



# 28th Annual Conference of the Dermatopathology Society of India

## DERMATOFIBROSARCOMA PROTUBERANS - a case series

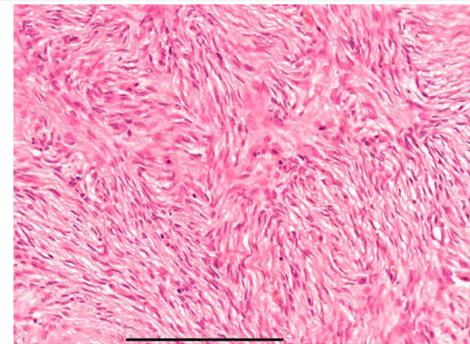
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### INTRODUCTION

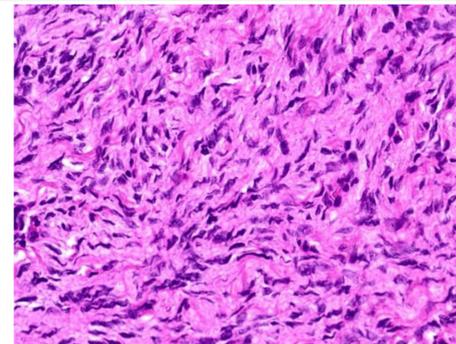
- ✓ Dermatofibrosarcoma Protuberans (DFSP) is a locally invasive tumor of low malignant potential & high rate of recurrence.
- ✓ It constitutes 1-6% of sarcomas and 18% of all cutaneous soft tissue sarcomas.

We present a series of 4 cases of Dermatofibrosarcoma Protuberans -

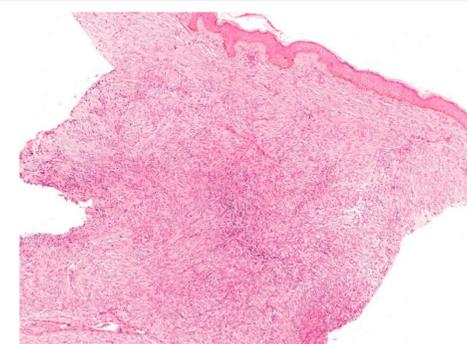
### HISTOPATHOLOGICAL IMAGES



(A) Tumor cells arranged in Storiform Pattern



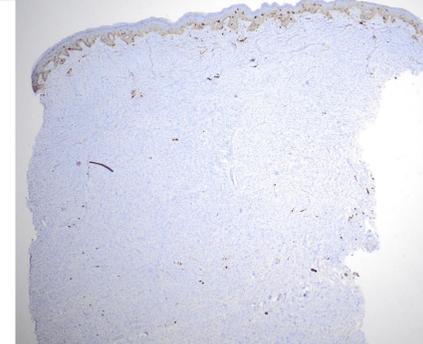
(B) Elongated spindle shaped tumour cells with nuclear pleomorphism



(C) Tumor extending into subcutaneous tissue



(D) CD 34(+) seen on IHC {seen in all 4 cases}



(E) S100 (-) seen on IHC

### CASE SERIES

- A 40-year-old female presented with a 1 year history of firm, protuberant nodules and hard indurated plaque, of size 8X5cm over the left inner thigh, which was associated with pain for the past 3 months.
- Histopathology(HPE) showed a diffuse tumour of round to spindle cells in oedematous, myxoid stroma with dilated vascular spaces.

#### CASE - 1



- A 47-year-old male presented with a lesion over the left groin since 4 years, with accelerated growth for past 10 days.
- On examination 3 soft nodules surmounted a hyper-pigmented indurated plaque of 8X10cm over the left groin, extending into the lower abdomen.
- HPE showed a dermal neoplasm of spindle cells in a storiform pattern, with nuclear pleomorphism and mitotic activity.

#### CASE - 2



- A 56-year-old male came with a lesion over the right lower abdomen since 2 years.
- On examination 2 firm, flesh coloured nodules were seen over a hard indurated base.
- HPE showed a tumour of spindle cells in storiform pattern with mitotic figures.

#### CASE- 3



- A 38-year-old male presented with a mildly pruritic, raised lesion over chest for past five years.
- On examination, a 10 × 8 cm, well-defined plaque, showing an irregular nodular surface and areas of hyperpigmentation, was noted over the left chest wall.
- HPE revealed spindle cells arranged in fascicles and foci with a storiform pattern in dermis.

#### CASE- 4



### DISCUSSION

- DFSP is seen in 3<sup>rd</sup> to 5<sup>th</sup> decade of life and affects males and females equally. Most often occurs on the trunk, proximal extremities & rarely over the head & neck.
- DFSP is diagnosed by typical pathology of spindle cells in storiform pattern, with pleomorphism and CD34 positivity on IHC.
- Surgical excision is the treatment of choice, however, recurrence after excision is seen in 25% cases, with a 5% risk of high grade sarcomatous transformation. Mohs Micrographic Surgery is the preferred treatment.
- Post Surgical Radiotherapy & Imatinib Mesylate have also been employed.

### CONCLUSION

- DFSP is not an uncommon cutaneous soft tissue sarcoma.
- The clinical resemblance to keloid leads to delay in diagnosis.
- Histopathological identification with IHC marker is the basis of diagnosis.
- Early diagnosis may be the key to complete surgical treatment with lesser recurrences.
- Surgical excision is treatment of choice.

### REFERENCES

- 1.Lim SX, Ramaiya A, Levell NJ, Venables ZC: Review of dermatofibrosarcoma protuberans. Clin Exp Dermatol. 2023, 48:297-302. 10.1093/ced/llac111
2. Hao X, Billings SD, Wu F, Stultz TW, Procop GW, Mirkin G, Vidimos AT: Dermatofibrosarcoma protuberans: update on the diagnosis and treatment. J Clin Med. 2020, 9:1752. 10.3390/jcm9061752