LONGSTANDING DERMATOFIBROSARCOMA PROTUBERANS- A CASE REPORT

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PRESENTATION OF CASE

A 42-year-old male presented to surgical OPD with a slow growing painless swelling in the left inguinal region since 3 years. He did not have history of similar swellings anywhere in the body. He did not have history of cough, fever, weight loss or any neurological deficits on the limb. On examination, the swelling was about 10 x 8 cms. on the left inguinal region with irregular margins, uneven surface, reddish discolouration, with no local rise of temperature and with areas of pressure necrosis.

CLINICAL DIAGNOSIS

Soft Tissue Swelling of Inguinal Region.

DIFFERENTIAL DIAGNOSIS

- Inguinal Adenitis
- Lymphoma
- Metastatic Neoplasm
- Liposarcoma
- Fibrosarcoma
- Dermatofibrosarcoma Protuberans

PATHOLOGICAL DISCUSSION

Dermatofibrosarcoma is a rare locally invasive soft tissue sarcoma with a propensity for extensive subclinical involvement local invasion which may include subcutaneous tissue, muscle, fascia and bone. The growth pattern of DFSP resembles that of fibroblasts with pseudopod like extensions from a central mass that penetrate tissue over a time.¹ Histologically, DFSP is characterised by a fibroblastic proliferation of tumour cells arranged around a central storiform pattern.² DFSP tumour cells exhibit a chromosomal translocation of genes COL1A9 and PGDF β (t17, 22) which encode to alpha chain of type I collagen and beta chain of platelet derived growth factors.³ In the last several decades, Mohs surgery has emerged as a promising treatment option and achieves superior results to surgical excision.⁴ It provides a method of eradicating tumour that rests on intraoperative evaluation of tumour margins.

Figure 1. Microscopic Appearance of DFSP

Figure 2. Clinical Appearance of The Lesion

DISCUSSION OF MANAGEMENT

He underwent a wide local excision with primary closure and the specimen was sent for HPE examination HPE examination which showed spindle cells arranged in storiform pattern with deep margin showing tumour tissue and concluded it to be dermatofibrosarcoma protuberans. The patient was referred to radiotherapy and is on follow up there.

DFSD is a low grade soft tissue sarcoma described in 1924 by Darier and Ferdinand.⁵ It accounts for 0.1% of all malignancies with an annual incidence of 0.8% million.⁶ A slight male preponderance has been reported.⁷ The tumour has lower chances of metastasis but is aggressive locally.⁸ The tumour commonly presents as a slow growing asymptomatic, skin-color, plaque that eventually turns to reddish or violaceous nodules MMS is the treatment of choice for DFSP. Prior to MMS, surgical excision with 3 cms to 5 cms wide margins was recommended but associated with high rates of local recurrence. A recurrence rate of up to 20 percent with 3cm surgical margins has been described.⁹

Radiotherapy has been used as an adjuvant therapy after wide surgical excision or in those patients who have inoperable disease. Post-operative radiotherapy has been associated with a cure rate of >85 percent.¹⁰

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FINAL DIAGNOSIS

Dermatofibrosarcoma Protuberans

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