

Dermatofibrosarcoma Protuberans of Abdominal Wall: A Case Report

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Abstract

Introduction: Dermatofibrosarcoma protuberans of abdominal wall is very rare. Surgery is treatment.

Case Report: A 36 year old female presented with painless abdominal swelling with cosmetic blemish. Excision of swelling was done. Histopathology was consistent with diagnosis of dermatofibrosarcoma protuberans.

Conclusion: A painless abdominal swelling is usually present. Wide excision is treatment.

Keywords: Abdominal Swelling; Dermatofibrosarcoma; Malignancy

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue neoplasm with long latency period that persists for years without symptoms [1]. Secondary complication usually make patient to seek medical advice. DFSP accounts for less than 0.01% of all malignant neoplasms [2]. This cutaneous malignancy arises from the dermis with a pattern of slow, progressive growth. These affect men more than women, commonly present on trunk and proximal extremity [3]. The presentation is asymptomatic elevated, firm nodular lesion free from underlying deep fascia. This has origin from fibroblastic, histiocytic or neuroectodermal cells and usually has benign course, but sometimes it exhibits malignant potential. Complete surgical excision is essential for DFSP of the abdominal wall [4]. Chemotherapy plays limited role. Radiotherapy is used adjunct to surgery. There is to be long term follow up for detecting recurrence and metastasis.

Case Report



Figure 1: Showing Abdominal wall swelling which was diagnosed as Dermatofibrosarcoma Protuberans

A 36 year old female presented with painless swelling of 27 months duration having gradual increase in size on abdominal wall. There was no antecedent history of any previous repeated skin trauma, irritated skin or scar. General physical and systemic examination was normal. Local examination 9.3×4.6×3.7 cm swelling with erythematous patches and the hypo pigmented areas present (Figure 1).

The swelling was firm mobile having nodular surface. Adherent to overlying skin, free from underlying fascia. High resolution USG abdomen and Computed tomography showed anterior abdominal wall mass with no invasion to underlying structures. Wide excision of swelling along with skin and primary closure of wound was done. Histopathological examination of specimen was consistent with diagnosis of dermatofibrosarcoma Protuberans. Immunohistochemical staining was positive for CD34 and 3 years with no evidence of metastasis.

Discussion

The term 'Dermatofibrosarcoma Protuberans' was coined by Hoffman in 1953 [5]. Dermatofibrosarcoma protuberans (DFSP) is a rare fibroblastic skin tumor of intermediate malignancy and is characterized by triad of rarity, slow growth and infiltrative nature. Its pathogenesis has not yet been fully clarified. Any age can be affected, characteristically appears in the younger age group (20–50 years), but it may also affect children and the elderly [6]. DFSP is usually affixed to overlying skin and is firm and indurated. Common site of DFSP is trunk, lower extremities, head and neck [7]. Size at the time of presentation is variable, may present initially as slightly raised sclerotic plaque like mass or small nipple like projection on the surface of skin of repeated trauma, vaccination site, irradiated skin, scar or in decorative tattoo [8]. There are multiple satellite nodules at periphery of main lesion to obtain its typical protuberant appearance. There can be infarction and spontaneous involution in areas of extremely large and neglected tumour. Pressure on the surface of lesion cause blanching and tumour is usually skin coloured with a brown yellow or red tinge. Occasionally this discoloration precedes the development of definite tumefaction [9]; therefore this type of presentation can be misinterpreted as a keloid [10]. Dermatofibrosarcoma protuberans has been compared clinically as a morphea type of basal cell carcinoma or scleroderma [2,7]. Despite the enormous size of some DFSP patient may appear well without signs of cachexia. With progression of time, there is rapid growth of tumour and patient can present with ulceration and bleeding and gets adherent to deeper structures, such as muscle and fascia. 1% to 4% of DFSP may develop distant metastases [11]. Cytogenetic observations show some chromosomal anomalies in etiology of the DFSP in the literature [12]. Dermatofibrosarcoma protuberans is genetically characterized by the unbalanced chromosomal translocation usually in the form of a supernumerary ring chromosome. The extent of the tumors can be estimated by Computed Tomography and more precisely with Magnetic resonance imaging. The diagnosis is made histologically. Differential diagnosis includes atypical dermatofibroma and dermatomyofibroma, as well as from malignant fibrous histiocytoma, Giant cell dermatofibroma, neurofibroma, atypical fibroxanthoma, classic fibrosarcoma, fasciitis, myxoid liposarcoma, myxofibrosarcoma. Immunohistochemistry differentiates these from DFSP. Modality of treatment of DFSP is wide resection, namely with margins of 3 cm beyond the evident disease and histological negative margins. The significant prognostic factor is the extent of resection. Distant metastasis is rare with a rate of 4-6%, predominantly to the lungs and metastasis is preceded by multiple local recurrences [13]. Local recurrence rates are high (13% to 60%), especially in DFSP with fibrosarcomatous histopathology [11]. Moh's micrographic surgery has been associated with lower rates of recurrence [14]. Difficulty in implementing this intraoperative pathology limits its practice use. Histographic surgery involving re-excision of tumor-positive areas until tumor-free margins are obtained leads to high cure rate (97 %) and preserving normal tissue [15]. Imatinib in unresectable, recurrent, metastatic dermatofibrosarcoma protuberans that are not amenable to surgical resection show marked response [16]. Radiation therapy may be recommended if the margins of resection are tumour positive or wide excision alone may result in functional deficits and repeated surgery may cause functional impairment [17,18]. Close follow-up care after radiation therapy is recommended as some of the tumors may become more aggressive.

Conclusion

A painless abdominal swelling is usually present. Patient seeks advice for secondary complications. Excision is treatment.

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