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CASE REPORT

# Recurrent occipital dermatofibrosarcoma protuberans tackled with wide local excision: A case report and current management

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**SUMMARY**

**Dermatofibrosarcoma protuberans (DFSP) is a slow-growing, locally invasive tumour of the dermis. It commonly presents in the trunks and proximal extremities but is seen to a lesser extent in the head and neck regions. We present a case report of a recurrent DFSP in a 48-year-old Iban woman at the occipital region. The patient underwent wide local excision and removal of outer table of cranium, dressing followed by split thickness skin graft. Histopathological examination confirmed dermatofibrosarcoma protuberans with clear lateral surgical margins and a deep margin of 0.5mm. She is currently undergoing radiotherapy and is planned for 50Grey 25cycles.**

## INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous tumour known for its high rate of recurrence and low risk for metastasis. Darrier and Ferrand first described it in 1924, however Hoffman coined the term DFSP in 1925.[1] The most observed site of presentation is the trunk, followed by the extremities and the head and neck.[1,2]

## CASE REPORT

A 48-year-old Iban woman presented to a district hospital with a left occipital swelling in 2013. Histopathological Examination (HPE) post excision showed dermatofibrosarcoma protuberans, with all surgical margins involved. She refused further treatment due to logistic and financial issues, and subsequently defaulted further follow up.

In 2015, she developed a recurrent left occipital swelling which progressively increased in size over the past year. It was painless, smooth surfaced, fixed, no erythema and not fluctuant. A Computer Tomography (CT) scan of the head showed a well-defined exophytic heterogenous enhancing solid mass measuring 1.2cm x 4.0cm x 4.4cm (Fig.1). There was no skull vault or intracranial extension. A wide local excision with a 5cm circumferential surgical margin was taken, complimented with removal of the outer table of the cranium by burring and osteotomy (Fig. 2,2e). The wound was allowed to granulate while awaiting HPE confirmation, then closed by split thickness skin graft after 35 days. Histopathological examination confirmed DFSP with all margins more than 2cm from the tumour, 0.5mm away from deep margin (Fig. 2b). The bony cranium was still undergoing decalcification which may take up to a year to process. Immunohistochemical staining was positive for CD 34 expression, negative for SMA, Desmin, S100 and CKAE1/AE3 (Fig.2c). The wound healed well, and the patient was referred to Oncology team. She is currently undergoing radiotherapy and is planned for 50Grey 25cycles.

## DISCUSSION

DFSP is a rare cutaneous neoplasm, locally infiltrative and has a high frequency of recurrence.[1,2] DFSP accounts for less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas.

It may initially present as a raised, firm pink to red-bluish painless lesion which may ulcerate in time. Its slow growing indolent nature allows it to go unnoticed for many years. Its characteristic microscopical appearance consists of an arrangement of spindle-shaped tumour cells in a "cartwheel" pattern with no nuclear atypia nor increased mitotic activity.[1] However, infiltration of the dermis and surrounding fat is characteristic.[1] It has strong positivity for CD34 on immunohistochemical staining, as well as Vimentin. To differentiate DFSP from benign fibrous histiocytoma (dermatofibroma) or other malignant tumours such as Malignant Peripheral Nerve Sheath Tumours (MPNST) and rare variants of spindle-cell malignant melanoma, Stromelycin 3 (ST3) and factor XIIIa may be employed.[1-2] West et. al proposed Apolipoprotein D as another novel marker to differentiate DFSP from dermatofibroma. If there arises difficulty in diagnosis, fluorescence in situ hybridisation (FISH) or multiplex reverse transcriptase-polymerase chain reaction (RT-PCR) which detects chromosomal translocations and fusion gene transcripts may be considered.[1-3]

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