Journal of Cardiovascular Disease Research

ISSN: 0975-3583, 0976-2833 VOL13, ISSUE 08, 2022

Case report

A MISDIAGNOSED CASE OF RECURRENT DERMATOFIBROSARCOMA PROTUBERANS

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Introduction:

Dermatofibrosarcoma (DFSP) is a rare tumour of skin. It is a locally aggressive tumour and highly recurrent, metastasis is less common. DFSP is more common in women than men (53% in a study of 6817 individuals) [(1)]. It is predominantly seen in individuals aged between 20 to 60 years. Mean size of the lesion is 4.4 to 4.9 cm [(2,3)]. Key challenge with this tumour is diagnosis. Diagnostic delay is very common. Median delay in diagnosis was 3 to 5 years [(2,3)]. Mean size of the lesion is 4.4 to 4.9 cm [(2,3)]. Wide local excision with Mohs microsurgery is usually the treatment of choice. Non-Protuberance is also in many cases which causes delay in diagnosis. Entire deep and circumferential margins are sent for pathological assessment. Conservative excision is followed for tumours in sensitive areas wide local excision or Mohs microsurgery is not possible. DFSP is radiosensitive and Imatinib therapy may be used [(6,7)].

Clinical scenario: A 48-year-old female from Kanchikacherla came to OPD with a chief complaint of swelling over left infraclavicular region since 15 months. Patient was apparently normal 15 months back swelling started in peanut size and gradually increased. Rapid growth was observed in last 5 months. A healed ulcer was seen over pedunculated swelling. No history of pain in the swelling. No history of fever. No H/O trauma, weight loss and chronic cough. Patient had history of excision of lipoma near the present swelling 6 years back. Patient is known case of Hypothyroidism. Bowel and bladder habits are normal. No history of Diabetes mellitus, hypertension and Ischemic heart disease. Patient is on thyroxine 25 mg since 10 years. On inspection, a swelling of size 9*7 cms is seen over the left infraclavicular region. surface of the swelling is nodular. Healed ulcers are seen over the swelling. No expanse cough impulse. No discharge is seen. On palpation No local rise of temperature. No tenderness. Consistency is firm and swelling is arising from the skin. No deeper extension. Swelling was freely mobile vertically and limited horizontally. It is not reducible. No generalized lymphadenopathy. Patient is moderately built, cooperative, coherent and conscious. B.P is 120/70.Respiratory rate is 12-16 breaths/minute. Pallor, icterus, cyanosis, clubbing was not seen.

Investigations:

Incisional biopsy:

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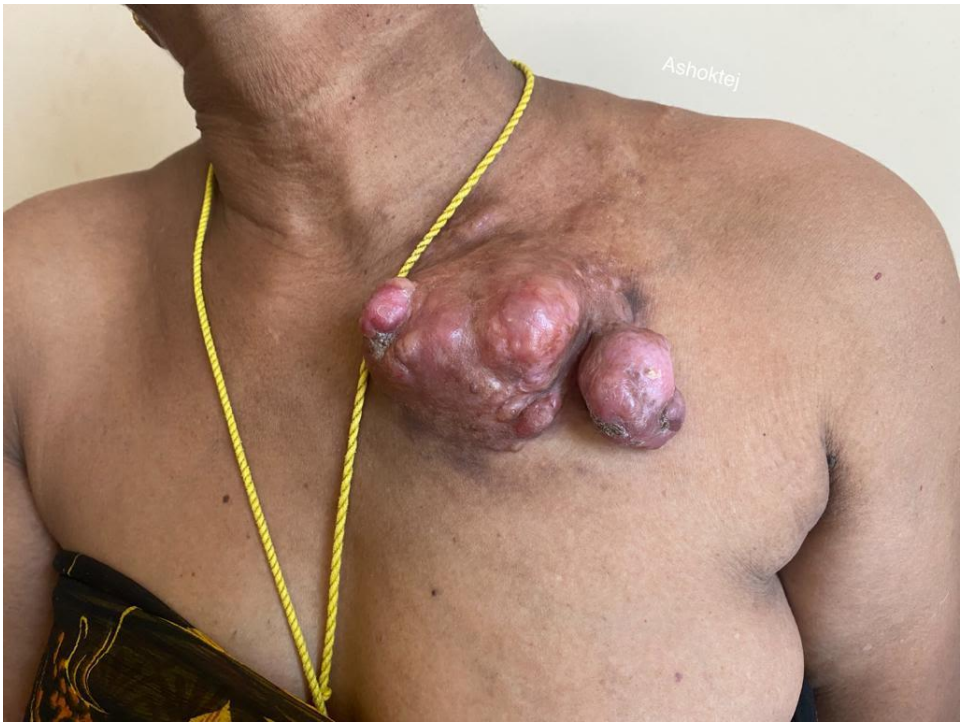
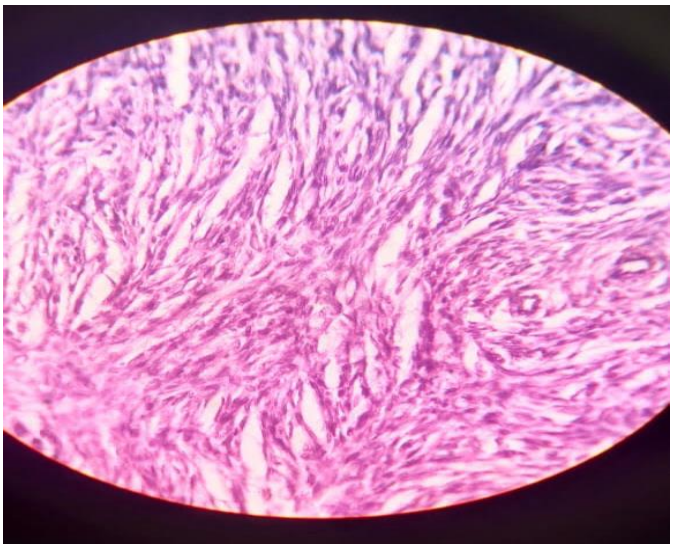


An incisional biopsy was done and sent for histopathological study and the histopathological features favored dermatofibrosarcoma protuberans. The tumour composed of interwoven bundles of spindle cells with plump nuclei arranged in a storiform or cartwheel pattern. CT scan showed no bone involvement. Chest x-ray was normal. Thus final diagnosis of Dermatofibrosarcoma protuberans was made. Wide local excision was done. Edges were approximated and sutured under General anesthesia. Radiotherapy was given. Patient was advised follow up after 3 months.



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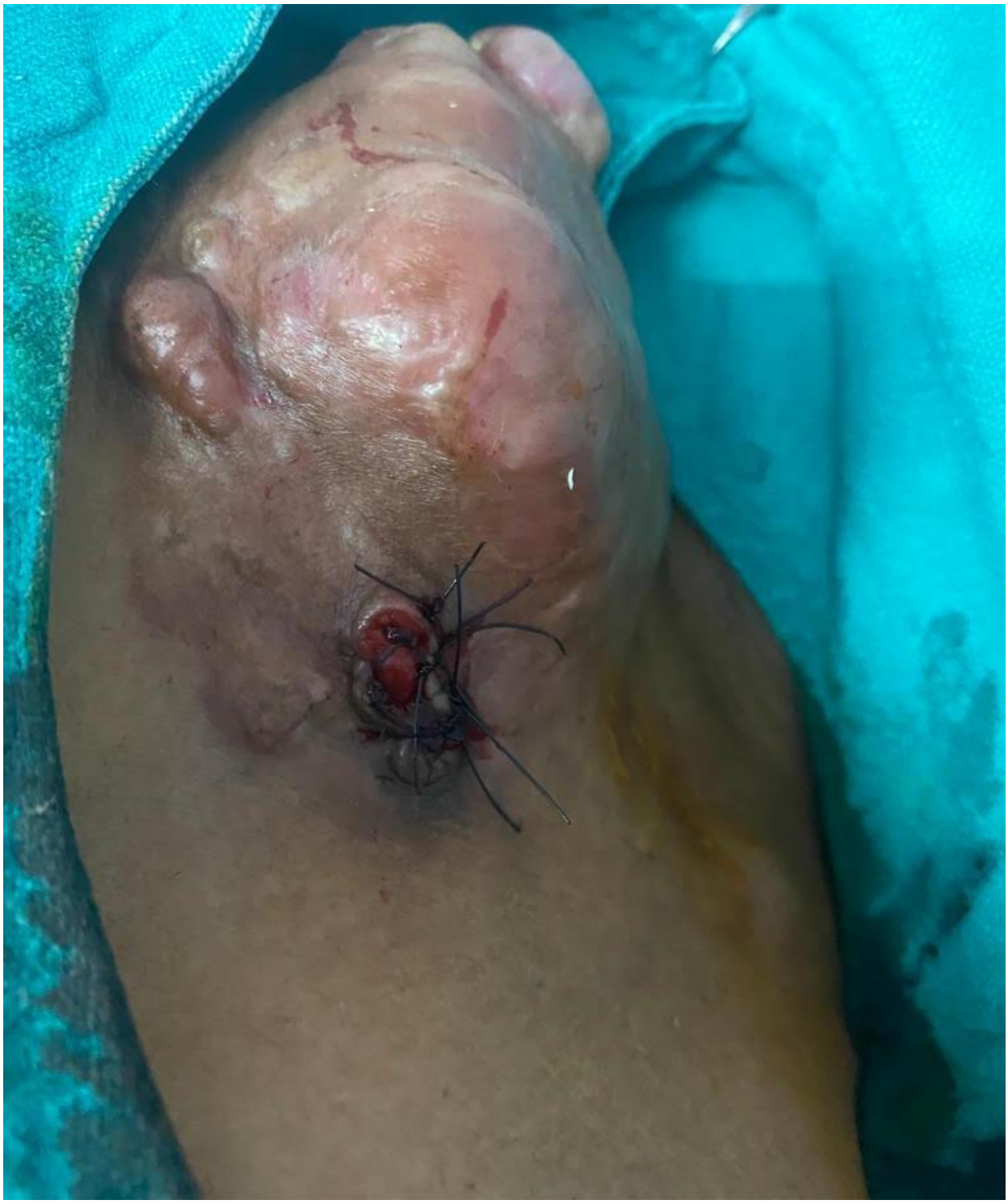

Chest x ray:

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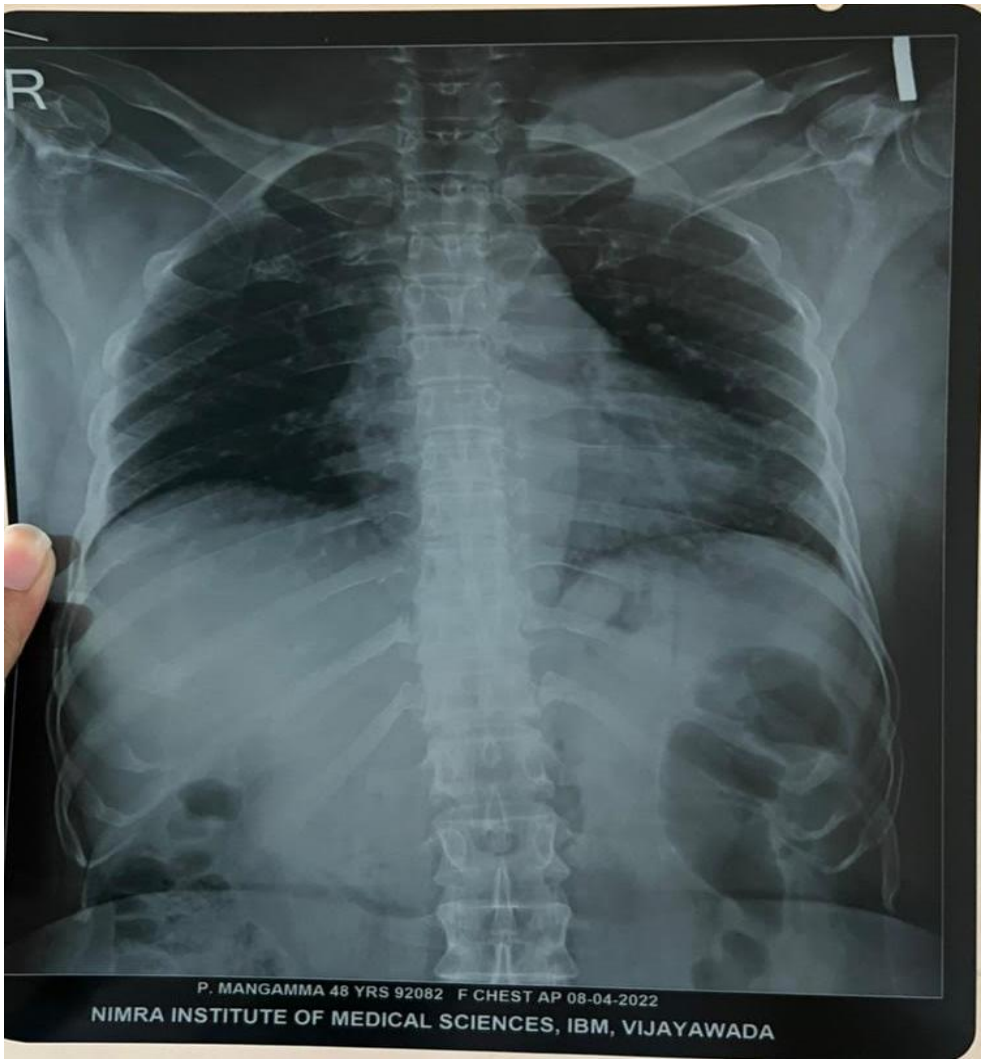

Discussion: Key challenge with this tumour is diagnosis. Diagnostic delay is more prevalent as this tumour starts of flat resembling benign cysts, lipoma, hypertrophic scar and Keloid. Recurrence rates are equal in both Wide local excision and Mohs microsurgery. In our case there was no need of reconstructive surgery.

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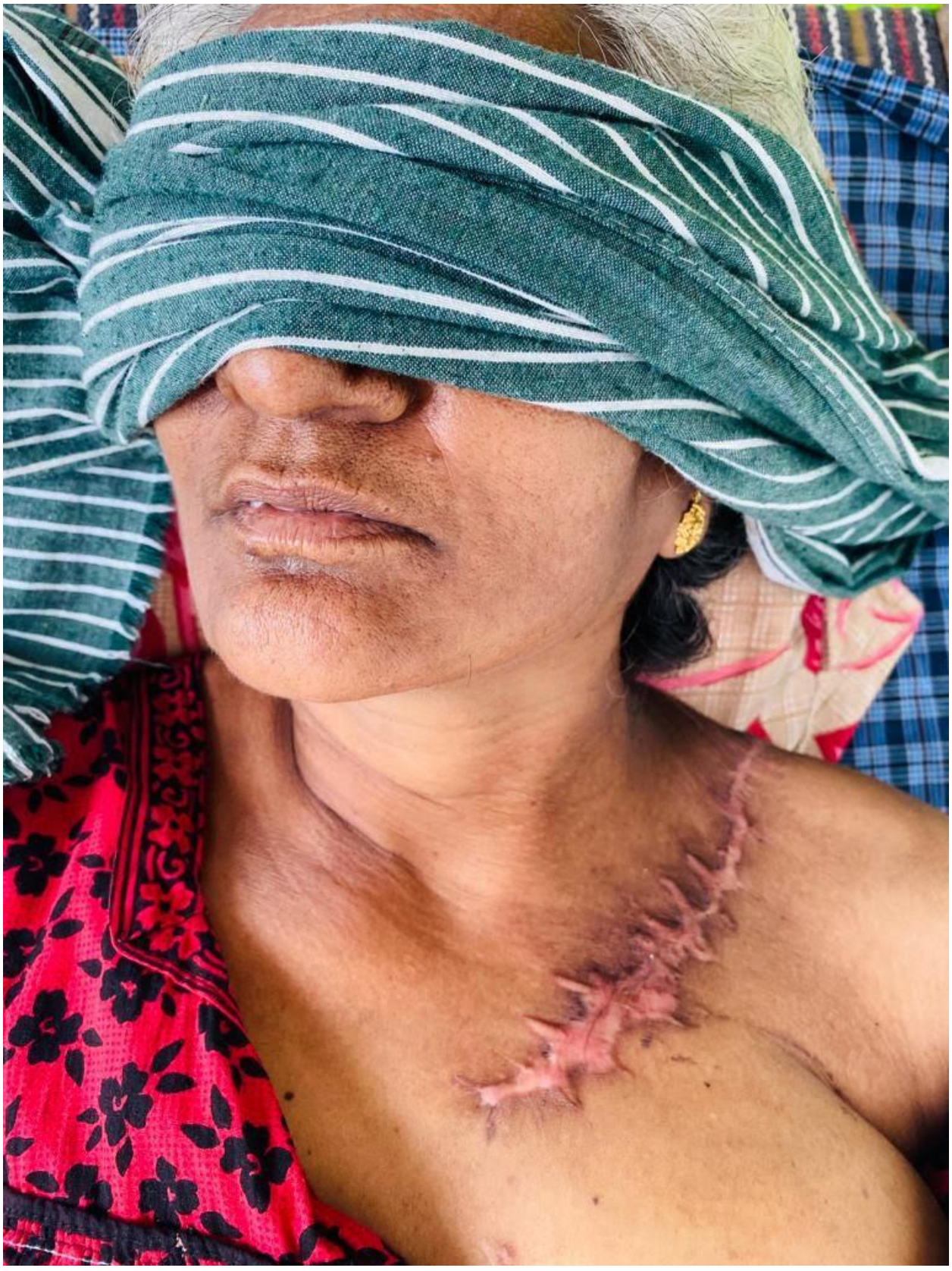

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Conclusion: Lesions were first seen by patient in 20's with a different perception. Diagnostic delay is highly prevalent. Early detection is necessary. Recurrent tumours can be treated with surgery, radiotherapy and Imatinib therapy.

Keywords: Mohs, DFSP, Keloid, hypertrophic scar, radiotherapy

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