A large primary dermatofibrosarcoma protuberans of the right upper eyelid: A case report and literature review

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SUMMARY

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing but locally aggressive dermal sarcoma. A 50-year-old woman is reported with a seven-year history of progressive right upper eyelid swelling, who had a prior benign mass removed from the same site. Imaging showed a lobulated right periorbital mass, and surgical excision identified a 4.4 × 4.2 × 3.1 cm tumour confirmed as DFSP with positive margins. Although she declined radiotherapy, there was no recurrence at 12 months. DFSP should be considered in the differential diagnosis of periorbital masses, as it can mimic benign lesions. Given its high risk of recurrence and aggressive local invasion, achieving complete surgical excision remains a challenge.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is an uncommon dermal neoplasm with gradual enlargement, local aggressiveness, and frequent relapse after treatment.[1-2] DFSP typically appears as a purple or pink plaque that most commonly arises on the trunk in 50-60% of cases, on the extremities in 20-30% of cases, and with the head and neck region involved in approximately 10-15% of cases.[2-4] Among head and neck cases, periocular or ocular involvement is very rare and occurs in approximately 3.5% of cases.[2-3] The medial canthus is the most frequent site for reported periorbital DFSP cases.[3] Orbital involvement typically occurs due to local invasion from adnexal lesions or, very rarely, from distant metastases.[3]

Based on PubMed search of DFSP cases affecting the eyelid region, only three instances of DFSP originating from a previous surgical scar were identified, including the present case.[5] This case of primary DFSP originating in the right upper eyelid is presented to emphasise the need for a high index of suspicion, as DFSP can be diagnostically challenging due to its resemblance to benign lesions and its occurrence at uncommon sites.

CASE PRESENTATION

A 50-year-old woman with a background of uterine fibroids presented initially with a five-year history of progressive, painless swelling and drooping of the right upper eyelid. The lesion initially appeared as a small papule at the medial aspect of the right upper eyelid and gradually increased in size. Magnetic resonance imaging (MRI) of the orbit demonstrated a superomedial eyelid mass measuring approximately 1.7 × 2.2 × 2.2 cm. She had undergone excision of a right upper eyelid mass, and the histopathological examination was reported as a benign fibrohistiocytic lesion. She remained well postoperatively for one year, but then she subsequently defaulted on follow-up.

She now presented with a gradually enlarged eyelid mass with an estimated duration of seven years at the past surgical site. On presentation, her best-corrected vision (BCVA) was 6/9 (20/30) in the right eye and 6/6 (20/20) in the left eye, with no relative afferent pupillary defect in either eye. A firm, non-tender, immobile mass was palpated over her right upper eyelid region, which caused complete mechanical ptosis (Figure 1A). The extraocular movement was normal except for elevation in the right eye. Ocular examination was normal in both eyes.

MRI of the orbit revealed a well-defined, lobulated, enhancing soft tissue mass located anteromedially in the right periorbital region. The mass had an iso-intense signal to grey matter on T1-weighted images and a heterogeneous intermediate signal on T2-weighted images (Figs. 2A and 2B). It was located near the right eye globe, resulting in a slight inferior and posterolateral displacement (Figs. 2C and 2D). Additionally, the mass was in proximity to the frontal sinus at the front.

The patient underwent an excisional biopsy through a transcutaneous anterior orbitotomy and upper eyelid reconstruction surgery, which included wedge resection, levator advancement, and upper blepharoplasty under general anaesthesia. A large multilobulated encapsulated mass with overlying dilated vessels was carefully removed intraoperatively. The mass, measuring 4.4 x 4.2 x 3.1 cm, was rounded with a smooth outer surface and an area of irregular, papillary-like tan-whitish tissue measuring 4.0 x 2.3 cm (Figs. 3A and 3B). The cut section showed a soft tan-pink mass with an irregular border.

Microscopic examination of the specimen revealed a partially circumscribed tumour composed of spindle cells

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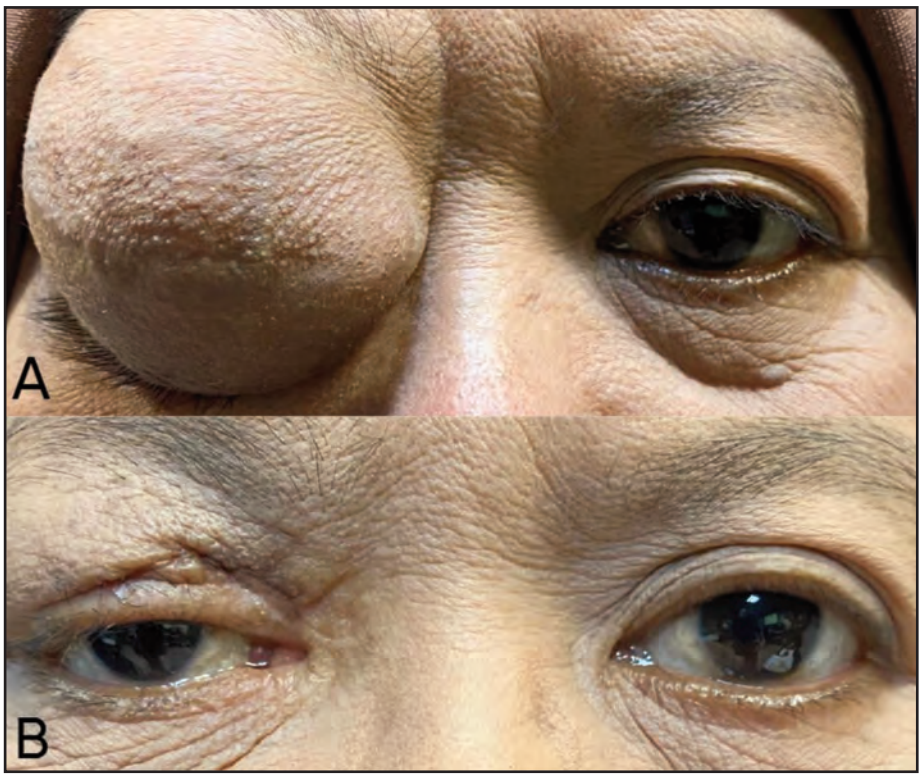

Fig. 1: A) Preoperatively, swelling due to a mass completely under the skin at the right upper eyelid, causing complete mechanical ptosis. B) Photo at 12 months post-operation showing right residual ptosis with minimal keloid formation at the previous surgical wound site.

arranged in various patterns, including diffuse sheets, cartwheels, storiform, and fascicles, with occasional mitotic figures (Figure 3C). The tumour was infiltrative and present at the irregular resected margins. Immunohistochemistry showed strong and diffuse positivity for CD34 (Figure 3D), but negativity for S100, desmin, and cytokeratin (CK AE1/AE3). Smooth muscle actin (SMA) highlighted scattered small vessels within the tumour, and the Ki-67 stain indicated a proliferative index of approximately 15%. These findings were consistent with a diagnosis of DFSP.

The patient was advised to undergo radiotherapy because of the positive surgical margins. However, she declined the recommended treatment. The ocular examination revealed residual ptosis of the right upper eyelid three months post-surgery. Hence, right upper blepharoplasty and frontalis flap were performed three months later. Currently, 12 months post-tumour excision, she exhibits mild ptosis of the right upper eyelid but, fortunately, shows no clinical signs of tumour recurrence (Fig. 1B).

Her BCVA was 6/9 (20/30) in the right eye and 6/6 (20/20) in the left. Refraction revealed marked corneal astigmatism of –3.75 dioptre in the right eye, likely secondary to chronic pressure exerted by the mass, in contrast to mild astigmatism of –0.75 dioptre in the left eye. She was counselled regarding another lid reconstruction surgery but expressed a lack of interest at this time, as she is content with her current ocular condition.

DISCUSSION

DFSP is a rare cutaneous malignant neoplasm, occurring at a rate of 0.8 to five cases per one million people annually.[1,4] It represents less than 0.1% of all malignancies and 1-6% of all soft tissue sarcomas.[1] DFSP grows slowly but is locally aggressive, as it can infiltrate subcutaneous tissues, including muscles and bones. It has a high recurrence rate with a low risk of distant metastasis in 1-4%.[2-4] Although DFSP is commonly diagnosed between the ages of 20 and 50, it can be manifested across all age groups, affecting both sexes equally.[1-2] Table I summarises the published reports of cases with DFSP of the eyelid in relation to a previous history of trauma or multiple surgical procedures, including the patient.[4,6-8] The age at presentation is reported to be in the range of 35 to 62 years of age, with both sexes equally affected.

DFSP may develop within scars from prior trauma, surgery, or vaccination, in areas exposed to ionising radiation, or may arise spontaneously.[3-4] Patel et al. published a retrospective study of 364 DFSP patients and identified that 31 patients (8.5%) acquired DFSP at the location of a prior skin biopsy or small excision, whereas six patients (1.7%) developed DFSP at the site of a previous major surgical scar.[9] Persistent inflammation was postulated to play a role in DFSP development by driving the malignant transformation of scars.[9] The present case involved primary DFSP of the right upper eyelid, which arose from a scar at the site of a previous mass excision.

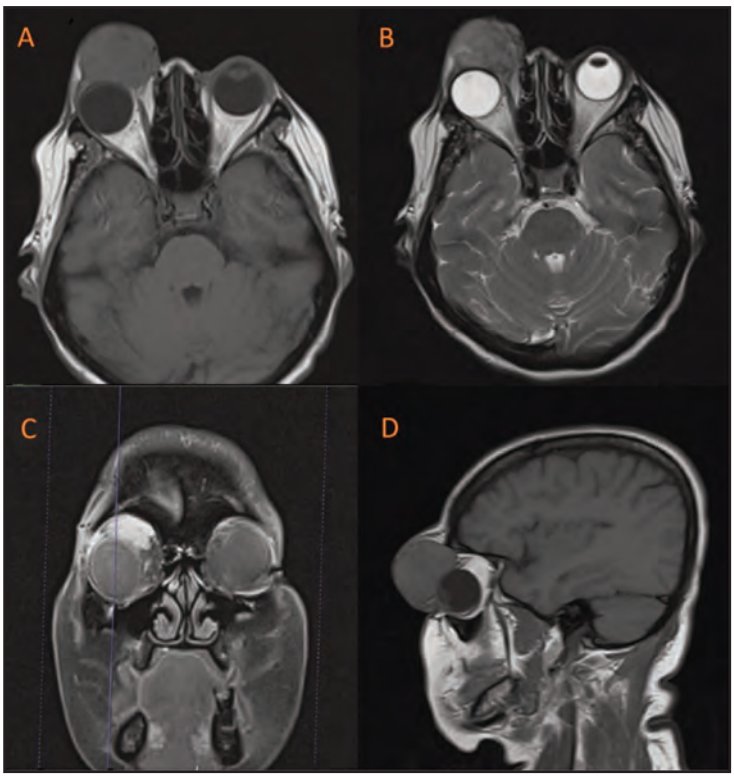

Fig. 2: The axial section of the orbital MRI scan shows a focal, well-defined soft tissue mass at the anteromedial aspect of the right periorbital region, demonstrating isointense signal on T1-weighted imaging (A) and heterogeneous intermediate signal intensity relative to grey matter on T2-weighted imaging (B). The coronal section of the MRI shows part of the mass causing displacement of the right eye globe inferolaterally (C). The sagittal section of the MRI shows the mass causing displacement of the right eye globe posteriorly (D)

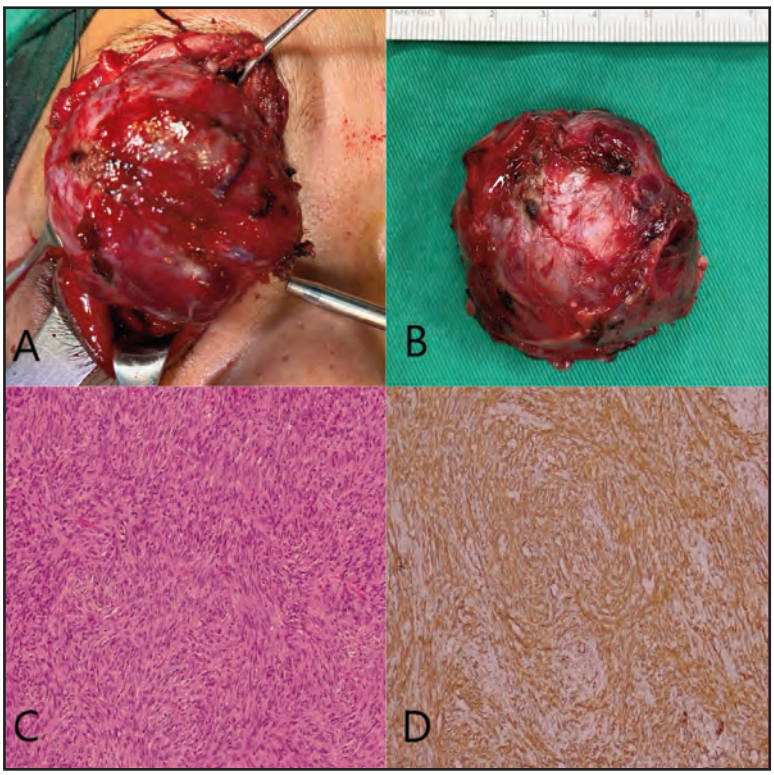

Fig. 3: Intraoperative image showing the location of the mass (A) and a round encapsulated mass with smooth outer surface post-surgical excision (B). Photomicrograph showing spindle cells arranged predominantly in storiform pattern (hematoxylin and eosin stain, magnification ×100) (C). Immunohistochemistry demonstrating diffuse CD34 positivity (magnification x100) (D)

Table I: Summary of published case reports of patients with DFSP of the eyelid in relation to trauma/scar.

Age/Gender/ Country	Initial tumor location	Site of extension	Risk Factor	Histopathology	Duration of illness	Treatment	Outcome/ Disease duration (month)	Author/ Year
62/M/China	Right lower eyelid	None	Local trauma (cat scratch)	Lace-like pattern. Atrophic variant	6 months	Resection of mass with wide margins and reshaped the lower eyelid	6 months	Li et al. 2011.[7]
38/M/USA	Left eyebrow	Left medial canthal, glabella and nasal	Struck by a wooden board	Spindle cell in storiform pattern	6 months	1) Mohs micrographic surgery 2) Exenteration and dacryocystectomy	6 months	Goshe et al. 2012.[6]
44/F/Turkey	Right medial canthus	None	Post mass excision	N/A	15 years	1) En-bloc resection 2) Intensity-modulated radiation therapy with 5400 Gray (180 Gray/fraction/day)	15 years	Kaya et al. 2022.[4]
35/F/Nigeria	Right side of the head	Right Upper eyelid	Post mass excision twice	Storiform pattern	18 years	1) Local resection of the tumor involving 3 cm of normal margin and split thickness skin grafting from the right thigh 2) Right cutler beard procedure and excisional biopsy with primary wound closure and graft of the scalp wound	18 years	Monsudi et al. 2023.[8]
50/F/ Malaysia	Right upper eyelid	None	Post mass excision	Diffuse sheets, cartwheel, storiform, and fascicles patterns	7 years	1) Excisional biopsy via transcutaneous anterior orbitotomy and upper eyelid reconstruction surgery 2) Right upper blepharoplasty and frontalis flap	7 years	Our case

F: female; M: male; N/A: not available

Goshe et al. reported a case of DFSP in a 38-year-old man who developed a tumour in a scar on his left eyebrow, two years after being hit by a wooden board.[6] Li et al. reported a case of primary DFSP of the right lower eyelid region with a history of trauma by a cat scratch at the tumour lesion, which may speed up its progression.[7] Kaya et al. reported a case of a 44-year-old woman who presented with primary DFSP in the right medial canthus.[4] She had a history of a previously excised mass at the same location 20 years earlier, though no pathology report was available.[4] Similarly, Monsudi et al. reported a recurrent case of DFSP on the right side of the head that extended to the right upper eyelid region with a previous history of excisional surgeries twice, but no histopathological examination of the sample was performed.[8]

The histopathological appearance of DFSP is characterised by uniform spindle neoplastic cells arranged predominantly in a storiform pattern parallel to the epidermal surface with minimal pleomorphism and scant cytoplasm.[1] It is surrounded by collagenous stroma and often displays a characteristic honeycomb appearance due to irregular projections infiltrating the subcutaneous tissue and fat.[2,4] In cases of periocular and ocular adnexa involvement, the histopathology of DFSP has been classified into storiform, atrophic, pigmented, diffuse sheets, and cartwheel patterns.[3,10] Immunohistochemistry typically demonstrates strong positivity for CD34 and vimentin,[1,3] with some tumours occasionally demonstrating focal positivity for SMA but not S100.[4] Satellite appearance around the lesion can occur due to intradermal infiltration of malignant cells, potentially extending to muscle and bone.[3] In the present case, the tumour was positive for CD34 and SMA but negative for S100, desmin, and CK AE1/AE3.

The presence of spindle cells arranged in a storiform pattern, combined with histopathological features consistent with DFSP and positive immunohistochemical staining for CD34 and vimentin, constitutes the key diagnostic criteria for DFSP. However, it is important to note that CD34 positivity can also be observed in other spindle cell tumours, such as solitary fibrous tumour, sclerotic fibroma, cellular digital fibroma, cutaneous neurofibroma, dermatofibroma, intradermal spindle cell lipoma, desmoplastic melanoma, and nuchal-type fibroma.[1] Sharudin et al. presented a case of recurrent painless swelling over the left medial canthus region that was previously reported as nodular fasciitis before developing DFSP.[10] Another published report also described DFSP developing after recurrent leiomyomas.[3] The preliminary histopathological interpretation in the present case resembled a benign fibrohistiocytic lesion, illustrating how DFSP may closely mimic benign entities. This highlights the diagnostic challenge and the importance of considering DFSP in recurrent or atypical eyelid lesions. False-negative pathological diagnoses delay treatment, allowing the tumour to extend, recur, and cause significant local damage. Accurate pathological diagnosis is therefore crucial for successful DFSP management.[1]

The principal treatment for DFSP is surgical excision, employing two primary techniques: wide local excision (WLE) and Mohs micrographic surgery (MMS). WLE involves resection of the tumour with a margin of adjacent healthy tissue of 3 to 5 cm, which is important for reducing recurrence rates.[4] The dermal defect resulting from WLE should be reconstructed using flaps or grafts.[3] However, WLE may not be feasible for head and neck tumours. MMS allows for immediate evaluation of margins, making it advantageous in cosmetically sensitive areas. It is associated with lower recurrence rates compared with WLE.[5] In cases where complete excision is not feasible or recurrence or metastasis is present, radiotherapy and chemotherapy are considered alternative treatments.[3] Adjuvant radiotherapy, such as brachytherapy or external beam radiotherapy, can enhance local control and reduce recurrence.[5] Jamor et al. reported a case of DFSP in the right upper eyelid, treating it with tumour excision and surface mould high-dose-rate brachytherapy.[5] They suggested this is a viable option when radical surgery is not possible and cost-effective in low-resource environments.[5]

The National Comprehensive Cancer Network Guidelines for soft tissue sarcoma recommend adjuvant radiotherapy in the setting of positive surgical margins.[2] In the patient, the combination of positive margins and refusal of radiotherapy increases the risk of local recurrence, emphasising the need for close and long-term follow-up. Molecularly targeted therapy, such as Imatinib, inhibits the platelet-derived growth factor receptor and other tyrosine kinase receptors. It effectively treats locally inoperable and metastatic DFSP by inhibiting tumour growth and promoting cell death.[2-3]

The prognosis for DFSP is favourable, with a ten year survival rate of 99.1%. However, patients with metastatic disease usually have a survival period of about two years after diagnosis.[2] Recurrence is determined by the tumour's location, adequacy of surgical margins, and accuracy of the initial pathological diagnosis.[1,4] Local recurrence rates vary between 20-50%, particularly with positive margins.[1-2] Monsudi et al. reported recurrence at 14 months post-operatively.[8] The head and neck region is the most common site for DFSP recurrence, likely due to the difficulty of achieving free surgical margins.[1,4] Life-long surveillance is recommended, as local recurrence may arise many years after initial treatment.[3]

DFSP arising from a previous surgical scar is uncommon and may mimic benign tumours, leading to diagnostic and management delays. Therefore, early recognition of any mass developing at a prior surgical site, thorough patient counselling, and close long-term follow-up are essential to ensure timely treatment of this locally aggressive tumour and to prevent recurrence.

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