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Dermatofibrosarcoma Protuberans - Cutting through the Complexity

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Abstract: ***Dermatofibrosarcoma protuberans (DFSP) is an uncommon low-grade malignant soft tissue sarcoma, representing less than 0.1% of all malignant neoplasms and 1% of soft tissue sarcomas. It generally manifests in the third and fourth decades of life, with a greater prevalence in males. DFSP originates from dermal fibroblasts and is distinguished by gradual, infiltrative proliferation, local recurrence, and a minimal propensity for metastasis. This case report describes a 17-year-old female with a two-year history of a rapidly growing nodular mass on the anterior chest wall. Diagnostic assessments, comprising MRI, FNAC, and histology, validated the diagnosis. The patient had a broad local excision with negative margins, and immunohistochemistry indicated CD34 positivity. The postoperative follow-up indicated no recurrence. Timely identification, interdisciplinary care, and prolonged surveillance are crucial to enhance outcomes and reduce recurrence risk in DFSP cases.***

Keywords: Dermatofibrosarcoma protuberans, Histopathology, local recurrence, Rare Neoplasms, Soft tissue sarcoma.

1. Introduction

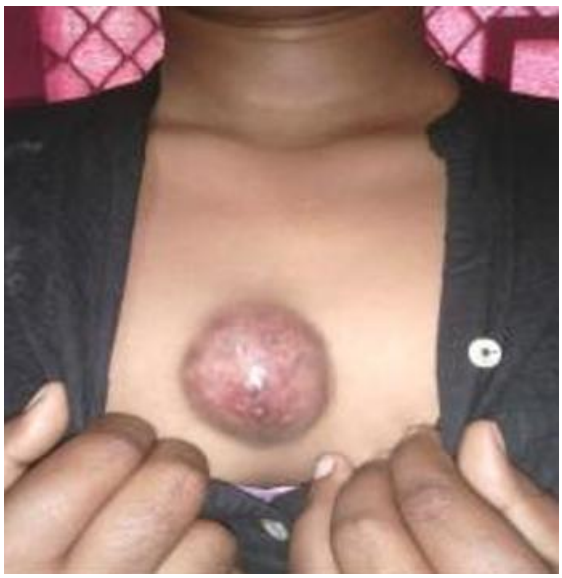
- Less than 0.1% of all malignant neoplasm.1% of STS.
- M: F =4:1 and in 3rd and 4th decade.
- Uncommon, low grade sarcoma, incidence rate of 4.1 to 4.5 cases/ million /year
- Characterized by infiltrative growth and local recurrence.
- Orginates from dermal fibroblasts.
- Low to intermediate grade. Rarely metastasizes (lungs and regional lymphnodes).
- Rare locally aggressive malignant cutaneous soft-tissue sarcoma.
- Develops in the connective tissue cells in the middle layer of the skin.
- 40%arise on the trunk, with most of the remaining tumors distributed between the head and neck and extremities.

2. Case Presentation

- 17 years old female presented with swelling in anterior chest wall for past 2 years gradually increasing in size and not associated with pain or bleeding.
- O/E: - 5*4 cm firm swelling over mid chest wall, nodular, surrounding skin telangiectasia with central ulceration and well defined borders. Nodule fixed to dermis but moves freely over deeper tissues.
- The lesion presents as a protuberant nodular, cutaneous mass that shows slow and persistent growth

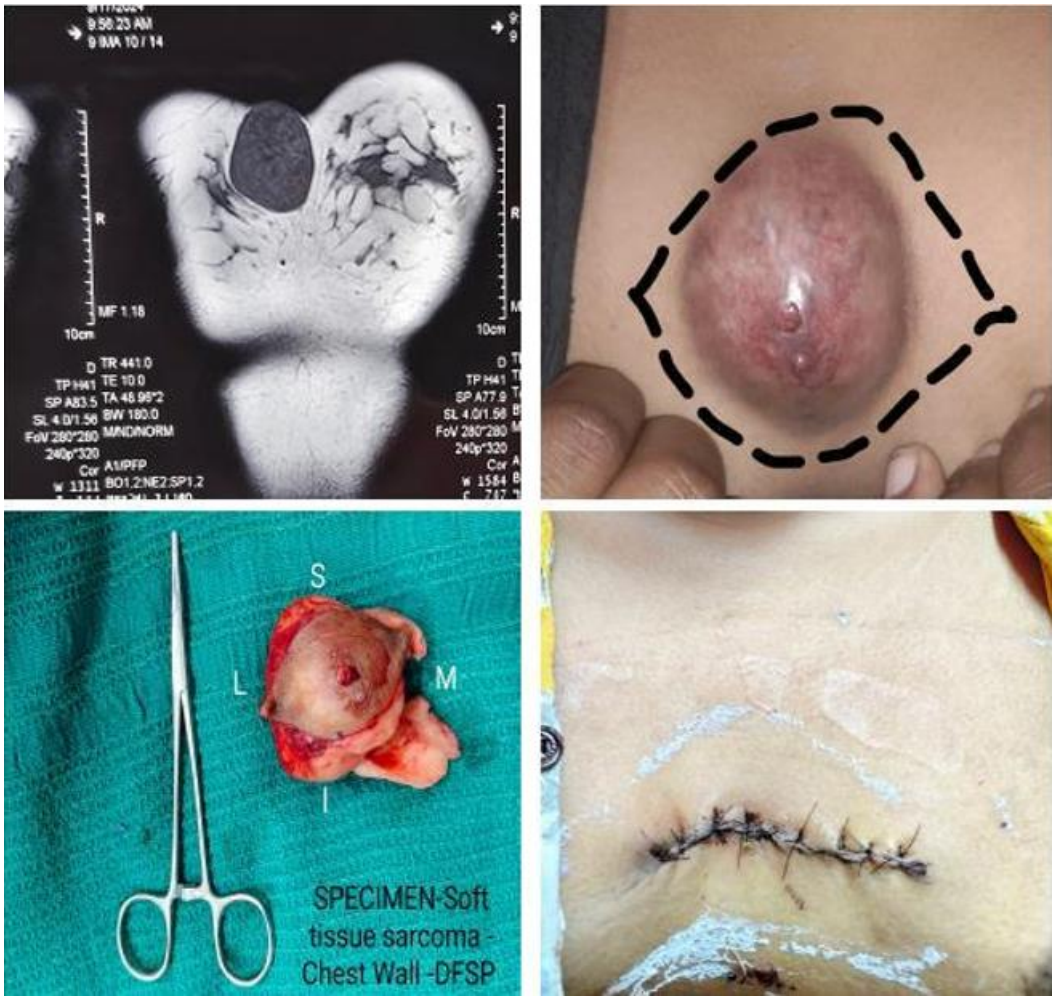
3. Diagnostic Approach

- CT chest: - Soft tissue sarcoma in subcutaneous plane.
- FNAC: - spindle cell neoplasm
- MRI: - well defined T1 isohyperintense and T2 hyperintense lesion showing diffusion restriction, low ADC values, involving skin and subcutaneous plane possibility of aggressive fibroma.
- Incisional biopsy features of soft tissue sarcoma - spindle cell neoplasm consistent with dermatofibroma.



Management

- Surgical: - wide local excision with 3 cm margin clearance.
- HPE: - stratified squamous epithelium with underlying neoplasm arranged in storiform pattern, short fascicles, cell are elongated to ovoid with vesicular ovoid to spindle shaped nucleus.no necrosis identified. All margins are free from tumor.
- IHC: - CD 34 - positive, SMA: - negative
- STAGING: - T2a N0M0



4. Discussion

- Dermatofibrosarcoma protuberans-neoplasm arising in dermis that may occur anywhere in the body.
- Recurrence rate -10 to 60%.
- NCCN recommendation- Debulking specimens from all excisions should be examined to identify fibrosarcomatous transformation of DFSP.
- Radiation - positive margins if further reresection not possible.
- Genetic analysis- COL1A1-PDGFB fusion gene + and responds to Imitinib.

NCCN Guidelines: -

- Follow up - every 6 to 12 months for 3 to 5 years.
- Physical examination and MRI surveillance for deeply invasive disease.

5. Conclusion

- This is a rare low grade soft tissue sarcoma in an adolescent female age, after multidisciplinary approach we achieved successful excision with no complications. Follow up of patient showed no signs of recurrence.
- High suspicion, early detection, wide excision.
- Long term follow up is necessary to detect recurrence. wide excision of recurrent tumor is necessary with adjuvant radiotherapy.

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