

Desmoplastic fibroblastoma of the thigh

A case report



Ann. Ital. Chir., 2011 82: 225-228

Michelangelo Miccini, Ottavia Borghese, Diletta Cassini, Matteo Gregori, Adriano Tocchi

Department of Surgery, "Sapienza" University Medical School, Rome, Italy

Desmoplastic fibroblastoma of the thigh. A case report

Desmoplastic fibroblastoma (DF) is an extremely rare benign soft tissue tumor, prevalent in adult men, mostly arising in deep regions of extremities. The tumor presents with a slowly growing and no recurrence or metastases after surgical excision. Histologically, DF is characterized by a collagenous stroma that contains spindle- and stellated-shaped fibroblastic cells positive for vimentin. Differential diagnosis with locally aggressive soft tissue tumors could be difficult. This case report deals with the clinical, pathological and immunoistochemical features of a DF of the left thigh in a 63-years old man.

KEY WORDS: Desmoplastic fibroblastoma; Soft tissue tumor.

Introduction

Desmoplastic fibroblastoma (DF) is a rare benign fibroblastic soft tissue tumor.¹

The tumour is prevalent in adult men (4:1), aged 50 to 60 years, with wide anatomic distribution, and typically it arises in the subcutaneous or subfascial tissue, mainly, of skeletal muscles. The tumor behaves in a benign fashion, presents a slowly growing with no recurrence or metastases, it can achieve a large volume².

Gross pathology generally shows an elongated, lobulated mass with a firm consistency and a pearl-gray colour. Histologically, DF is characterized by a collagenous stroma that contains spindle- and stellated-shaped fibroblastic cells³.

This case report deals with the clinical, pathological and immunoistochemical features of a DF of the left thigh.

Case report

A 63-years old man was referred to our Institution because of a painless swelling of the left thigh slowly growing since six months. No history of significant antecedent trauma, no pain, and no functional or vascular disorders of the lower limb were associated. At physical examination an irregularly round, mass was observed on the anterior aspect of the left thigh. A solid, pasty mass mobile in deep sections of the subcutis was palpated deeply seated in the quadriceps muscle. No cutaneous dischromic alterations were observed and no pulsations were detected.

An ultrasonographic examination revealed a hyperechoic mass (14x10x3 cm) located in the subfascial plane of the anterolateral side of the left thigh. A contrasted MRI scan evidenced a well-delineated, hyperintense (T1 and T2 phases) soft-tissue mass adjacent to the vastus lateralis of quadriceps muscle (Fig. 1). The lesion was characterized by smooth surface, regular margins and well-

Pervenuto in Redazione Agosto 2010. Accettato per la pubblicazione Ottobre 2010

Correspondence to: Adriano Tocchi, MD, FACS, Via Bruno Bruni 94, 00189 Rome, Italy (e-mail: adriano.tocchi@uniroma1.it).

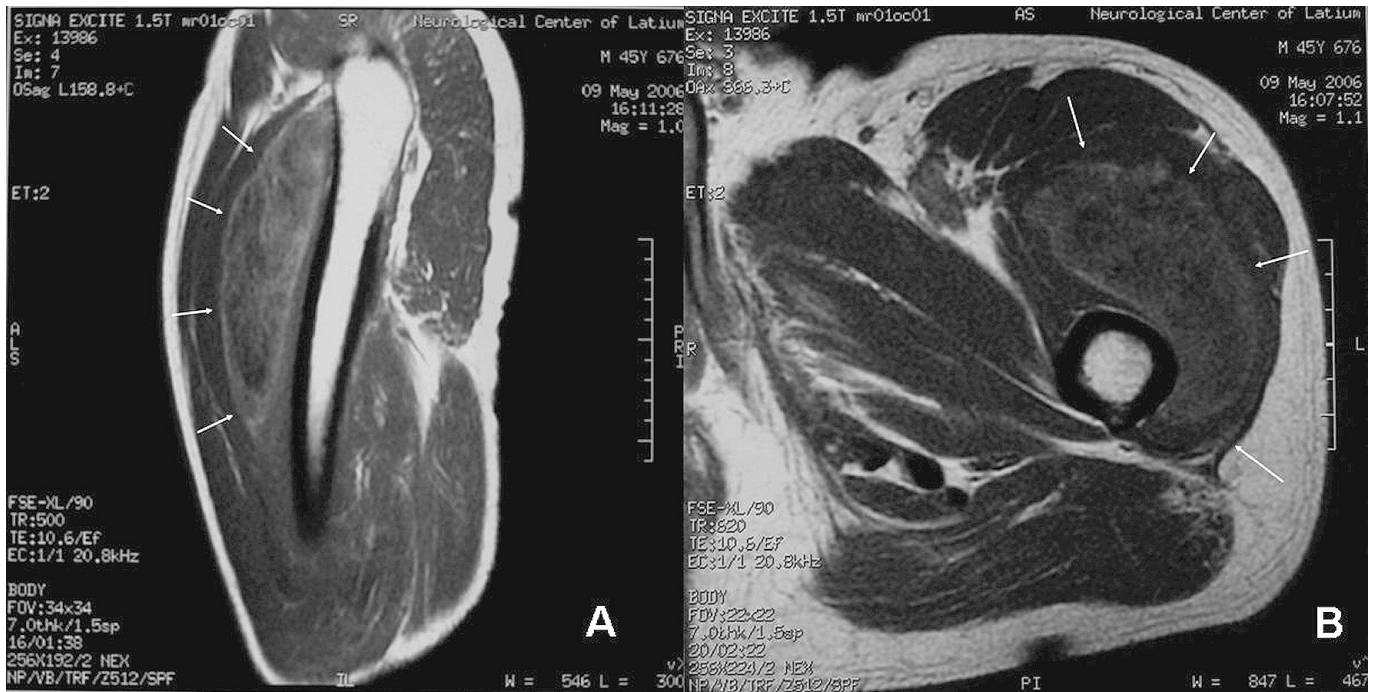


Fig. 1: Left thigh MRI: sagittal (A) and transverse (B) T1 images of the soft-tissue mass (arrows).



Fig. 2: Surgical exposure of mass deeply located under the rectus femoris.

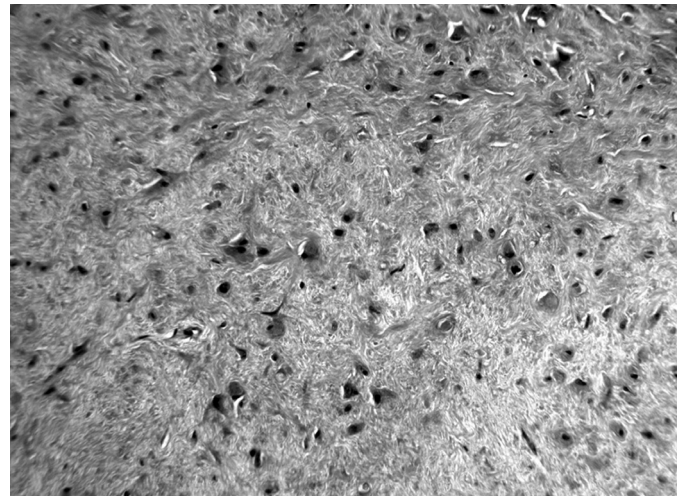


Fig. 3: Histological picture of desmoplastic fibroblastoma showing low cellularity, variable dense collagenous stroma and stellate or "reactive" fibroblasts. (H&E, 100x).

defined capsule. No evidence of calcification and continuity with surrounding (vascular and nervous) structures was observed. Complete surgical excision of the neoplasm respectful of the muscles, nerves and great vessels was performed (Fig. 2). A drainage system, located in remnant space, was removed on 8th postoperative day. *Pathology.* Macroscopically the neoplasm appeared as a well circumscribed firm discoid mass with slightly irregular external surface and homogeneous whitish colour. On cut section, the specimen showed fascicular pattern and light grey to tan colour. Histopathological exami-

nation revealed a low to moderate cellular lesion, made of spindle and stellate "reactive" fibroblastic cells with plumped nuclei in a variably dense collagenous stroma (Fig. 3). No necrotic foci or mitotic figures were observed. Immunohistochemistry analysis of tumour cells showed positive results for vimentin but negative for smooth muscle actin, desmin and CD34. The diagnosis of DF was defined on the base of these findings.

Follow up. Physical examination and MRI were performed at 6,12 and 24 months after surgery with no evidence of recurrence or metastases.

Discussion

First described in the 1995 from Evans, the DF is a very rare type of benign soft tissue tumour⁴. Cases reported in literature are anecdotal so overall incidence of this neoplasm is unknown. According to results of the larger study by Miettinen, DF is prevalent in adult people, aged over 40 with a sex distribution four times greater in the males. Site of occurrence of the tumour widely varies including both superior and inferior extremities, posterior neck and trunk, abdominal wall and hip region. The neoplasm is usually subcutaneous but occasionally it may be deep seated in fascial and sub-fascial structures².

The typical history of these patients is that of painless, slowly growing mass, with a duration of more than six months, that can reach in case of deep location considerable sizes, up to 20 cm. Infiltrative growth of surrounding structures is uncommon even if small area of fat, fascial, and muscle involvement may occur. According to the behaviour of all benign neoplasms no recurrences or metastases are observed after complete surgical removal⁵.

When superficial, the tumour is usually mobile with respect of the skin and in deep sections of subcutis, its growth is painless and no functional disorders are associated¹. When the tumor is deeply sited, especially in the limbs, the patient typically displays an enlargement and disfigurement of the afflicted region. Very small tumours may be occasionally detected in the course of ultrasonography or other radiological scanning.

Imaging diagnosis of DF depends mainly on ultrasonography, possibly associated with MRI and/or CT.⁵ The tumour appears as a homogeneous, capsulated, sharply demarcated, lesion exempt from contrast uptake. Conservative, complete surgical excision with functional preservation is the treatment of choice because of the benign nature of DF^{1,7}.

The process, on gross examination, does not exhibit specific patterns enabling differential diagnosis with other benign, locally aggressive, and low-grade malignant soft tissue tumours. No macroscopic patterns are characteristic of DF which can be confused with other benign and malignant soft tissue tumours such as desmoid tumours and low-grade fibromyxoid sarcoma which request more demanding therapeutic strategies^{1,2}. On the contrary diagnosis of DF may be confirmed on the basis of specific microscopic findings. Tumor cellularity is low, lesional cells are stellate or spindle shaped fibroblasts dispersed in collagenous or myxoid matrix. Nuclei are small, oval with delicate chromatin, but they can be plumped as in reactive fibroblasts. No atypical or hyperchromatic nuclei are observed. Even if exceptionally present, mitotic activity is usually absent as well as necrosis and angiogenesis. Tumour cells are characteristically positive for vimentin, and focal α -smooth muscle immunoreactivity has been observed especially at the periphery of

tumour. Absence of any reactivity for S100, CD34/CD68, and desmin have been generally reported as typical immunohistochemical patterns of DF. Since it was first described, it has been debated whether DF is a neoplasm or a reactive lesion^{8,9,10}.

Most recent genetic findings such as 11q12 breakpoints, diploid DNA and fibrous proliferation seem to support the neoplastic origin of DF^{11,12}.

In conclusion the reported case underlines the peculiar pathological patterns of DF and the consequent possibilities of a correct diagnosis and adequate treatment strategy.

Riassunto

Il fibroblastoma desmoplastico (DF) è un raro tumore benigno dei tessuti molli che insorge prevalentemente in soggetti in età adulta. Classificato come tumore dei tessuti molli, esso insorge frequentemente agli arti, mostrando lento e progressivo accrescimento. Istologicamente, il DF è costituito di cellule fusiformi immerse in uno stroma collagene, positive alla reazione immunoistochimica con la vimentina. La diagnosi differenziale con alcuni tumori dei tessuti molli localmente aggressivi può risultare difficile. Viene riportato un caso clinico relativo ad un paziente maschio, di anni 63, affetto da DF della coscia sinistra.

References

- 1) Watanabe H, Ishida Y, Nagashima K, Makino T, Norisugi O, Shimizu T: *Desmoplastic fibroblastoma (collagenous fibroma)*. J Dermatol, 2008; 35(2):93-97.
- 2) Miettinen M, Fetsch JF: *Collagenous fibroma (desmoplastic fibroblastoma): A clinicopathologic analysis of 63 cases of a distinctive soft tissue lesion with stellate-shaped fibroblasts*. Hum Pathol, 1998; 29(7):676-82.
- 3) Wilson C, Summerall J, Lubin J, Mesko TW: *Collagenous fibroma (desmoplastic fibroblastoma): A unique presentation as a goiter in an 88-year-old man*. Ann Diagn Pathol, 2000; 4(3):165-69.
- 4) Evans HL: *Desmoplastic fibroblastoma. A report of seven cases*. Am J Surg Pathol, 1995; 19(9):1077-81.
- 5) Jang JG, Jung HH, Suh KS, Kim ST: *Desmoplastic fibroblastoma (collagenous fibroma)*. Am J Dermatopathol, 1999; 21(3):256-58.
- 6) Shuto R, Kiyosue H, Hori Y, Miyake H, Kawano K, Mori H: *CT and MR imaging of desmoplastic fibroblastoma*. Eur Radiol, 2002; 12(10):2474-476.
- 7) Weisberg NK, DiCaudo DJ, Meland NB.J.: *Collagenous fibroma (desmoplastic fibroblastoma)*. Am Acad Dermatol, 1999; 41(2 Pt 2):292-94.
- 8) Sappino AP, Schürch W, Gabbiani G: *Differentiation repertoire of fibroblastic cells: Expression of cytoskeletal proteins as marker of phenotypic modulations*. Lab Invest, 1990; 63(2):144-61.

- 9) Schmitt-Gräff A, Desmoulière A, Gabbiani G: *Heterogeneity of myofibroblast phenotypic features: An example of fibroblastic cell plasticity*. Virchows Arch, 1994; 425(1):3-24
- 10) Bosman C, Boldrini R, Pierro V, Corsi A: *Unusual ultrastructural findings in giant-cell fibroblastoma*. Tumori, 1995; 81(4):283-89.
- 11) Sakamoto A, Yamamoto H, Yoshida T, Tanaka K, Matsuda S, Oda Y, Tsuneyoshi M, Iwamoto Y: *Desmoplastic fibroblastoma (collagenous fibroma) with a specific breakpoint of 11q12*. Histopathology, 2007; 51(6):859-60.
- 12) Bernal K, Nelson M, Neff JR, Nielsen SM, Bridge JA: *Translocation (2;11) (q31;q12) is recurrent in collagenous fibroma (desmoplastic fibroblastoma)*. Cancer Genet Cytogenet, 2004; 149(2):161-63.