Dermatofibrosarcoma protuberans: A review

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Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumour of the dermis. Diagnosis may be very difficult from both a clinical and histopathological standpoint. DFSP has a peak incidence in patients between age 10 and 50 years old, occurs with relatively equal frequency in males and females and has a predilection to occur on the trunk or proximal limbs. The tumour has a very low metastatic potential but can be very locally aggressive, requiring wide surgical resection. The possible variants of DFSP may obscure diagnosis especially in the case of fibrohistiocytic lesions such as dermatofibroma, fibromatosis and malignant fibrous histiocytoma. Immunohistological testing, while not completely specific, shows promise in improving diagnostic accuracy. Wide local excision continues to be the mainstay of treatment, although Mohs' micrographic surgery may ultimately provide superior cure rates.

Key Words: *Case report, Dermatofibrosarcoma protuberans, Fibrohistiocytic tumours, Literature review*

Dermatofibrosarcome protuberans : revue de synthèse

RÉSUMÉ : Le dermatofibrosarcome protuberans (DSFP) est une rare tumeur des tissus mous du derme. Le diagnostic peut être très difficile, tant d'un point de vue clinique qu'histopathologique. L'incidence de pointe du DSFP se situe entre la deuxième et la cinquième décennies de vie (soit à partir de l'âge de 10 ans [début de la deuxième décennie] jusqu'à l'âge de cinquante ans [fin de la cinquième décennie]) et la maladie affecte à peu près également les hommes et les femmes, surtout au niveau du tronc et des membres proximaux. La tumeur a un très faible potentiel métastatique, mais peut être très agressive localement, exigeant une résection chirurgicale large. Certaines variantes possibles du DSFP nuisent parfois au diagnostic, surtout dans les lésions fibrohistiocytaires, comme le dermatofibrome, la fibromatose et les histiocytomes fibreux malins. Les épreuves immuno-histologiques, bien qu'elles ne soient pas entièrement spécifiques, sembleraient prometteuses pour un diagnostic plus précis. L'incision locale large continue d'être au cœur du traitement, bien que la chirurgie micrographique de Moh puisse ultimement donner des taux de guérison supérieurs.

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon soft tissue tumour that is locally aggressive but rarely metastatic. The first published case report of the tumour was by Taylor (1) in 1890. However, the tumour was not characterized as a clinicopathological entity until 1924 (2). The current name, dermatofibrosarcoma protuberans or "lump-producing fibrosarcoma of the skin", is accredited to Hoffman (3), who published three cases from his practice in 1925. Clinically and histopathologically the tumour may be very difficult to differentiate from other fibrohistiocytic tumours such as dermatofibroma, fibromatosis and malignant fibrous histiocytoma. Other lesions commonly included in the differential diagnosis include keloid, localized scleroderma and fascitis (4,5). DFSP develops slowly and may infiltrate widely, giving rise to high recurrence rates. This paper comprises a case report, followed by a detailed review of the clinicopathological features of DFSP and its management.

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CASE PRESENTATION

A 73-year-old man was referred with a four-year history of a progressively enlarging nodule on the posterior aspect of the right shoulder (Figure 1). Examination revealed a 20 mm red plaque with a firm and adherent subcutaneous circumferential extension for 2 cm in all directions. Initial biopsy did not identify a cellular dermatofibroma or a DFSP. Because the

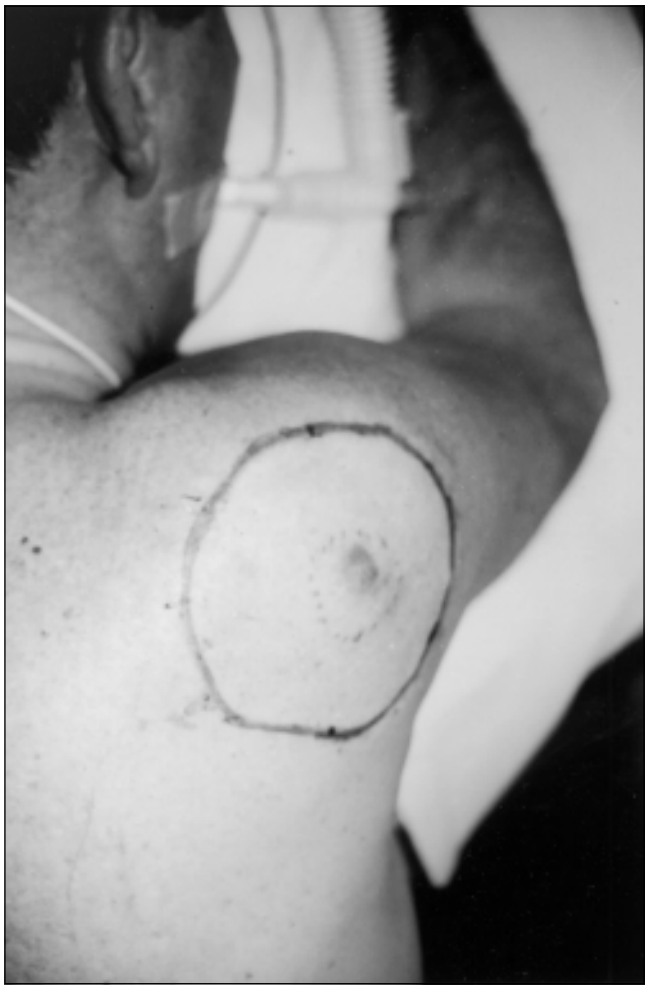

Figure 1) *Planned wide local excision of dermatofibrosarcoma protuberans from right shoulder. Dotted line indicates palpable subcutaneous extension of tumour*

lesion was clinically most consistent with DFSP, the patient underwent a wide excision, with a 3 cm margin of normal tissue around the grossly visible and palpable limits of the tumour (Figure 1). Intraoperative frozen section confirmed that the deep margin, including the deep investing fascia, was clear. The defect was reconstructed with a split thickness skin graft.

Histological examination of the tumour revealed a vague storiform arrangement of slender spindle cells, rare mitotic figures and immunoreactivity with CD 34 antibody, features consistent with DFSP (Figures 2-4). Two years after treatment, the patient remains free from local recurrence.

DISCUSSION

Clinical features

Clinically the tumour usually presents as a solitary raised cutaneous nodule with a red to blue hue (4). Growth of the neoplasm is most often painless and may progress from a dermal plaque to form adjacent nodules that can eventually coalesce into a single structure (6). DFSP lesions usually remain mobile on the underlying fascia but often are very adherent to the overlying epidermis (7). Expansion of DFSP can proceed at various rates ranging from three weeks (6) to several decades (7). Later stages of tumour progression may feature skin atrophy that can result in local trauma, infection and ulceration of the lesion (6). Patients often do not seek medical attention until such local changes develop.

DFSP is most often found on the trunk or proximal limbs, although it may occur at any anatomical location (8,9). The tumours are aggressive locally; their reported size ranges from a few millimetres to well over 20 cm in diameter (4,6,7). The larger tumours are usually seen when medical attention has not been sought or in cases of repeated recurrence.

The incidence of DFSP is relatively uncommon compared with that of other soft tissue tumours. Over 1200 cases of DFSP have been reported in the literature (5), comprising slightly less than 0.1% of all soft tissue tumours (6,7,10). In a series reported by Bendix-Hansen et al (11), DFSP represented 7% of all soft tissue sarcomas and had an annual incidence of 0.08/100,000. DFSP occurs in all age groups, ranging from the neonate (12-14) to the elderly, with a peak incidence between ages 10 and 50 years (9,15,16). DFSP is slightly more common in males (11,16).

Pathology

On gross examination, DFSP is usually a single, sometimes multinodular, greyish-white tumour. While the tumour may appear to be circumscribed, microprojections of the tumour can be far-reaching, contributing to the high local recurrence rate of DFSP (15). Some lesions may display a foci of myxoid change where areas of the tumour appear translucent or gelatinous (5).

Histologically the tumour is a dermal-based proliferation of plump, relatively monomorphic spindle cells showing the classical 'storiform' or 'cartwheel' pattern with varying vascularity and collagenization (17). The storiform pattern is not unique to DFSP because dermatofibroma, malignant fibrohistiocytoma and atypical fibroxanthoma may all display a similar cellular arrangement (18). Cellularity is greatest at the centre of the lesion and gradually diminishes at the periphery where the cells become slender, gradually blending into normal tissue (9). One of the characteristic, almost diagnostic features is the lace-like or 'honeycomb' infiltration of subcutaneous fat. Irregular projections of tumour tissue extend laterally and superficially, often causing atrophy and enveloping sweat glands and pilosebaceous units (7). Later stages of DFSP infiltration may become aggressive, with the tumour extending into fascia and muscle planes (17).

Mitotic figures are relatively rare, even in cellular areas and in rapidly growing tumours (15,16). Chattopadhyay et al (10) suggested that tumours with fewer than five mitoses per 10 different high power fields have a less aggressive course. In one series, three of five lesions with metastases were shown to have eight mitoses per 10 different high power fields (6). Others have speculated that aneuploidy may predict a more aggressive tumour (19). Necrosis is rare and, if present, minimal.

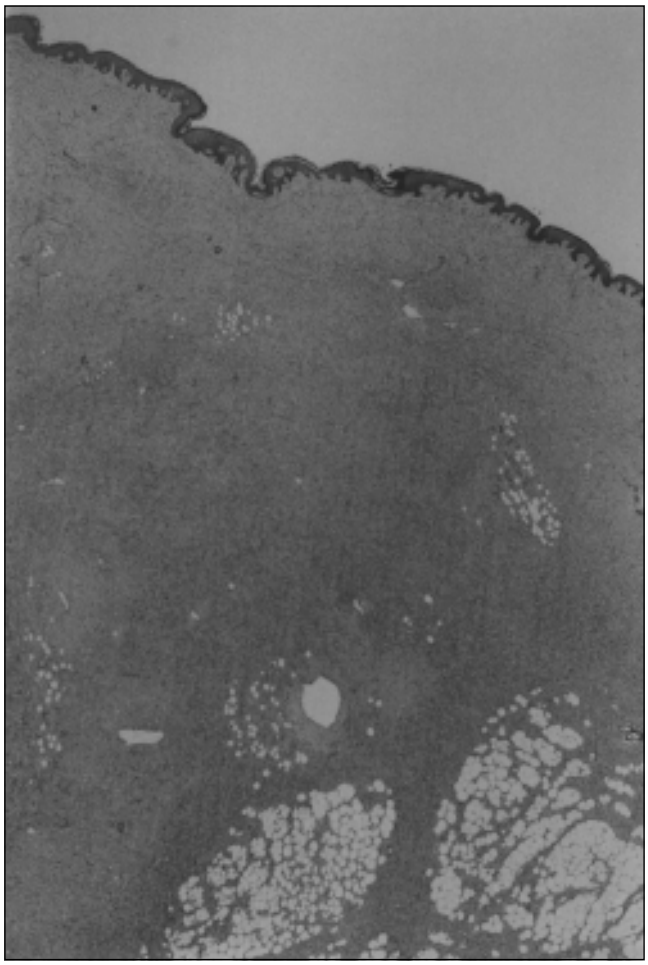

Figure 2) *Dense proliferation of spindle cells occupying the entire dermis and infiltrating the subcutaneous fat with the characteristic lace-like or honeycomb pattern are presented (hematoxylin, phloxine and saffron stain, original magnification x20)*

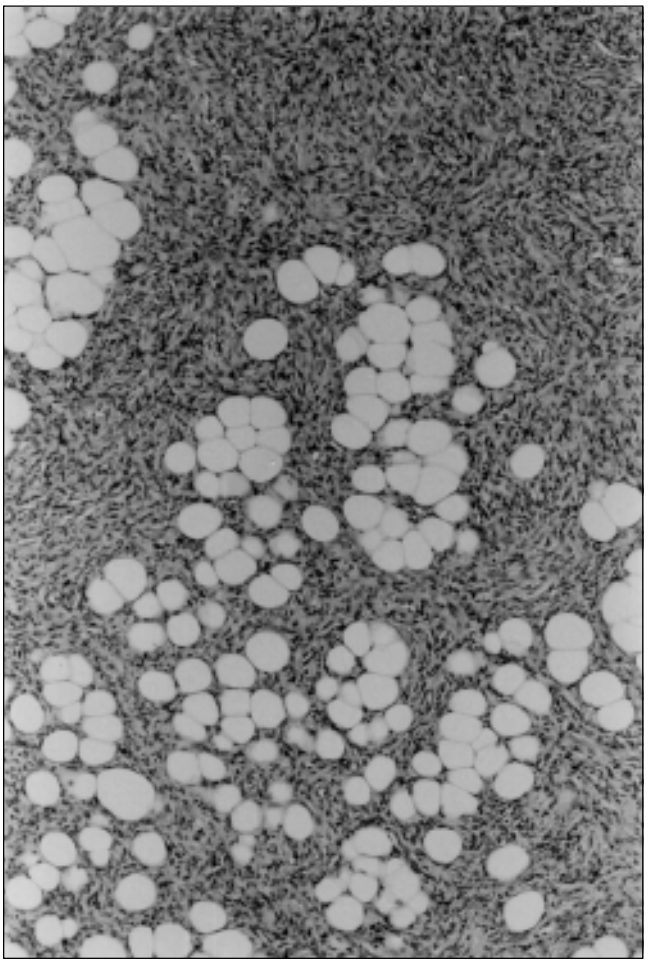

Figure 3) *Relatively monomorphous plump spindle cells arranged in a storiform pattern and infiltrating subcutaneous fat are presented (same view as in Figure 2 but with greater magnification) (hematoxylin, phloxine and saffron stain, original magnification x100)*

Variant DFSP lesions

A major obstacle in diagnosis is that DFSP may exist in one of several variant forms. One of the most frequent abberations is a lesion with myxoid features. It has been estimated that up to 15% of all DFSP tumours display some degree of myxoid change (18,20,21). Previously, myxoid change was thought to be predominantly a feature seen in recurrences, but recent studies have refuted this because microscopic myxoid areas are often seen in initial lesions (22-24). Myxoid foci are characterized by a decrease of storiform or cartwheel patterns, and accumulations of interstitial ground substance containing mucopolysaccharides (5). Myxoid change may interfere with both gross and microscopic determination of tumour boundaries (22).

The so-called Bednar tumour or pigmented DFSP is an unusual variation containing scattered melanin-bearing dendritic cells. Although too rare to be appreciated clinically, approximately 5% of all DFSP tumours may possess melanin-synthesising cells (25). Bednar (26) first described this pigmented lesion in 1957 as a neurofibroma variant. However S-100 protein, present in neural and melanocytic tumours, has not been reported in the pigmented variant or any other

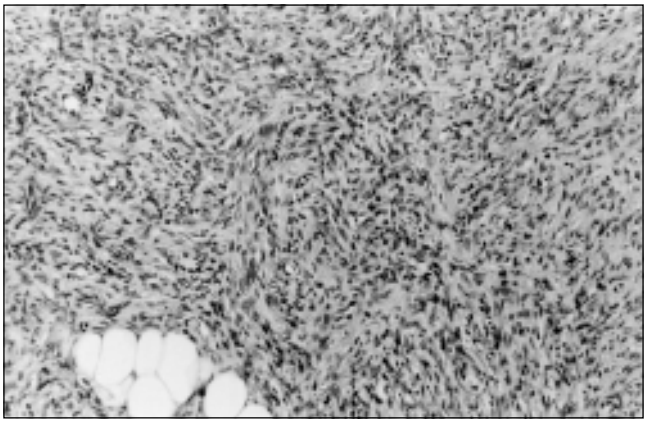

Figure 4) *Cells showing little atypia in a densely collagenous stroma; mitoses are not apparent. The storiform pattern is very visible (hematoxylin, phloxine and saffron stain, original magnification x200)*

DFSP (5). These pigmented tumours range from the rare grossly darkened lesions to others where pigmentation can only be appreciated microscopically (25). Typically, it is only the grossly pigmented variety of DFSP that adds confusion to the clinical diagnosis.

Another variant that creates a diagnostic challenge is DFSP lesions with hemorrhagic features, which may mimic vascular neoplasms. Final diagnosis may require immunohistochemical staining and electron microscopy to exclude vascular origin (27). While this variant is very rare it is important to recognize the possibility of a vascular neoplasm. Angiomatoid features of DFSP may also be confused with vascular variants of other fibrohistiocytic sarcomas (28,29).

One final DFSP variant that may create a diagnostic difficulty is DFSP with dermal atrophy, which can be confused with dermatofibroma (30,31). Atrophic variants of both DFSP and dermatofibroma may present with greater than 50% dermal loss superficial to their respective lesions, increasing the likelihood of misdiagnosis (30).

Histogenesis

A controversial aspect of DFSP is its origin. Since its discovery, researchers have debated the histogenesis of DFSP. Historically the tumour was assigned a broad spectrum of etiologies including parasitic infections (2), misplaced embryonic breast tissue (7,16) and previous trauma (6,7,16). Although histological examination does not support these theories, antecedent trauma continues to be cited as a possible precipitating factor in up to 20% of DFSP cases (5). This figure may reflect that the trauma calls attention to previously unrecognized lesions.

Historically, light microscopy and electron microscopy revealed characteristics suggestive of peri- or endoneural origins (31,32). Concentric layers of tumour cells enveloping capillaries and ultrastructural features such as elaborate convoluted nuclei, extensive cytoplasmic processes and fibrous long-spacing collagen all pointed towards a neural origin (20). Further supporting these observations was Bednar (33) who felt the characteristic DFSP storiform pattern to be a specific feature of neural differentiation and who described the pigmented melanin containing DFSP (26).

Currently, few researchers subscribe to a neural origin theory; instead they view the endoneural and perineural features to be characteristic of fibroblastic genesis. Classic immunohistochemical markers for neural derivation (s100 protein, Leu 7 antigen and neuron-specific enolase) are absent in DFSP (34,35).

Immunodiagnosis

Immunohistochemical staining has generated considerable research into the diagnosis and origins of DFSP, with CD 34 and factor XIIIa being the most prominent markers under investigation. Positive CD 34 staining for DFSP was first described by Ramani et al (36) in 1990; subsequently great interest has been shown in this marker (37-43). The antigen CD 34 is a transmembrane glycoprotein encoded by a gene located on chromosome 1 (44). Cells that possess the membrane protein include endothelial cells, dermal dendritic cells and hematopoietic progenitor cells. Approximately 88% of DFSP tested for CD 34 are positive, with a strongly diffuse pattern (41). However CD 34 is not specific to DFSP and is present in a wide range of neoplasms, including endothelial cell tumours, epithelioid sarcomas, some smooth muscle tumours and peripheral neural sheath tumours. CD 34 is useful in distinguishing DFSP from dermatofibroma because dermatofibroma is usually CD 34 immunonegative (41,42).

Factor XIIIa is an intracellular analogue of the fibrin stabilizer formed during the clotting cascade (35,42). This antigen has been identified in various dendritic cells found in reactive lymph nodes (45), fibrohistiocytic tumours (46,47) and some epithelial tumours (42). Factor XIIIa is not found within DFSP tumour cells (35,42,48); however, the lesional cells are positive in 90% of dermatofibroma cases (42). While dermatofibroma contains factor XIIIa, it may only stain weakly positive (49,50).

Treatment

The treatment of choice for DFSP is wide surgical excision. A 3 cm margin of normal tissue around the visible tumour was recommended as early as 1903 by Johnson (51). Margins of 2.5 to 3 cm are still commonly used (5,6,7,16,52-55). Invasion of DFSP into surrounding tissue requires a 3 cm margin of grossly normal tissue, both peripherally and deep to the tumour (5,11). This margin usually results in resection of the deep fascia (5) and, in certain anatomic locations such as the scalp, resection of the periosteum (19,53,56). A recent study (57) recommends an even more aggressive approach in which deep resection is combined with removal of tissue 5 cm peripheral to the tumour.

Local recurrence is a well known phenomenon and is reported to occur in 11% to 73% of cases (6,11,16,53). Recurrence is related to the locally aggressive biology of DFSP. These tumours have the ability to microscopically infiltrate adjacent dermis, fat, fascia, muscle and even bone well beyond the limit that can be appreciated on gross inspection of the tissues at surgery (6). The variability among studies of local recurrence rates is a function of imprecise definitions of 'wide' resection and different follow-up periods. Most local recurrences develop within one to three years (55), but recurrences as late as 19 years have been reported (7,16). The risk of local recurrence is inversely related to the width of surgical resection (53,54). Lymphatic invasion does not generally occur; hence prophylactic lymph node dissections are not routinely performed (6).

The low but potential metastatic ability of DFSP is now accepted (9). Large tumours (greater than 15 cm) or a preceding history of multiple local recurrences may be associated with a higher risk of systemic metastases (6).

The wide resection of DFSP usually necessitates skin grafts or flaps for closure of the surgical wound. Mohs' micrographic surgery (MMS) has been advocated as a very useful method of treating DFSP. MMS has the capability of providing more complete surgical resection of the tumour while maximizing tissue preservation (56,58-62). While the literature to date supports MMS as a highly effective treatment for DFSP, it has been used in only a relatively small number of cases compared with traditional wide local resection. Thus its true role, while very promising, is not yet clear. Adjuvant therapies such as radiation and systemic chemotherapy have no role in the treatment of DFSP.

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