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REVIEW

Mohs micrographic surgery for dermatofibrosarcoma protuberans (DFSP): A single-centre series of 76 patients treated by frozen-section Mohs micrographic surgery with a review of the literature☆

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Summary Dermatofibrosarcoma protuberans (DFSP) is a rare low-grade sarcoma that typically presents with local invasion but rarely metastasises. Surgical excision remains the first-line treatment for DFSP. There are no randomised controlled or prospective studies comparing wide local excision (WLE) with Mohs micrographic surgery (MMS), but available evidence from the retrospective studies and case series available has consistently shown higher recurrence rates for standard surgery and WLE than for MMS. Combined recurrence rates of data within the last 20 years for WLE have been reported at 7.3% compared with 1.1% for MMS. Our aim was to review the clinical details and recurrence rates of DFSP cases treated with frozen-section MMS in our centre between 1996 and February 2013. The relevant data were collected from the case notes. It involved 76 patients with nine of these patients lost to follow-up. In the remaining 67 (67/76) cases, the recurrence rate was 1.5% during the mean follow-up period of 50 months (2—132). This is comparable to recurrence rates for the MMS in the literature [20,21]. Our series is the largest series for frozen-section MMS reported to date.

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Based on these findings and the current literature evidence, we advocate MMS as the treatment of choice for DFSP in all locations.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumour of mesenchymal origin that is locally aggressive. It extends deep into subcutaneous tissue and has a propensity to invade through fascial planes and into muscle with tentacle-like extensions contributing to a high risk of local recurrence after surgical excision. It is a rare tumour with a prevalence of 0.8—4.2 cases per million persons per year and accounts for between 2% and 6% of all soft tissue sarcomas and is the most common skin sarcoma.[2] It involves the trunk in 50—60% of cases, the upper limbs in 25% of cases, followed by head and neck in 10—15% of cases, but can occur anywhere in the body.[3] Although it can occur at any stage in life, the lesions are most common in the second to fifth decades, with a slight male predominance.[3,5]

Clinically, DFSP is characterised by nodular or plaque-like lesions with a skin-coloured, brown-yellow, red-tinged, sclerodermiform, telangiectatic or atrophic surface (Photograph 1). Usually, it is fixed to the dermis, but freely moving over the deeper tissues except late in the course of the disease or in recurrent tumours. Long-standing lesions may ulcerate. DFSP may increase in size over a period of months to years, to produce large protuberant nodules, for which it is named.[2] It has a propensity to invade through fascial planes and into muscle.[1] Histologically, DFSP is characterised by a monomorphous proliferation of bland-looking spindle cells (frozen-section Photomicrograph 1) with a fascicular and storiform architecture, low mitotic activity and deep, honeycomb infiltration into subcutaneous adipose tissue (frozen-section Photomicrograph 2). Early lesions may have a grenz zone histologically. There are several variants described, which include pigmented (Bednar tumour), myxoid, myoid, granular cell, sclerotic, atrophic, giant cell fibroblastoma variants and DFSP with fibrosarcomatous areas. The tumour margin can be difficult to gauge clinically because the irregular, tentacle-like projections of neoplastic cells can diffusely infiltrate the surrounding dermis and subcutis and deeply invade fascia and muscle. Approximately 85—90% of all DFSPs are low-grade lesions. The remaining 10—15% contain a high-grade fibrosarcomatous (DFSP-FS) component showing a 'herringbone' pattern histologically (frozen-section Photomicrograph 3), and this accounts for 5% of the tumour volume. These lesions have a significantly higher rate of local recurrence and increased risk of distant metastases.[6]

Low-grade DFSP carries a small risk of metastasis (0.5%), and this is usually preceded by multiple local recurrences. In cases of metastasis, the lung (4%) is the most common site, with metastasis to other areas including regional lymph nodes (1%), brain, bone and heart being reported.[7,8] Metastasis is associated with a poor prognosis over the next 2 years. Metastasis usually occurs in the setting of multiple failed treatments of the primary lesion or in the context of fibrosarcomatous changes (DFSP-FS).

A characteristic feature of DFSP is the overexpression of platelet-derived growth factor B (PDGFB), a result of a fusion gene consisting of collagen type Ia1 (COLIA1) and PDGFB-chain genes from the rearrangement of chromosomes 17 and 22, leading to a supernumerary ring (r(17; 22)) or to reciprocal translocation (t(17; 22)).[2] PDGFB activates PDGF receptor b and its tyrosine kinase that results in cell growth and proliferation. In about 8% of DFSPs, this fusion script is not found.[9] CD34 is one of the useful stains to differentiate DFSP from dermatofibroma and other soft tissue tumours with a sensitivity of 84—100%.[2]

Surgical excision remains the first-line treatment for DFSP. The National Comprehensive Cancer Network (NCCN 2014) DFSP guidelines recommendations are that the surgical technique used should aim to achieve clear surgical margins with some form of complete histological margin examination whenever possible. Suggested surgical techniques include frozen-section Mohs micrographic surgery (MMS), Modified/'Slow' Mohs, complete circumferential peripheral and deep margin assessment (CCPDMA) and wide local excision (WLE) with 2—4-cm margins to fascia with clear histological margins.[25] The NCCN 2014 guidelines also suggest that, whenever possible, recurrent tumours be treated surgically.

Our centre has been performing frozen-section MMS for DFSP for the past 17 years. We analysed our cases retrospectively to look at the excision margins required for clearance with MMS and to ascertain the recurrence rate in our series.

Methods

All patients who underwent MMS for DFSP from 1996 to February 2013 were identified from the pathology database. This included patients with both primary and recurrent lesions. The diagnosis of DFSP was established histologically with haematoxylin and eosin (H&E) stain and in some cases with the CD34 staining prior to patients undergoing MMS. The primary disease was defined as individuals who had not previously been treated or who had recently had previous surgery with histological positive margins. The recurrent disease was defined as that occurring in a patient within or juxtaposed with the previous surgical excision site 6 months or longer after the initial excision at a peripheral centre. Medical records of all these patients were reviewed and information on demographics, tumour characteristics, treatment course and follow-up information was collected. Data compiled from each case included the patient's age, sex, tumour location,