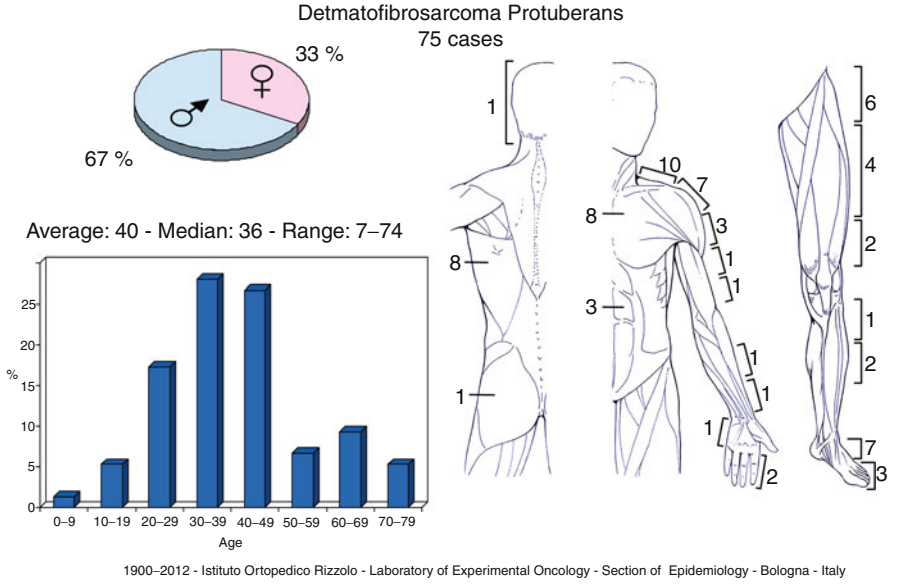
Chapter 64
Dermatofibrosarcoma Protuberans

Marco Gambarotti

Definition: Low-grade fibroblastic tumor of the skin in adult patients. There is a juvenile form of DFSP, called giant-cell fibroblastoma.
Epidemiology: Infrequent, males. DFSP: young to middle-aged adults, rare in children. Giant-cell fibroblastoma: infants and children younger than 5 years, exceptional in adults.



M. Gambarotti, MD
Department of Anatomy and Pathological Histology, Istituto Ortopedico Rizzoli,
Via del Barbiano 1/10, Bologna 40136, Italy
e-mail: marco.gambarotti@ior.it

P. Picci et al. (eds.), *Atlas of Musculoskeletal Tumors and Tumorlike Lesions*,
DOI 10.1007/978-3-319-01748-8_64, © Springer International Publishing Switzerland 2014

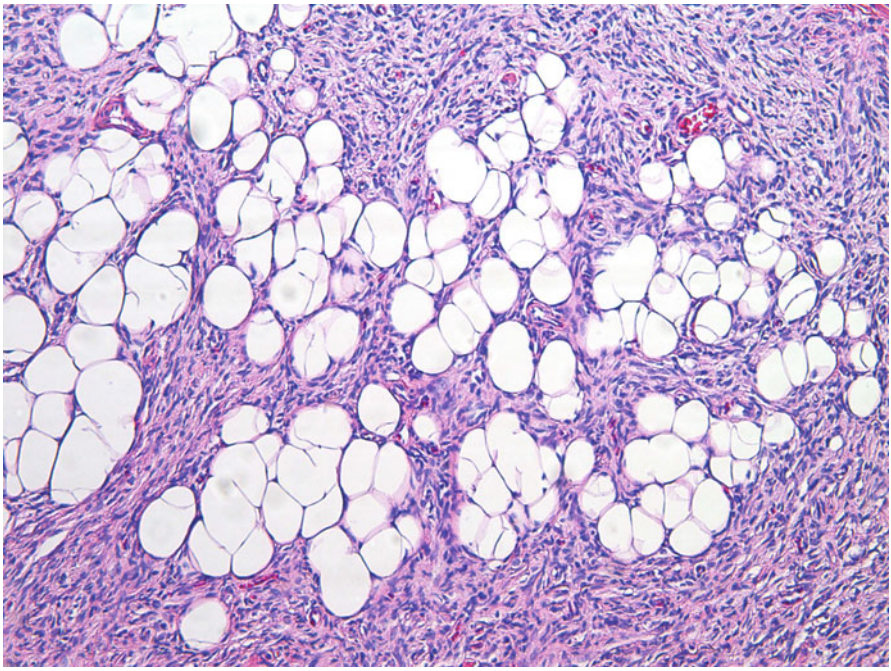
Location: Trunk, groin, and proximal extremities.
Clinical: Nodular or plaque-like painless cutaneous tumor, slowly growing.
Diagnosis: A well-defined superficial tumor. On MRI, in T1 usually iso- or hypointense to skeletal muscle. In T2 intermediate or high signal intensity compared to fat. On STIR high signal, similar to water or blood vessels. Uniform enhancement of gadolinium.
Histopathology: DFSP: Monotonous spindle cell proliferation, diffusely infiltrating the dermis, subcutis, or both, spindle cells with elongated wavy nuclei. Cells are organized in a storiform pattern, there is minimal cellular atypia, and mitoses are rare. Characteristically there is infiltration of the subcutaneous fat in a honeycomb pattern. Giant-cell fibroblastoma: spindle to pleomorphic cells, variably collagenized matrix, multinucleated-appearing giant cells bordering pseudovascular spaces. Mitoses are rare and necrosis is absent. Fibrosarcomatous DFSP is defined by herringbone pattern and increased atypia and mitoses. On immunohistochemistry DFSP are positive for CD34. Apolipoprotein A1 has also been reported as a sensitive marker of DFSP. Giant-cell fibroblastoma: positive for CD34 and negative for S-100, CD31, and epithelial markers. DFSP and giant-cell fibroblastoma share similar molecular abnormalities: the presence of supernumerary ring chromosomes consisting of amplified sequences from chromosomes 17 and 22 and/or the presence of t(17;22), a balanced reciprocal translocation that results in the fusion of COL1A1, a gene of collagen, and PDGFβ, a gene that encodes a growth factor. Ring chromosomes are predominantly observed in DFSP of adult patients, whereas the t(17;22) translocation is mostly seen in DFSP of children and in giant-cell fibroblastoma.
Course and Staging: Conventional DFSP recurs locally in 10–50 % of cases, often after incomplete excisions. Distant metastases are observed in less than 5 % of cases, almost all of which are associated with high-grade fibrosarcomatous changes. Giant-cell fibroblastoma recurs in up to 50 % but does not metastasize. Recurrence rates are closely related to surgical margins.
Treatment: Wide excision with tumor-free margins is curative. Radiotherapy has been proposed for unresectable tumors or after margin-positive resections. Imatinib mesylate (Gleevec), a tyrosine kinase inhibitor, may have potential value in the treatment of recurrent or metastatic DFSP.

Chromosomal Translocations

t(17;22) (q22;q13)	COL1A1-PDGFβ	>90 %
ring 17q, ring 22q, der(22)	COL1A1-PDGFβ	75 %

Immunohistochemical Panel

Vimentin	+
CD34	+



Bland-appearing spindle cells organized in a monotonous storiform pattern, with infiltrative margins that frequently surround lobules of fat

Selected Bibliography

Archontaki M, Korkolis DP, Arnogiannaki N, Konstantinidou C, Georgopoulos S, Dendrinos P, Zarkadas G, Kokkalis G (2010) Dermatofibrosarcoma protuberans: a case series of 16 patients treated in a single institution with literature review. Anticancer Res 30(9):3775–3779. Review

Gooskens SL, Oranje AP, van Adrichem LN, de Waard-van der Spek FB, den Hollander JC, van de Ven CP, van den Heuvel-Eibrink MM (2010) Imatinib mesylate for children with dermatofibrosarcoma protuberans (DFSP). Pediatr Blood Cancer 55(2):369–373. Review

McArthur G (2007) Dermatofibrosarcoma protuberans: recent clinical progress. Ann Surg Oncol 14(10):2876–2886. Review

Palmerini E, Gambarotti M, Staals EL, Zanella L, Sieberova G, Longhi A, Cesari M, Bonarelli S, Picci P, Ruggieri P, Alberghini M, Ferrari S (2012) Fibrosarcomatous changes and expression of CD34+ and apolipoprotein-D in dermatofibrosarcoma protuberans. Clin Sarcoma Res 2(1):4. doi:10.1186/2045-3329-2-4

Stadler FJ, Scott GA, Brown MD (1998) Malignant fibrous tumors. Semin Cutan Med Surg 17(2):141–152. Review

Sundram UN (2009) Review: Dermatofibrosarcoma protuberans: histologic approach and updated treatment recommendations. Clin Adv Hematol Oncol 7(6):406–408. Review