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Orbitofacial dermatofibrosarcoma protuberans with intranasal extension

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ABSTRACT
A 25-year-old Chinese woman presented with recurrent painless swelling over the left medial canthus region for 3 months and intranasal mass for an indeterminate duration. Initial incision biopsy of the mass was reported as nodular fasciitis but the lesion recurred 3 weeks later. Intraoperative findings during repeat biopsy showed a mass extending from the deep dermal tissue into the anterior orbit and polyp-like nasal mass. Histopathology findings were that of dermatofibrosarcoma protuberans (DFSP). The mass recurred 4 months later without orbital or intranasal recurrence. Wide excision biopsy under frozen section guidance was attempted however; clear surgical margins could not be achieved despite extensive resection. She was subsequently referred for adjuvant radiotherapy. We report an exceptionally rare case of local recurrence of DFSP in an unusual anatomic location. This case was surgically challenging in achieving negative margins, and thus neoadjuvant therapy may improve overall outcome to prevent local relapse.

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Dermatofibrosarcoma protuberans; intradermal tumor; orbital extension; wide local excision

Introduction
Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing mesenchymal tumor of the dermis accounting for approximately 0.1% of all malignancies and 1.8% of soft tissue sarcoma.1 This locally aggressive tumor involving the periorbital area has been infrequently reported.2 The rarity of the disease lessens their clinical awareness which offers a diagnostic and therapeutic challenge. Here, we report a rare case of a local recurrence of DFSP following standard surgical excision in an unusual anatomic location.

Case report
A 25-year-old Chinese woman presented with painless swelling over the left medial canthus region for 3 months. There was no history of trauma and she had no previous significant medical illness. Initial incision biopsy done in a private center was reported as nodular fasciitis. However, the lesion recurred 3 weeks later associated with left intranasal mass.

Upon presentation, her visual acuity was 6/6 in both eyes. Examination revealed a palpable, non-tender, indurated subcutaneous mass measuring about 3 cm × 2 cm over the left medial canthus. There was also a polyp-like mass in the left-nasal cavity with an indurated skin lateral to the nose (Figure 1). The remainder of the ophthalmic examination was normal. Systemic evaluation including cervical lymph nodes examination and chest radiograph was unremarkable.

A repeat incisional biopsy of the left recurrent medial canthus mass revealed a homogenous, greyish, deep friable mass extending into the anterior orbit without distinct margin. The mass measured about 9 mm x 1 mm x 1 mm. Histopathological analysis demonstrated a poorly demarcated intradermal lesion composed of spindle cells proliferation in diffuse sheets. There were areas of storiform growth pattern involving all resected margins and subcutaneous tissue extending up to papillary dermis. The nuclei were elongated, spindled with inconspicuous nucleoli and ill-defined cytoplasm. Mitotic activities were present up to 5/10 per high-powered field (hpf). At the periphery of the tumour, focal loose area with myxoid background was detected. There was no necrosis or cytologic atypia noted (Figure 2A and 2B). Immunohistochemistry was
strongly positive for CD34 and vimentin, and negative for smooth muscle actin (SMA) and S100 (Figure 2C and 2D). These findings were consistent with DFSP.

Excision biopsy of the left-nasal mass and nasolabial tissue was performed by the otorhinolaryngology team. The specimen obtained was a grey whitish tissue measuring 45 mm x 25 mm x 10 mm and an exophytic tumor mass measuring 40 mm x 17 mm x 6 mm. Microscopically, the tumor cells were spindled with no definite pattern of arrangement embedded within slightly fibromyxoid stroma. The cells were mildly pleomorphic with ill-defined cytoplasmic borders and inconspicuous to small nucleoli. Mitosis is occasionally seen (2/10 hpf). There was no necrosis or carcinomatous differentiation seen. The tumor cells were positive for CD34 and vimentin stains, and negative for desmin and S100. These findings were consistent with low-grade fibromyxoid sarcoma.

The patient was strongly urged to undergo wide excision biopsy for proper tumour clearance, however, she refused. Post-excision magnetic resonance imaging (MRI) 3 months later showed a recurrence of subcutaneous lesion over the left medial canthus extending down to the left nasolabial fold. It measured about 1.3 cm x 1.3 cm x 4.5 cm. However, there was no orbital recurrence noted (Figure 3A and 3B). Six months later, the growth recurred as an indurated mass over the previous incision site which gradually increased in size involving the lateral side of the nose and maxilla (Figure 4A).

The patient was finally agreed for surgical with the aim of tumor clearance. She underwent wide excision biopsy under frozen section guidance by plastic surgery team. However, clear surgical margins could not be achieved despite extensive resection. She was subsequently advised for intensity-modulated radiation therapy (IMRT) and planned for reconstruction later. However, she declined any intervention including serial imaging and had been lost to follow up in view of unacceptable cosmetic outcomes post-extensive surgical resection. Patient presented to our clinic 12 months later with extensive indurated mass over the left peri-orbital region and yet still not keen for any intervention.

Figure 1. Indurated subcutaneous mass over the left medial canthus with polyp-like mass in the left-nasal cavity (arrow).

Figure 2. (A) The presence of intradermal spindle cell proliferation arranged in a storiform pattern (hematoxylin–eosin (H&E), magnification x 4). (B) (H&E), magnification x 40. (C) Immunohistochemistry which was strongly positive for CD34 (magnification x 40). (D) Positive immunostaining for vimentin (magnification x 40).
Repeated MRI of the orbit revealed a subcutaneous mass at the anterolateral aspect of the left eye without intraorbital or intracranial extension (Figure 3C).

**Discussion**

This case demonstrates the locally aggressive behavior with high risk of local recurrence of DFSP involving the
periorcular region. The clinical presentation of DFSP can vary widely and often masquerades as a benign, indolent tumor leading to a delay and misdiagnosis of the disease.3

Our patient presented with recurrent mass over the previous incision site with negative initial biopsy. The lesion recurred and expanded rapidly invading the orbit and the nasal cavity within the following 3 weeks. Therefore, a high index of suspicion of the disease should be considered for a recurrent tumor and a repeated biopsy is crucial. A definitive diagnosis can be made based on the classical histopathologic findings characterized by a storiform pattern of spindle cells and CD34 positivity on immunohistochemical studies.4

DFSP involves most frequently the trunk accounting for 42–72% and the least common in head and neck which accounts for 10–15%.5 In patients with head and neck disease site, only 3.5% involve the periorcular region.2 The unusual location of the tumor in our patient involved the medial canthal region extending into the orbit and the nasal cavity making her a surgically challenging candidate. The higher recurrence rate in this patient was due to the difficulty in obtaining wide margins following a critical anatomic site as well as functional and cosmetic concerns.

The literature search showed seven prior case reports of DSFP involving the orbit.2,6–11 Of the cases mentioned, three cases presented with medial canthal mass which was similar with our patient’s initial presentation. The age of the patients involved was reported to be range between 38 and 71 years old and our patient was reported to be the youngest patient of DSFP with orbital extension. Being extremely rare in the orbit, ours is the eighth reported case.

The first case was reported in 2004 of a 61-year-old man with metastasis to the orbit from the primary DSFP of the lumbar region.7 Due to the advanced stage of the disease and multiple concurrent systemic metastases, he was treated palliatively with orbital radiation, chemotherapy, and orbital exenteration. Goshe et al. reported a case of a 38-year-old man with primary DFSP of the glabella and medial canthus invading the anterior orbit in 2012.6 The patient was treated with exenteration and dacryocystectomy after failed Mohs surgery. The third case is that of a primary DFSP of the orbit in a 70-year-old woman who presented with left eyelid swelling with superolateral orbital mass.8 She underwent orbital exenteration due to extensive involvement of the orbit.

Schittkowski et al. reported a case of a 45-year-old woman who presented with primary DFSP of a palpable medial canthal mass invading the orbit.9 She remained recurrence free for 24 months after a successful tumor excision via transcutaneous orbitotomy. Erickson et al. reported a case of a 71-year-old man who presented with left medial canthal mass extending into the medial orbit and the proximal part of the nasolacrimal duct.2 This was the first case of a recurrent DFSP invading the orbit managed with conservative resection and adjuvant therapy. There was no evidence of recurrence at 11 months of follow-up. Most recently, Bashir et al. reported a case of orbital DFSP with intracranial extension which was treated with imatinib mesylate and radiotherapy.10

The standard of therapy is complete surgical excision with appropriate reconstruction.6 Mohs micrographic surgery (MMS) has been found to be the more superior technique with markedly lower recurrence rate especially in head and neck lesions.12 The lesions in our patient continue to exhibit positive margins after multiple resections which MMS may not be feasible due to the extension of the lesion into the deeper structures. Therefore, she was advised for adjuvant therapy since the complete surgical resection was unachievable. Radiotherapy is one of the treatment options in patients with locally advanced tumor and recurrent lesions.13 Moreover, chemotherapy and more recently imatinib mesylate have been reported to be effective.14

In view of unacceptable cosmetic outcomes post-extensive surgical resection, our patient declined any further intervention including serial imaging and had been lost to follow up. She may carry the risk of distant metastases even though it has been reported to be as low as 4% of the cases occurring mainly in lung and lymph nodes.14 For this reason, all DFSP patients should be followed up for life for evidence of metastatic disease when feasible.

### Conclusion

We report an exceptionally rare case of local recurrence of DFSP in an unusual anatomic location. We emphasize the value of a highly aggressive local resection in the primary treatment of the disease to minimize local recurrence and potential malignant transformation. This case is surgically challenging in achieving negative margins, and thus neoadjuvant therapy may improve overall outcome to prevent local relapse. Better awareness of this entity can lead to a prompt diagnosis and proper management of the disease.

### Patient consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the editor of this journal.
Disclosure statement

No potential conflict of interest was reported by the authors.

References