ORIGINAL RESEARCH PAPER

General Surgery

DERMATOFIBROSARCOMA PROTUBERANS -A RARE CUTANEOUS TUMOUR PRESENTING AS BENIGN LIPOMA – CASE REPORT

KEY WORDS:
Dermatofibrosarcoma protuberans, revision surgery,local excision , recurrence.

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ABSTRACT

Background: Dermatofibrosarcoma protuberans is a rare cutaneous tumor occurs in less than 0.1 % of all tumors.Often we misdiagnose it as a lipoma. It is treated by wide local excision. **Description :** A 45 yr old female present with swelling in back on left side for 3months. Preoperative diagnosis lipoma and underwent local excision. Pathological report turned out to be dermatofibrosarcoma protuberans.Margins (2sides) found to be positive. Planned for revision surgery.Patient under follow up. **Conclusion:** Dermatofibrosarcoma protuberans is a rare cutaneous sarcoma . It originates in dermis tends to invaginate nearby muscle, tendons, fascias.In our case tumor is attached to muscle. Revision surgery planned .Long term follow up is needed to prevent recurrence.

INTRODUCTION:

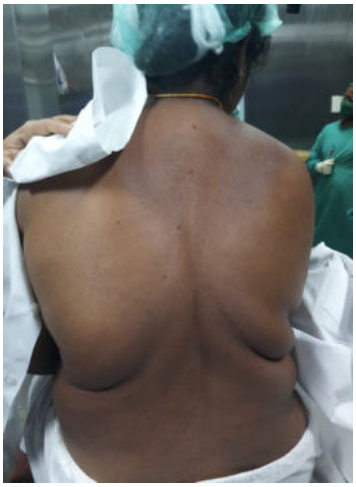
Dermatofibrosarcoma protuberans is a rare slow growing soft tissue sarcoma [1] with fibroblastic origin with high rates of local recurrence . DFSP is most commonly seen on trunk and extremities followed by head and neck [2]. Because of its unspecific appearance and slow growth it is often misdiagnosed as benign lesions. DFSP is treated by wide local excision followed by adjuvant radiotherapy and chemotherapy. Here in this case we initially misdiagnosed as lipoma and underwent local excision .This was presented to being more awareness to benign nature of this rare malignant tumour.

CASE REPORT:

A 45 yr old female came with the complaints of swelling in left back for 2months , insidious onset ,not progressive not associated with pain, restriction of movements,pt not having fever,no swelling elsewhere . Patient known case of rheumatic heart disease on treatment.no history malignancy in family.

General examination were normal ; local examination revealed 4*4 cm Mass in the left infrascapular region ,smooth surface,no dilated veins,skin appears normal,not warm non tender soft in consistency.No regional lymphoadenopathy.

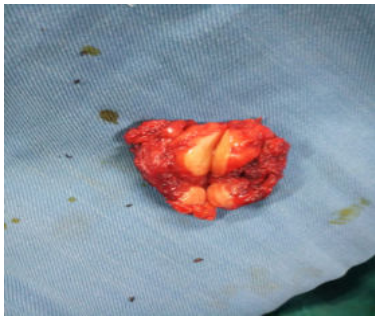
PREOPERATIVE PICTURE OF PATIENT:



High frequency ultrasound revealed lipoma ; FNAC was performed which showed features of folliculitis or abscess ;With these investigation we came to conclusions of lipoma and proceeded with excision biopsy of mass.

Intraoperatively mass found adherent to surrounding structures and excision of tumour done . postoperative period were uneventful, wound healed well. Patient discharged on 4th post operative day. On follow up Histopathological examination revealed DERMATOFIBROSARCOMA PROTUBERANS with immunohistochemistry marker CD 34 +. Patient referred to radiation oncology department for adjuvant radiotherapy.

EXCISED SPECIMEN:

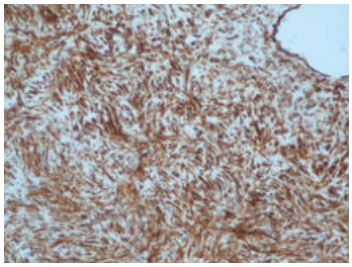


DISCUSSION:

DFSP is an uncommon soft tissue sarcoma which affects 1 in 4.2 million patient.Men and women are equally affected .Incidence was between 40 and 70 years.site of occurance is trunk ,lower extremity followed by head and neck. On physical examination these are firm,indurated nodules which are reddish or brown in appearance.

On histological evaluation DFSP is a dermal or subdermal tumour without penetration into epidermis.Cytogenetically,it displays translocation of **t(17;22)(q22;q13)**which fuses **COL1A1** and platelet derived growth factor **PDGF.** This can be detected by FISH ;markers are **CD34** and **VIMENTIN.**

Immunohistochemistry of the tumour cells showing positivity forVIMENTIN.



Immunohistochemistry of the tumour cells showing positivity for CD 34 antigen.

It can be classified as low grade – favourable prognosis after surgical resection.

High grade – aggressive tumour with metastatic potential. Despite high recurrence rate, metastatic disease occurs in 1-2% . Mortality is rare They have excellent outcome.

Ultrasound shows mostly hypoechoic or mixed hyperechoic, with mostly well defined margins to irregular margins showing psedopodia[3] . Lipoma also has same feature in ultrasound so distinction between two is not possible[4].

MRI studies also not specific[5].Histological examination is the only definitive diagnostic method.

Other types of DFSP are myxoid DFSP and bednar DFSP [6]. Immunohistochemically , tumour stain for VIMENTIN,CD 34,apolipoprotein D,nestin [7,8].

Treatment of choice is wide local excision with margin of 3-5cm from tumour edge. The rate of recurrence depends on the resection margins. Reconstructive surgery may be required to restore tissue defects after excision using local skin flap,skin graft or myocutaneous flap[9].

Alternative to wide surgical resection is Mohs micrographic surgery[10,11].Local cure rates of 93-100% [12].

Adjuvant chemotherapy using imatinib mesylaye, a tyrosine kinase inhibitor,is used in treatment of unresectable , recurrent or metastatic disease[13].

Combination of conservative excision and adjuvant radiotherapy has demonstrated reduced local recurrance of 5%

CONCLUSION:

DFSP is a rare malignant tumour with presentation similar to lipoma ,so one must have high index of suspicion and rule out DFSP before excision of benign tumour.Wide local excision is the treatment of choice .Though there is high local recurrance ,survival is excellent .

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