) 10× magnification: Spindle-shaped cells stained positive for CD34.

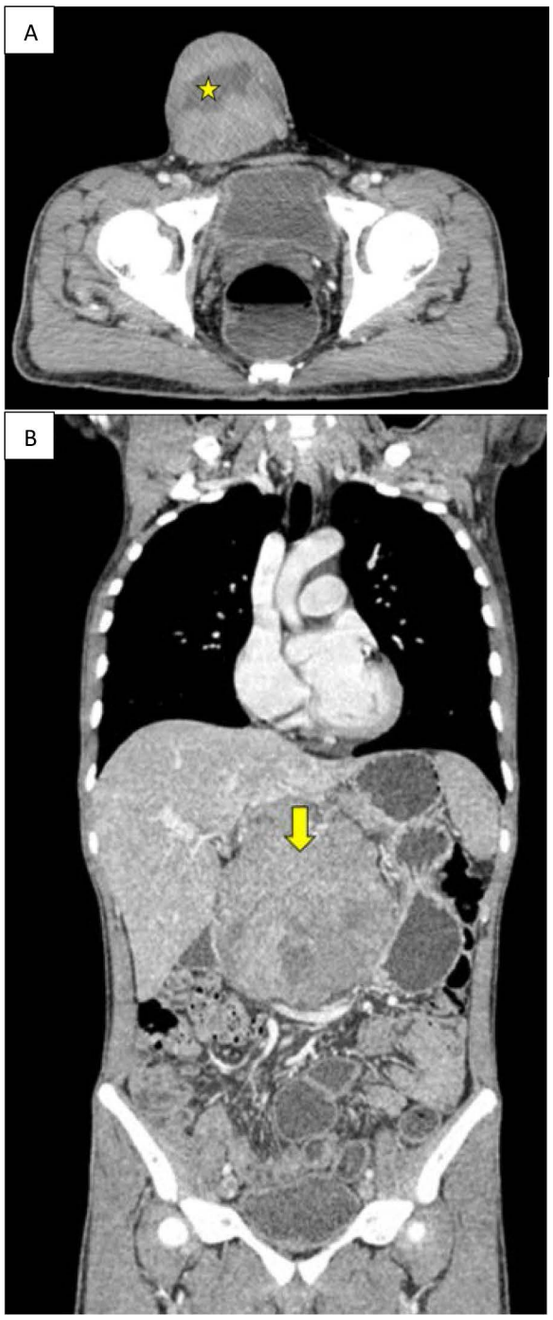

Fig. 5 Extensive metastases with new heterogenous mass at (A) right inguinal measuring 7.8 cm × 7.7 cm × 7.6 cm (star); and (B) abdomen likely originating from omentum measuring 17.7 cm × 16.3 cm × 9.8 cm (arrow).

DISCUSSION

DFSP especially the fibrosarcomatous variant is a locally aggressive disease and should not be underestimated. In our case, the patient ignored the lesion after it was misdiagnosed as benign, which allowed the tumour to grow into huge dimensions, followed by failure to receive optimal treatment and follow-up post-surgery which resulted in extensive metastasis and systemic involvement of the disease.

DFSP is rare and presentation at the infra-auricular region as seen in our patient is atypical. One may come to a clinical misdiagnosis of salivary gland tumours such as pleomorphic adenoma or mucoepidermoid carcinoma which are more commonly seen in that region. However, a good understanding of the clinical progression of DFSP may guide us to a more accurate diagnosis. DFSP is a mesenchymal neoplasm that originates from the skin's dermal or subdermal tissue and progresses from superficial to deep (Alnemare *et al.*, 2018). It is rarely the case for salivary gland tumours where the skin is involved only in the late stages of the disease. Early presentation of DFSP may appear as a slow-growing plaque-like mass or nodule on the skin which may be pink or reddish blue and blanches upon pressure. It is mobile upon palpation as it fixes to the skin but not underlying tissue, except in lesions involving the periosteal layer where scalp fixation may occur in earlier stages (Lemm *et al.*, 2009). As the disease progresses, the tumour infiltrates deeper and becomes fixed, developing into a protuberant growth that may become ulcerative and bleed as the stretched skin becomes thin (Bogucki *et al.*, 2012). Nevertheless, due to its slow growth and inert nature, this progression is hard to observe and patients often present late.

The second dilemma would be to obtain a representative sample for a definitive histopathological diagnosis. DFSP is characterised by uniform monomorphous proliferation of spindle cells arranged in a storiform, cartwheel-like pattern, with immunohistochemical staining typically demonstrating strong positivity for CD34 (Bogucki *et al.*, 2012). In regard to the mode of biopsy, open biopsy or trucut biopsy would be superior when compared to fine needle aspiration (FNAC). The former would provide information on the architecture and pattern of invasion of the tumour as well as more samples for immunohistological and other ancillary studies for a more accurate differential. Another concern especially for large tumours is the heterogenicity of the tumour. Necrotic and cystic areas may result in poor yield which may explain the discrepancy in pre and postoperative histopathology results of the patient. This can be remedied by ultrasound-guided targeted biopsy which provides the advantage of avoiding vital neurovascular structures as well (Haldar *et al.*, 2016).

Management of DFSP can be very challenging as it could be mimicking features of a benign tumour while being locally aggressive with a distant metastasis rate of 0.5% to 5% (Bogucki *et al.*, 2012) if left unchecked. Surgeons need to have a high clinical suspicion of a more aggressive fibrosarcomatous variant of DFSP which shows atypical spindle cells with higher mitotic activity (20 vs 2 mitosis/10 high power field), lower CD34 expression, and higher p53 expression (Abbott *et al.*, 2006). There are also reports that radiological findings of intramural hypodensities in larger tumours may represent necrotic degeneration which should raise the concern of fibrosarcomatous transformation (Mentzel *et al.*, 1998) and may warrant further imaging to evaluate for distant metastasis (Bowne *et al.*, 2000). Metastasis most commonly occurs via the hematogenous route to lungs and bones but other sites such as lymph nodes, brain, visceral organs, and soft tissue have also been noted (Mahajan *et al.*, 2015; Hayakawa *et al.*, 2016). Therefore, a thorough systemic examination should be exercised with caution before subjecting the patient to surgery. If fibrosarcomatous DFSP is suspected, it should be treated as an advanced disease, and consultation with a

multidisciplinary tumour board is needed to plan for more aggressive treatment (Bowne *et al*., 2000).

One of the most common pitfalls in the treatment of DFSP is preoperative planning and surgery. Radiological imaging may be a useful tool, especially in larger tumours to evaluate the extension and relation to nearby vital structures. CT often reveals a solitary, well-defined cutaneous or subcutaneous mass with homogenous to heterogenous soft tissue attenuation and enhancement post-contrast, while magnetic resonance imaging (MRI) would show isointense compared to muscle on T1-weighted image and hyperintense on T2 image with post-contrast intermediate to marked enhancement (Zhang *et al*., 2015).

A simple excision, however, would result in a high recurrence rate of 10% to 60% which is seen in earlier reports in the 1960s to 1980s (Bogucki *et al*., 2012). This can be attributed to the deceptively well-circumscribed margins, outlined by a false capsule because of compression to the surrounding tissue (Garça *et al*., 2013) which was also seen intraoperatively in the case discussed. The tumour was easily separated from the underlying tissue with a smooth rubbery white surface and despite what appears to be a complete excision of the tumour post-total parotidectomy, the margins post-op is still positive.

Various studies have shown that DFSP has microscopic irregular tentacle-like projections that infiltrate into deeper structures and failure of a complete clearance would increase the risk of recurrence (Hao *et al*., 2020). The ideal treatment for resectable tumours is wide local excision (WLE) with a 2 cm to 4 cm margin (Bogucki *et al*., 2012; Hao *et al*., 2020) or by Mohs micrographic surgery (MMS) where margins post-tumour removal is examined with frozen section intraoperatively and deeper or wider resection is done until clear margins are achieved (Chang *et al*., 2004). Multiple studies comparing the outcome for the recurrence rate of both techniques favour MMS (0% to 6.6%) instead of WLE (1.7% to 30.8%) (National Comprehensive Cancer Network, 2022).

However, total resection of the tumour is not always feasible especially when critical areas are involved in the excision margin for example in the head and neck region where cosmetic and functional considerations come into play (Angouridakis *et al*., 2011). Such cases, along with unresectable or metastatic tumours can be offered radiotherapy or targeted therapy such as imatinib mesylate (IM). Studies have shown DFSP to be radiosensitive, and in a retrospective study of 38 patients with DFSP, the local control after surgery alone is 67% compared to 82% where adjuvant radiotherapy is given postoperatively (Haas *et al*., 1997). Imatinib inhibits the tyrosine kinase of PDGF-R, reducing the proliferation of DFSP neoplasm and inducing apoptosis. A recent systemic review of 152 patients of metastatic or locally advanced DFSP receiving imatinib showed complete response in 5.2%, partial response in 5.2%, stable disease in 27.6%, and progression in 9.2% of patients (Navarrete-Dechent *et al*., 2019).

The patient should be well informed and involved in each step of management, understanding the risk and implications of the choices made. There is always a possibility of false negative histopathological results in the early stages of evaluation and the guard should not be let down until the final definitive diagnosis. The patients must adhere to the follow-up and treatment with vigilance and self-awareness for any new symptoms of recurrence. The median time for recurrence is 32 to 38 months and several studies recommend long-term surveillance every 6 to 12 months (Hao *et al*., 2020). Patients with poor socioeconomic status or from rural areas inaccessible to healthcare services should be offered as much help as possible from social welfare services to avoid cases of defaulted follow-up.

CONCLUSION

DFSP is rarely seen in the head and neck region and is difficult to manage. However, awareness and a good understanding of the clinical and pathological features, together with careful selection of the appropriate mode of biopsy to obtain the best representative sample for histopathological evaluation is crucial to come to a correct diagnosis. Prompt recognition of the signs of fibrosarcomatous transformation and the deceptive tumour margins due to microscopic infiltration of this disease is also important to plan for optimal treatment for the patient. The patient should be well informed on the management and every effort should be made to ensure patient compliance with the treatment and monitoring for recurrence.

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