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DERMATOFIBROSARCOMA PROTRUBERANS- A RARE CASE REPORT

General Surgery

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ABSTRACT

Dermatofibrosarcoma protruberans (DFSP) is a rare mesenchymal sarcoma with low to intermediate grade malignant potential that typically involves the dermis with an incidence rate of 0.8 to 4.5 cases per million persons per year. Typical presentation is an asymptomatic, indolent, exophytic protruberant tumour with a high local recurrance rate seen on the trunk. We report a typical case of DFSP with local recurrence on the left lower back in a 32-year old female which was managed with wide local excision with margins free of tumour infiltration confirmed by histopathology. This case is reported for its rarity.

KEYWORDS

dermatofibrosarcoma protruberans, recurrence, sarcoma, storiform

INTRODUCTION

Dermatofibrosarcoma protuberans is a rare mesenchymal tumour with an incidence rate of 0.8 to 4.5 cases per million persons per year.[1] It occurs most often in adults in 3rd to 5th decades of life.[1] It emerges as an asymptomatic slow growing tumour involving the trunk frequently.[1] Local recurrence after incomplete resection is common.[2] This case is reported for its rarity.

CASE REPORT

A 32 year old female hailing from Davangere, Karnataka, with no known comorbidities with a previous history of similar swelling excised in toto from the same site 12 years ago with no histopathological examination performed then, currently presented with a solitary exophytic lobulated tumour since 4 months with a size of 7x2cm in the left lower back, with excoriation of the skin since 2 months, nontender, no local rise of temperature, irregular well distinct borders, firm consistency throughout, nodular surface, skin not pinchable, swelling mobile in both horizontal and vertical planes, not fixed to the underlying muscles or fascia with an overlying scar of the previous surgery. Local and distant metastasis was dismissed with clinical examination and imaging.

Ultrasound imaging of the swelling suggested dermatofibrosarcoma protruberans, fine needle aspiration cytology yielded atypical cells. Tru cut biopsy was done which showed tumour cells suggestive of DFSP. Considering the history and investigations, after pre anesthetic evaluation, patient underwent wide local excision with 2cm margins. Post op period was uneventful.

Histopathology reported as cells arranged in storiform pattern and fascicles, with abundant eoisinophilic cytoplasm, elongated ovoid nucleus compatible with dermatofibrosarcoma protruberans with margins free of tumour infiltration. Immunohistochemistry showed tumour cells strongly positive for CD 34 immunostaining and factor XIIIa negative staining compatible with DFSP.

She was followed up on out patient basis and showed no signs of local recurrence or metastasis. Hence adjuvant radiotherapy was not considered during her follow up.

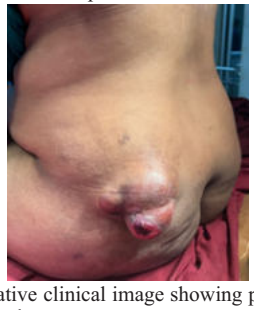

Figure 1: Pre operative clinical image showing protruberant tumour over the left lower back.

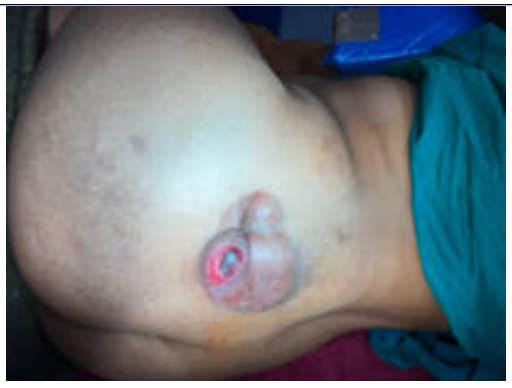

Figure 2: Pre operative clinical image with the scar of the previous surgery at the site of recurrance

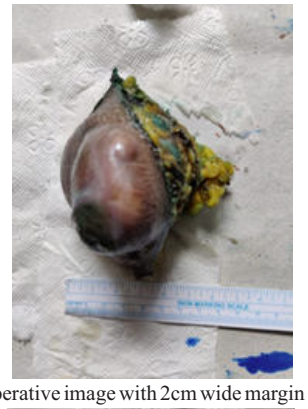

Figure 3: Post operative image with 2cm wide margin

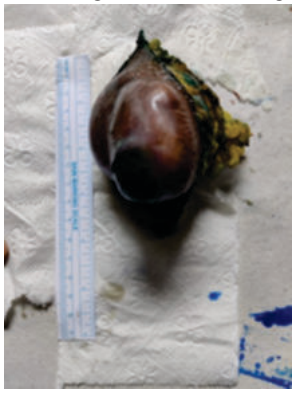

Figure 4: Post operative image with 2cm wide margin

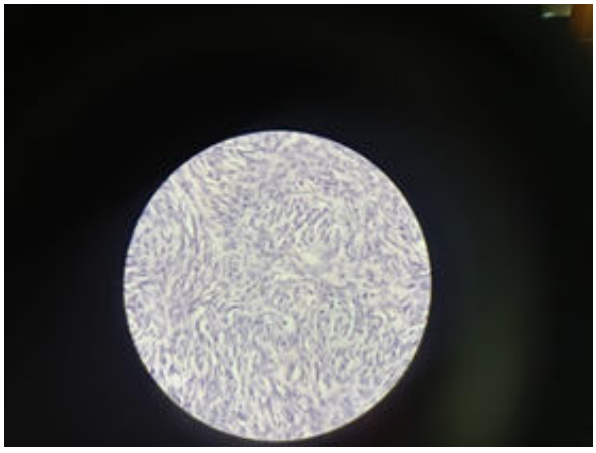

Figure 5: Histopathological image showing storiform pattern of tumour cells

DISCUSSION

DFSP is a rare, locally aggressive indolent dermal mesenchymal neoplasm. Clinically the typical presentation is asymptomatic, slow growing tumour most commonly on the trunk (50%),[1] which are asymmetric, protruberant tumour with root like projections over the years involving the dermis.[2] The cause is unknown, though proposed risk factor includes injury to skin in affected location,[2] like within preexisting scars or tattoos. Pathophysiology shows chromosomal translocation between t(17;22)(q22;q13) which is present in over 90% cases which leads to fusion of platelet derived growth factor- beta polypeptide gene (PDGFB) and collagen type 1A1 gene (COL1A1).[1] A thorough history, physical examination and lymph node examination is recommended. Diagnosis is made via skin biopsy. It is considered an intermediate-grade malignancy with high local recurrence rate.[1] Distant metastasis is rare in only <5% of cases.[2] The standard treatment consists of either Mohs micrographic surgery[1] or wide local excision with 2-4 cm margins.[2] Biopsy typically shows the lesion primarily in the dermis with irregular infiltration into subcutaneous plane in lace-like storiform pattern. Epidermis is usually spared.[1] Immunohistochemistry shows spindle cells strongly positive for CD 34 immunostaining and factor XIIIa negative.[4] The factors associated with high rates of recurrence are histological subtype, cellularity, size, location and high mitotic rate.[2] Radiotherapy has been used as an adjuvant therapy after wide surgical excision or in those with inoperable macroscopic disease. Post operative radiotherapy is associated with a cure rate of >85%.[5] Risks of adjuvant radiotherapy include acute and chronic radiodermatitis and further development of new skin cancers.[1]

CONCLUSION

This case motivated us to present an interesting typical case of dermatofibrosarcoma protruberance with local recurrence. As local recurrence is common even after wide excision, which is highest in the first 3 years of treatment,[1] follow up is necessary every 3 to 6 months for first 3 years, and then annually thereafter.[3] The key is to remove the entire lesion with clear margins. The overall prognosis is good with 10-year survival rate of 99.1% as metastasis is rare.[1] Morbidity is due to local recurrence, complications like post surgical scarring and cosmesis.

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