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Dermatofibrosarcoma protuberans

By Kristina M. Lim, DO, Emily Chea McEldrew, DO, and Cynthia L. Bartus, MD, FAAD, FACMS

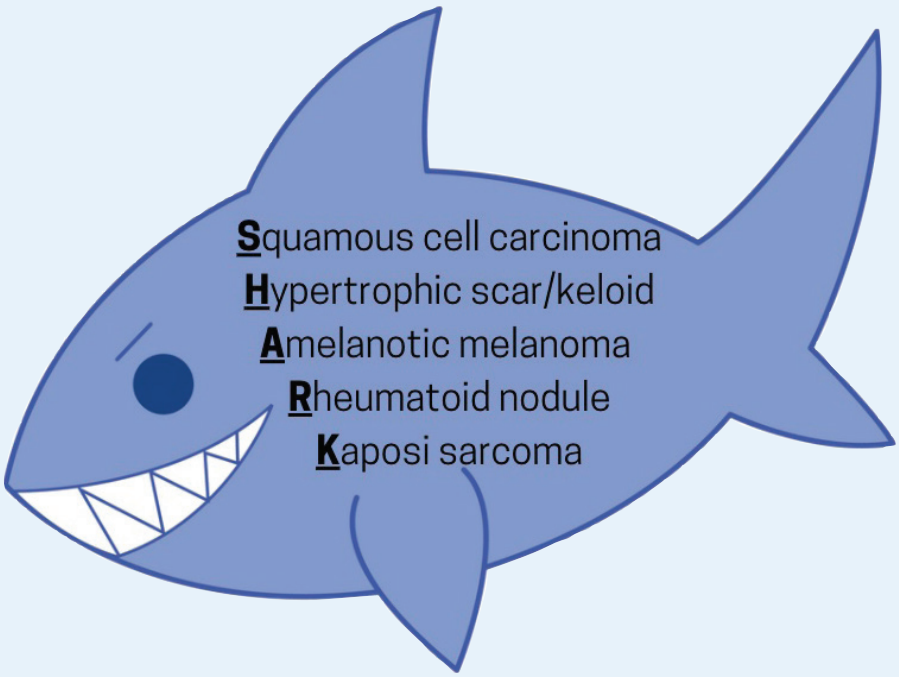
What	Rare soft tissue tumor of intermediate malignancy (low rate of metastasis, high rate of local recurrence) involving dermis, subcutaneous fat, occasionally muscle and fascia
Who/when	Young- to middle-age adults No sex predilection
Why	Translocation between chromosomes 17 and 22 → fusion of COL1A1-PDGFB (90% of cases)
Where	Shoulder or pelvic region are most common Trunk (50-60%) Proximal extremities (20-30%) Head and neck (10-15%)

Clinical features

- First appears slow-growing and asymptomatic
- Evolves into large, red-brown, indurated plaques or nodules that feel firmly attached to subcutaneous tissue
- May also grow rapidly during pregnancy

Clinical ddx

SHARK - squamous cell carcinoma, hypertrophic scar/keloid, amelanotic melanoma, rheumatoid nodule, Kaposi sarcoma



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Histologic features

Monotonous spindle-shaped cells arranged in a storiform or "herringbone" pattern. Cells obliterate adnexal structures and infiltrate the subcutaneous tissue in a "honeycomb" pattern.

\+ Stains:
- CD34

\- Stains:
- Factor XIII
- Stromelysin-3
- D2-40

Histologic ddx

Plaque stage: atrophic dermatofibroma, dermatomyofibroma, fibroblastic connective tissue nevus, and neurofibroma

Nodular stage: deep dermatofibroma, fibrosarcoma, and malignant peripheral nerve sheath tumor

Management

Surgical
- Mohs surgery (98-100% cure rate). Treatment of choice.
- Wide local excision (WLE): 2-4 cm margins extending to superficial muscular fascia. (93% cure rate)

Radiation
- For unresectable or recurrent tumors or postoperatively for positive surgical margins. (86-93% cure rate)

Systemic medications
- Imatinib - oral tyrosine kinase inhibitor for unresectable, recurrent, or metastatic DFSP in adults. Evaluate for a t(17:22) translocation prior to therapy. (65% response rate)

Metastasis: Lungs are the most common site.
- Multidisciplinary consultation for possible further treatment is recommended for tumors with fibrosarcomatous degeneration, due to its increased metastatic potential.

Recurrence: Variable depending on treatment; most likely to occur within three years.

Follow-up: Clinical follow-up q3-6 months for first three years, annually thereafter.

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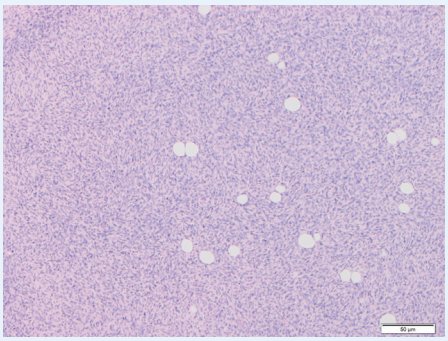
Additional histologic features

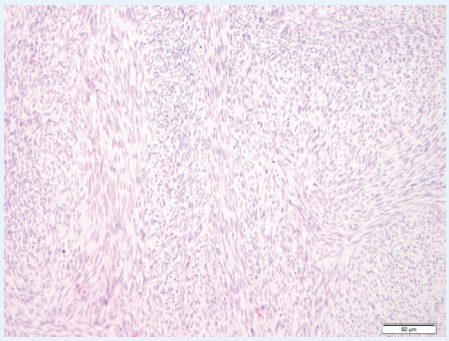
5% of DFSPs contain melanin and are referred to as Bednar tumors.

Fibrosarcomatous degeneration is considered a form of tumor progression, histopathologically demonstrated by a change from storiform to herringbone pattern with increased mitotic activity, cellularity, atypia, and often loss of CD34 staining. It is associated with a greater metastatic risk (15-20%) compared to classic DFSPs.

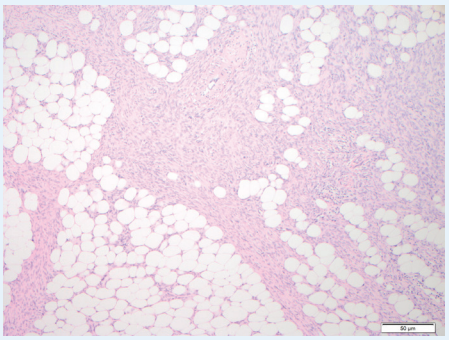
Punch or incisional biopsy, preferably of deeper subcutaneous layer for sufficient tissue sampling and accurate pathologic assessment.



Storiform pattern



Herringbone pattern



"Honeycomb" pattern

References:

1. Bolognia JL, Callen JP, Jorizzo JL, Schaffer JV. *Dermatology.* 4th ed. Elsevier; 2018.
2. Elston DM and Ferringer T. *Dermatopathology.* 3rd ed. Elsevier; 2019.
3. Alikhan A and Hocker TLH. *Review of Dermatology.* Edinburgh: Elsevier; 2017.
4. Patterson J. *Weedon's Skin Pathology.* 5th ed. Elsevier; 2021.
5. *NCCN Clinical Practice Guidelines in Oncology.* Dermatofibrosarcoma protuberans. Version 1.2022-Nov 17, 2021.
6. Hao X, Billings SD, Wu F, et al. Dermatofibrosarcoma Protuberans: Update on the Diagnosis and Treatment. *J Clin Med.* 2020;9(6):1752.
7. Brooks J, Ramsey ML. Dermatofibrosarcoma Protuberans. [Updated 2022 Jan 21]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan. Available from: https://www.ncbi.nlm.nih.gov/books/NBK513305/.
8. JJ Abbott, AM Oliveira, AG Nascimento. The prognostic significance of fibrosarcomatous transformation in dermatofibrosarcoma protuberans. *Am J Surg Pathol.* 2006;30(4):436-443.
9. Histology slides courtesy of Marisa Baldassano, MD.