Rare disease

Dermatofibrosarcoma: a rare neoplasm of skin

Rizwan Aziz, Anwer Jamil

Department of Surgery, South West Acute Hospital, Enniskillen, UK

Correspondence to Rizwan Aziz, drizwanaziz@hotmail.com

Summary

A 25-year-old lady presented with a small pea-sized lesion on left side of her neck, anteriorly. There was no history of any other lesion on her body. She was medically fit otherwise. An excision was planned, after taking consent from her, which was performed under local anaesthesia as a day procedure. Her postoperative recovery was uneventful. Histopathology showed dermatofibrosarcoma protuberance which is an extremely rare skin tumour. She was further investigated for metastasis. Her CT scan of chest, abdomen and pelvis was performed which showed prominent thymus. Other than this finding, no other abnormality was found in CT scan.

BACKGROUND

Dermatofibrosarcoma protuberance (DFSP) is a very rare neoplasm of the dermis layer of the skin, and is classified as a sarcoma. There is only about one case per million per year. DFSP is a fibrosarcoma, more precisely a cutaneous soft-tissue sarcoma. In many respects, the disease behaves as a benign tumour, but in 2–5% of cases, it can metastasise, so it should be considered to have malignant potential. It accounts for approximately 2–6% of soft tissue sarcoma cancers.

It starts off as a minor firm area of skin most commonly about 1 to 5 cm in diameter. It is a slow growing tumour. It is usually diagnosed through a biopsy and is treated primarily by surgical resection.

CASE PRESENTATION

A 25-year-old lady was examined in surgical outpatient clinic who was referred by general physician with the complaint of a pea-sized skin lesion on the left side of her neck anteriorly. She had no significantly related family history. She was a cooperative, nice lady sitting comfortably. On examination, she had about 1 cm lesion on the left side of her neck. All her routine blood counts were sent, which turned out to be in normal range for her age. So an excision was planned.

The lesion was excised under local anaesthesia and she was discharged home the same day. Her postoperative recovery was uneventful and the stitches were removed after 7 days. The excised lesion was sent for histopathology and the report showed dermatofibrosarcoma protuberance. The report was discussed at dermatology MDT and a wide local excision and metastatic review was planned on the basis of report because there are chances of spread and local recurrence of this tumour. The findings and further plan was discussed with patient and she was booked for a CT scan of chest, abdomen and pelvis, which showed a prominent thymus. Other than this, finding, the CT scan report was unremarkable. Her wide local excision was also performed during further course of treatment and the histopathology turned out to be negative for residual tumour.

She was referred to cardiothoracic surgeon regarding the incidental finding of prominent thymus which was slightly bigger for her age. According to his opinion, there was no need for any further management regarding thymus. Her PET scan was performed in the mean time which was also negative.

OUTCOME AND FOLLOW-UP

She was examined at the surgical outpatient department on regular intervals and there was no evidence found of local recurrence after 1 year of successful follow-up. Further follow-up visits are planned at the surgical outpatient department.

DISCUSSION

DFSP is a low-to-intermediate grade soft tissue sarcoma originating from the dermal layer of the skin. Although, historically, it has been attributed to fibroblastic origin, recent immunohistochemical evidence suggests that it may arise from the dendritic cell in the skin. In 1924, Darier and Ferrand first described the entity of DFSP as a 'progressive and recurring dermatofibroma,' underscoring its predilection for local recurrence.¹ DFSP is a locally aggressive tumour, and despite sharing some histological features with fibrohistiocytic tumours, it tends to grow in a more infiltrative manner. Three-dimensional reconstructions of DFSP have shown that the tumour can assume irregular shapes and extend in a villous or finger-like manner.² Its local recurrence is a common clinical dilemma; metastases are rare and usually occur late in the disease course.

DFSP comprises roughly 0.01% of all malignant tumours and approximately 2–6% of all soft-tissue sarcomas.^{3–4} The estimated incidence is 0.8–5 cases per 1 million persons per year.^{5–9} The incidence among African-Americans (6.5 per million) is almost double than among Caucasians (3.9 per million).¹⁰ It most commonly affects patients between 20 and 50 years of age, although it has been described in both children and in the elderly.¹¹ Congenital DFSP is a recognised entity but is extremely uncommon.^{11–14}

DFSP usually has a long-and-slow course, with early tumours appearing as painless areas of skin-thickening. They may have pink, red or bluish discolouration, particularly at its periphery. With the passage of time, they develop into a larger nodular mass, and ultimately, can develop into a large fungating lesion. When they grow into the epidermal layer of the skin, they may eventually ulcerate. Unlike tumours of the subcutaneous tissue, DFSP is adherent or intimate with its overlying skin. Typically, it is not adherent to underlying structures. Most tumours are superficial and less than 5 cm in size when they are diagnosed.¹⁵

DFSP can often be mistaken for lipomas, deep-seated epidermal cysts, scars, hypertrophic scars, keloid, dermatofibromas, nodular fasciitis and insect bites,¹⁶ and a delayed diagnosis is not uncommon. The trunk is the most common location (47%), followed by lower extremity (20%), upper extremity (18%) and finally, head and neck (14%).¹⁵

DFSP metastasises 1–4% of the time.¹⁵ Metastasis is a late clinical outcome and typically occurs only after several local recurrences.¹⁷

Although routine imaging is not necessary, MRI may be helpful to evaluate the local extent of the tumour and may be important in preoperative planning for larger tumours.

Diagnosis is made using either a core needle or an open incisional biopsy. Whereas the role of fine needle aspiration is established in cases of recurrent disease, initial biopsies should be larger samples that demonstrate the histologic architecture of the tumour.¹⁸

The mainstay of treatment of DFSP has been surgery. Because of the high rates of recurrence, historical recommendations have sought 5 cm margins.¹⁹

However, with the advent of Mohs surgery, complete excision with microscopic margins has yielded excellent outcomes and offers the benefit of decreased surgical morbidity. In a comparative study of wide resection versus Mohs surgery, wide resection was associated with a recurrence rate of 13% whereas Mohs surgery had no recurrences at 5 years.²⁰

The general prognosis for DFSP is excellent. The overall rate of distant metastasis is only 5% and regional metastasis is 1%.²¹ Historically, recurrence rates have been high, ranging from 11 to 53%,⁹ but with the advent of Mohs surgery, the rates have dropped. Even with recurrent DFSP, Mohs surgery has a 98% cure rate.²² Metastasis is associated with a poor prognosis, with few patients surviving past 2 years. With initial encouraging results using imatinib, improved prognosis even in cases of metastatic disease may be realised.

Competing interests None.

Patient consent Obtained.

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Learning points

- Any suspicious skin lesion should not be ignored and should be excised and sent for histopathology.
- A full metastatic workup should be done and patient should be regularly followed up, if it turns out to be malignant.

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