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.

References:
9. Histology slides courtesy of Marisa Baldassano, MD.