

CASE REPORT

A RARE CASE OF RECURRENT DERMATOFIBROSARCOMA PROTUBERANS

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ABSTRACT: Dermatofibrosarcoma protuberans is a locally aggressive, uncommon, cutaneous, soft tissue sarcoma. It most commonly occurs in the trunk and extremities but rarely may also occur in the head and neck region. The local recurrence rate is 20-50% in cases with incomplete resection. We are reporting a case of recurrent dermatofibrosarcoma protuberans which was seen over the supraclavicular region.

KEYWORDS: skin neoplasm, recurrence, soft tissue sarcoma, dermatofibrosarcoma-protuberans.

INTRODUCTION: Dermatofibrosarcoma protuberans is a locally aggressive, uncommon, cutaneous, soft tissue sarcoma. The tumour has low chances of metastasis, either to the regional lymph nodes or distantly, but it is aggressive locally. Wide excision with histologically negative margins is the cornerstone of treatment, but relatively high recurrence rates are described in the literature.

CASE REPORT: A 60 year old lady presented to the Surgery department with an ulcerated swelling in the left supraclavicular region since 1 year. It was associated with bleeding since 1 month. She was operated for the swelling at the same site 3 years back. She was asymptomatic for 2 years and had noticed a swelling gradually increasing in size at the same site.

EXAMINATION: An elderly female, moderately built and poorly nourished with stable vitals was found to have a globular swelling, firm to hard in consistency in the left supraclavicular region measuring 6cm x 5cm with bleeding ulcer over its summit. It was nontender and fixed to the underlying clavicle. A single nodular nontender lymph node 2cm X 1cm was noted in the left supraclavicular region. Her cardiac, respiratory, and abdominal examination was normal.

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INVESTIGATIONS: Routine blood count revealed her to be anemic (Hb- 8.9g %) Rest of the blood investigations were within normal limits. Chest X-ray, ECG and 2DECHO were normal. FNAC showed extensive hemorrhagic areas and necrosis. Biopsy was suggestive of spindle cell neoplasm with areas of ulceration. Her CT report revealed Soft tissue mass in cutaneous plane of Left supraclavicular region with intense enhancement and neovascularity with preserved fat plane with significant left axillary lymph nodes suggestive of recurrence of tumor with malignant transformation.

TREATMENT: Intraoperative globular swelling measuring 7 x 5 CM was noticed located transversely in the Left middle 1/3rd of clavicle to the Left acromion process, vertically from the 2cm above the left clavicle to left 2nd rib. Wide excision of the swelling along with adjacent lymph node was done and sent for histopathological examination. The defect was closed with pectoralis major myo cutaneous flap. Postoperatively, patient was shifted to ICU and was treated with IV antibiotics and IV analgesics. Patient was discharged after suture removal on postoperative day 8. The patient came for followup every week. Her surgical wound had healed well.

HISTOPATHOLOGICAL REPORT: It was suggestive of Recurrent Dermatofibrosarcoma with sarcomatous foci with reactive lymphadenitis.

DISCUSSION: Dermatofibrosarcoma protuberans is a rare tumour, involving the skin and subcutaneous tissue with an incidence of 1 to 5 cases per million persons per year. Most cases affect the trunk and proximal extremities, rarely affecting the head and neck. Surgical excision with 3 to 5-cm-wide margins is recommended, but high rates of local recurrence (20-50%) have been noted. An unresectable or positive margin should be treated with adjuvant radiotherapy to reduce the local recurrence rate and avoid mutilation by repeated surgery. Post-operative radiotherapy has been associated with a cure rate of 85 percent. According to Balogh, Zagars GK, et al, in patients who received a combination of conservative resection and adjuvant radiation, local recurrence of 5% was noted. The majority of local recurrences occur within the first three years but recurrences after five years have been reported. Thus, it is important to follow up these patients over a long period after the treatment. A multi-modality approach is necessary for the treatment of these patients to prevent and reduce further recurrences.

REFERENCES:

1. "Tumors and tumors-like proliferations of fibrous and related tissues," in Weedon's Skin Pathology, D. Weedon, Ed., pp. 833–835, Elsevier, 2010.
2. Antonescu CR, Leung DHY, Bowne WB, et al. Dermatofibrosarcomaprotuberans: a clinicopathologic analysis of patients treated and followed at a single institution. Cancer.2000;88:2711–2720.
3. Parker TL, Zitelli JA. Surgical margins for excision of dermatofibrosarcoma protuberans. J Am AcadDermatol. 1995;32(2 Pt 1):233–236.
4. Revol M, Arnaud EJ, Perrault M, , Servant JM, Banzet P. Surgical treatment of dermatofibrosarcomaprotuberans. PlastReconstr Surg. 1997;100:884 – 895.
5. S. Bhambri, A. Desai, J. Q. Del Rosso, and N. Mobini, "Dermatofibrosarcoma protuberans: a case report and review of the literature,"Journal of Clinical and Aesthetic Dermatology, vol. 1, pp. 34–36, 2008.

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6. Gloster HM Jr. Dermatofibrosarcoma protuberans. J Am Acad Dermatol 1996; 35:355.



INTRAOPERATIVE IMAGE OF THE TUMOUR

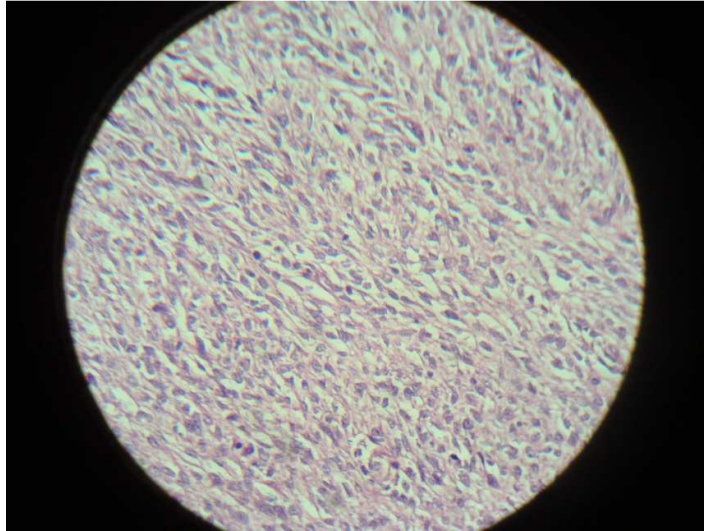


Cut section of the tumour along with the lymph node



Post operative day 8

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Spindle shaped cells, few showing atypical mitosis



Tumour showing increased vascularity