

**Dermatofibrosarcoma: a rare form of soft tissue. Management and review of the literature**

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**Abstract**

Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue tumor of cutaneous origin of intermediate grade malignant potential. The incidence of DFSP is 0.1% of all cancers and 1% of all soft tissue sarcomas. We present the case of a 65years old female with a palpable, painful mass on the right thigh. A surgical excision of the lesion was done and the histopathology, as well as the immunohistochemical analysis with CD-34, confirmed the diagnosis of DFSP. Two years later, the patient is free of disease and no local recurrences or metastases have been found. Wide radical excision is the preferred surgical method for therapy of DFSP without distant metastasis. Furthermore, DFSP resists to conventional chemotherapy and radiation therapy, while, in cases of metastasis, therapy depends on cytogenesis and molecular biology of the tumor, so new therapeutic strategies are under research.

Key words: Dermatofibrosarcoma protuberans (DFSP), soft tissue, tumor, translocation COL1A1-PDGFB, CD-34

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