ORIGINAL ARTICLE

Dermatofibrosarcoma Protuberans presenting in young male

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SUMMARY

Dermatofibrosarcoma protuberans (DFSP) is a rare tumour of skin and soft tissue. A 35 years old male presented in December, 2005 with a recurrent soft tissue swelling on the left anterior chest wall which had been increasing in size for the last one year. He had a nodule removed from same site 6 years back. The soft tissue swelling on biopsy turned out to be Dermatofibrosarcoma protuberans (DFSP). Case history is presented with review of literature.

Keywords: Dermatofibrosarcoma Protuberans, Cancer, Skin Cancer, Soft Tissue Sarcoma

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, intermediate to low-grade soft tissue malignancy thought to be of fibrohistiocytic origin, arising from dermis and subcuticular fat. It is locally aggressive but rarely metastatic skin tumor which tends to recur after excision1. Hoffmann2 first named this tumour as dermatofibrosarcoma protuberans. Other terms used to describe this neoplasm are hypertrophic morphea, progressive and recurrent dermatofibroma, fibrosarcoma of skin and sarcomatous tumour resembling keloid3,4. We describe case history of a patient who presented with similar tumour on the chest wall.

CASE HISTORY

A 35 years old male presented in the surgical outpatients of our hospital in December 2005 with a mass on the left anterior chest wall near the breast for the last 1year. Initially, it started as a slightly painful swelling on the left anterior chest wall. He had a nodule removed from same site 6 years back. Gradually, it increased in size and became a large mass. At the time of presentation he had a pinkish-brown, well-defined, firm mass of uneven surface measuring 8-10 cm in diameter. It was situated on the upper left anterior chest wall near the nipple of breast. There were multiple nodules of variable sizes, ranging from 1-3 cm size, within and close to the mass (Fig.1). The mass was slightly tender and fixed to the overlying skin but moveable over the deeper structures. There was no regional lymphadenopathy. The routine laboratory parameters were within normal range. Chest X-ray and abdominal ultrasonography were unremarkable. Biopsy was taken and histopathological examination showed normal epidermis with densely packed monomorphous spindle cells arranged in a storiform pattern throughout the dermis. There was mild pleomorphism and mitotic figures were infrequently present. A histological diagnosis of Dermatofibrosarcoma protuberans was made. We adopted a two-stage procedure. In the first stage the whole tumour was radically excised (Figs. 2 & 3). Post operatively histopathology confirmed tumor free margins of resection. In the second stage the chest wall defect was covered with latismus dorsi myo-cutaneous flap. The patient had uneventful recovery and was discharged after 5th day of second stage procedure. No recurrence of the tumor has been noticed in the 24-month follow up (Fig. 4).

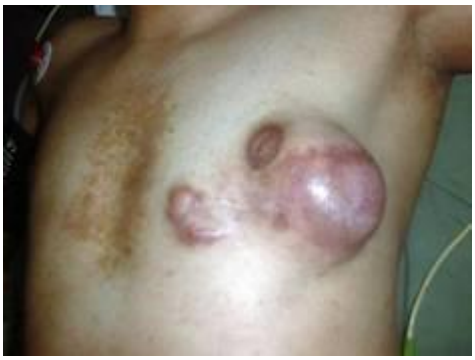

Fig. 1. The Pre-operative image of the lump on the anterior chest wall. Note the scar of previous surgery on the lump.

Fig 2: The radical excision of the lumps on the chest wal

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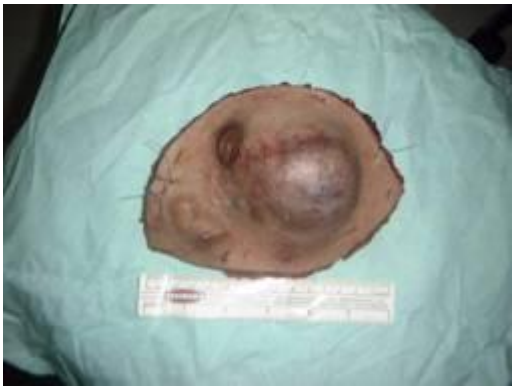

Fig. 3. The excised specimen. Contains all the nodules.

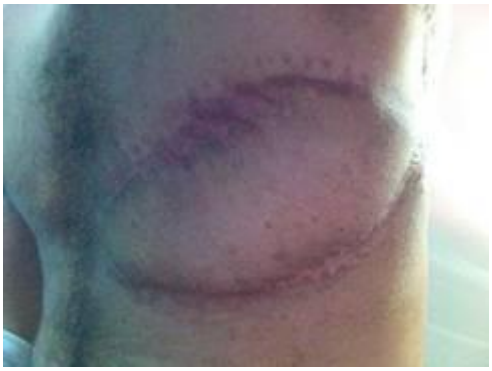

Fig 4.The post-operative result after one month of Latissmus dorsi flap cover of the residual defect on the chest wall after radical excision of the dermatofibrosarcoma protuberans.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) clinically presents as a reddish macule to protuberant nodule affecting the trunk and proximal extremities of young and middle-aged adults5. The tumour can appear as a single fibrous nodule in skin, with a pink or violaceous hue, which is freely mobile on deeper structures4. Our patient was a young man who presented with pinkish-brown mass affecting the left anterior chest wall with multiple nodules. Age at onset of the disease has ranged from 6-65 years while the maximum incidence is in 2nd and 3rd decade. A total of five cases1,6,7,9 have been published in the national literature. All of them were females with an average age of 37 years. We are reporting this disease in a male of 35 years.

Though no anatomical region is spared, the tumour has marked predilection for the trunk4, followed by head and neck regions. Tamoney [8], reported 261 cases, out of which 40 were involving head and neck. In the Pakistani reported cases, 4 were involving trunk while one was involving the lower extremity9.

Condensation of connective tissue at the periphery may give a false appearance of encapsulation but, actually, the tumour may extend well beyond margins in fine microscopic projection4. Microscopically it is composed of intermingling of capillaries, fibroblasts, histiocytes and inflammatory cells. It shows focal collection of foamy histiocytes and occasional tuton giant cells, large blood filled spaces and extensive hemosiderin deposition. Focal nuclear pleomorphism and mitotic activity is seen6.

The tumour enlarges by expansile growth, while the periphery of tumour infiltrates the adjacent skin and subcutaneous tissue. The spread by haematogenous route is seen in 4-6% of cases3. Recurrence is a well-known complication after excision reported in 80% of cases within first three years in one study3. In our case, the patient had a simple excision of a lump on the chest wall without any formal biopsy. 5 years later he had a recurrent mass in the same area. The local recurrence of the tumour stems from its infiltrative capability that is not widely appreciated. In most of the cases the treatment of choice is wide surgical excision ensuring adequate margins of 3 to 5cm10. We adopted a two-stage procedure. In the first stage the whole tumour was radically excised with wide margins. Post operatively histopathology confirmed tumor free margins of resection. In the second stage the chest wall defect was covered with latismus dorsi myocutanious flap. Two-stage procedure was chosen because it was a recurrent case and we wanted to make sure that the excised margins were free from any tumour infiltration by definitive histopathology. Otherwise one-stage procedure is possible with the help of frozen section histopathology to confirm the clear margins of excision.

In conclusion we are reminded of the principle not to dismiss any swelling or lump after excision as benign unless confirmed by histopathology. A high index of suspicion is required to pick up rare cases like Dermatofibrosarcoma. An unusual lump on an unusual site demands a definitive histpathology after biopsy.

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