

## Diagnostic and behavioural parameters differentiating proliferative musculo-fascial low grade lesions. Case reports

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**SUMMARY:** Diagnostic and behavioural parameters differentiating proliferative musculo-fascial low grade lesions. Case reports.

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*Pseudosarcomatous nodular fasciitis and desmoid tumors can be very similar at physical examination. Although their behaviours and cytologic aspects are very different, they both undergo the same surgical approach. Nevertheless, only desmoid tumors - because of their high rate of local recurrence - require a strict follow-up and further therapies when radicality of primary surgery could not be likely performed.*

**RIASSUNTO:** Parametri diagnostici e clinici delle lesioni proliferative musculo-fasciali a basso grado di malignità. Casistica personale.

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*La fascite nodulare pseudosarcomatosa e il tumore desmoide si possono presentare con aspetti clinici molto simili. Pur avendo comportamenti e aspetti istologici notevolmente diversi, prevedono lo stesso atteggiamento chirurgico ma solo per i tumori desmoidi l'alto rischio di recidiva locale suggerisce un attento follow-up e, nei casi di dubbia radicalità, terapie successive.*

**KEY WORDS:** Desmoid tumor - Extra-abdominal fibromatosis - Infiltrative fasciitis - Pseudo-sarcomatous nodular fasciitis - Pseudo-sarcomatous fibromatosis.  
Desmoide - Fibromatosi extra-addominale - Fibromatosi aggressiva - Fascite nodulare pseudo-sarcomatosa - Fibromatosi pseudo-sarcomatosa.

### Introduction

In 1832, Mc Ferlane first described a particular type of fibrous neoplasm originated in the muscular aponeuroses, characterized by a biologic behaviour intermediate between that of sarcomas and fibromas. In 1983, Müller coined the term desmoid tumor (1,2). In 1923, a case of extra-abdominal desmoid tumor was described by Nichols (3).

Desmoid tumors - also called extra-abdominal or aggressive fibromatosis - are rare fibromatous tumors of fibroblast origin which involve muscular connective tissue, muscular fascia and aponeuroses. They are locally very aggressive and tend to infiltrate adjacent tissue even if it is that thought their clinical behaviour appears to be quite different from that of sarcomas. In fact, desmoid

tumors are low-grade tumors which grow slowly but constantly, do not metastasize either through lymph nodes or through blood circulation; however, they have high rates of local recurrence (4,5).

As the scarcity of cases limited the performing of statistically significant perspective studies, desmoid tumors - although more frequently diagnosed nowadays - still stimulate the curiosity of the scientific community. Overall, desmoid tumors are reported to account for less than 0.03% of all neoplasms, with only 2-4 new cases per million of inhabitants/year (6). They are fibroproliferative disorders arising from musculoaponeurotic tissue and their high local recurrence rates suggested a reactive etiology; however, recently it has been shown that, even if this tumor does not metastasize, a true neoplastic process underlies fibroblast proliferation due to desmoid cell clonality (7).

Three main theories have been proposed to explain the pathogenesis of desmoid tumors: mechanical theory, endocrine theory, genetic theory.

According to the first theory, desmoid tumors may be associated with trauma, stretch injuries of the abdominal wall and especially enlarging pregnant uterus; the

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higher incidence of desmoid tumors in multipregnancies and some cases observed after actinic therapy seem to confirm this theory (8,9).

The endocrine theory is based on the fact that desmoid tumors are often associated with both endocrine disorders and high estrogens levels. In fact, women of fertile age who develop a desmoid tumor may have a predisposition to estrogen predominance over progesterone. In female, a direct relationship between tumor growth rate and endogenous estrogen levels has been reported: a significant increase in estradiol but not in progesterone receptor concentrations in the tumor cell cytoplasm has been observed; it suggests that desmoid tumor is highly influenced by sex hormones (10). Positive answers of patients undergoing antiestrogenic therapy (Tamoxifen) - strengthened by in vitro studies which have shown the inhibition of desmoid cell proliferation - support this theory (11,12).

The genetic theory is supported by the evidence that 2% of desmoid tumors have a genetic origin. They may appear due to APC mutation in patients with familial adenomatous polyposis (FAP) or Gardner's syndrome. Indeed, the fact that 10% of desmoid tumors occur in patients with FAP and 38% in those with Gardner's syndrome, suggests a common genetic origin of both pathologies. This theory is supported by the presence of trisomy 8 and 20 in desmoid cells (13-15). An autosomal dominant syndrome of hereditary desmoid disease (HDD) due to mutation at codon 1924 of the APC gene has been reported (16). However, in most cases, the etiology remains unknown (10). The neoplastic transformation of desmoid tumors into sarcoma is extremely rare; Literature reports only 5 cases (11,17,18).

Nodular fasciitis, which on the contrary is a benign, rapidly growing soft tissue tumor characterized by fibroblastic and miofibroblastic proliferation of uncertain etiology has a similar clinical presentation. Its macroscopic appearance and clinical behaviour make pre-operative diagnosis extremely difficult because of its ability to simulate a malignant process (19-21). Nodular fasciitis was first described by Konwaler et al. in 1955 (22), as a pseudosarcomatous fasciitis with a rich mitotic activity. In Literature the terminology for this type of lesion may be: proliferative fasciitis, infiltrating fasciitis and pseudosarcomatous fibromatosis (23).

It is a benign rapidly growing reactive fibroblastic proliferation of uncertain etiology, which may occur consequently to a non-specific inflammatory process, although trauma has also been implicated. More recently, clonal chromosomal abnormalities have been reported in a small number of cases, suggesting that at least some cases of nodular fasciitis may represent a clonal myofibroblastic tumor (24,25). Nodular fasciitis may occur in three main forms according to the location of the lesion: 1) the *subcutaneous form* is more frequently observed and presents

as a small subcutaneous defined nodule; 2) the *intramuscular form* is rare, about 10% of all the fasciitis, has a larger size and infiltrates; 3) the *fascial form* with irregular dendritic margins originates from the superficial fascia.

Clinical observation, study and surgical treatment of 4 patients with solid musculofascial neoformations – of which 3 turned out to be desmoid tumors (aggressive fibromatosis) – gave us the possibility to critically evaluate different problems related to diagnosis and treatment of these patients.

## Case reports

Four patients (3 females and 1 male) were observed in the Surgical Science Department, "Sapienza" University of Rome, over the last two years with proliferative intramuscular neoformations (Table 1), having quite similar clinical characteristics; the final diagnosis was aggressive fibromatosis (desmoid tumors) in 3 patients and nodular fasciitis in 1 of them. First two cases concerned two young women (FF, 28-year old and CG, 36 year-old), both with recent pregnancies in their history who reported a rapidly growing mass in the rectus abdominis muscle; they noticed the mass for the first time 6-8 months before. The first case was approximately 4 cm in diameter and it was located at the left side of the rectus abdominis muscle; the second one, was located at the right side of the rectus abdominis muscle and it was about 7 cm in diameter.

In both patients, physical examination revealed an irregular, smooth-surfaced mass of hard parenchymatous consistency with quite well defined margins; it was fixed to the muscles and followed muscle contraction; it was almost asymptomatic and only slightly painful upon palpation. In both cases, echographic examination showed an inhomogeneous mainly hypoechoic structure with polycyclic and irregular margins and minimal peripheral vascularization. An MRI was performed on the first patient and a CT scan on the second one. Both examinations showed the integrity of adjacent structures and confirmed both tumor size and its perilesional vascularization.

The first patient underwent an echoguided tru-cut needle biopsy: despite the small quantity of tissue collected, extemporaneous histological examination revealed a lesion made of fibrous tissue compatible with fibromatosis. Definite diagnosis was achieved by final histologic examination. Pre-operative biopsy was not performed on the second patient because of benign radiologic features of the lesion (well-defined margins and perilesional vascularization). Both patients underwent surgical intervention: a pararectal incision was performed (on the left and on the right side respectively) and the mass was excised in toto with most of rectus abdominis muscle and its aponeurosis trying to obtain clear surgical margins. As a clear cleavage plane was achieved in both patients, it was not necessary to open the peritoneum. Wall repair was performed using a double thickness of Marlex mesh cut to appropriate dimensions that was sutured to musculoaponeurotic structures using prolene sutures. Postoperative course was uneventful in both patients. At present - 2 years after surgery – there is no evidence of recurrence.

Third case concerned a 42-year-old man, with no major pathology at his past medical history who, over the past 4 months, noticed the onset of a hard and elastic lump of approximately 3 cm in maximum diameter in the medial third of the right thigh of quadriceps femoris muscle. Ecographic examination revealed a solid, encapsulated lump of 33 mm in maximum diameter in the quadriceps femoris muscle compressing the surrounding area with no evidence of neoplastic infiltration. Power-doppler did not reveal intralesional vascular spots.

TABLE 1 - PATIENTS.

| N | Patient | Sex | Age | Location / Muscle        | cm      | Vascularization | Histology         |
|---|---------|-----|-----|--------------------------|---------|-----------------|-------------------|
| 1 | F. F.   | F   | 28  | Left Rectus abdominis    | 4 x 1,6 | Perilesional    | Desmoid tumor     |
| 2 | C. G.   | F   | 36  | Right Rectus abdominis   | 7 x 5   | Perilesional    | Desmoid tumor     |
| 3 | G. F.   | M   | 43  | Right femoral quadriceps | 3 x 1,7 | Perilesional    | Desmoid tumor     |
| 4 | C. A.   | F   | 30  | Adductor magnum          | 3 x 3   | Intralesional   | Nodular fasciitis |

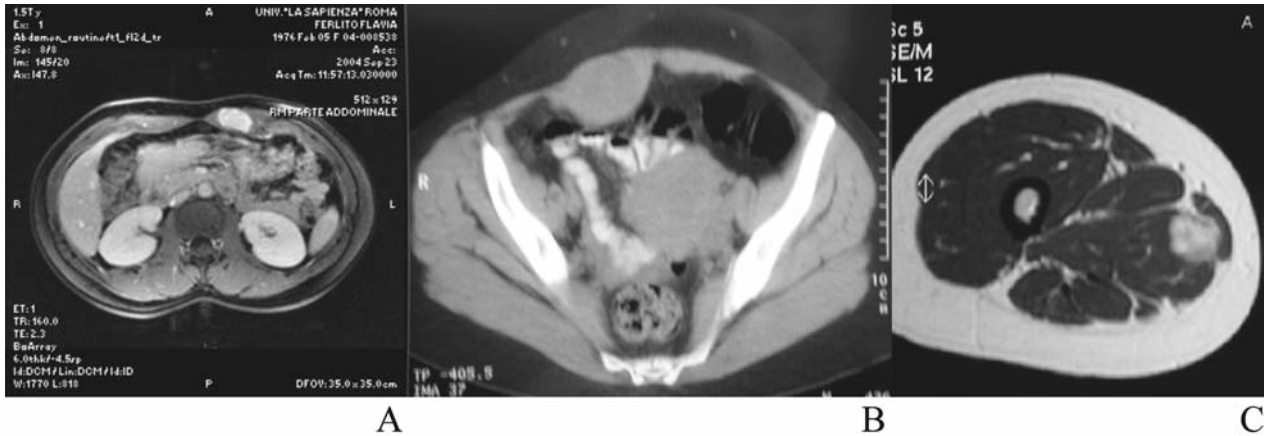


Fig. 1 - A) CT scan with intra-venous contrast injection showing the mass located in the left rectus abdominis wall. B) CT-scan showing the mass located inside the right rectus abdominis muscle. C) CT scan of the right thigh: the lesion appears to be very vascularized.

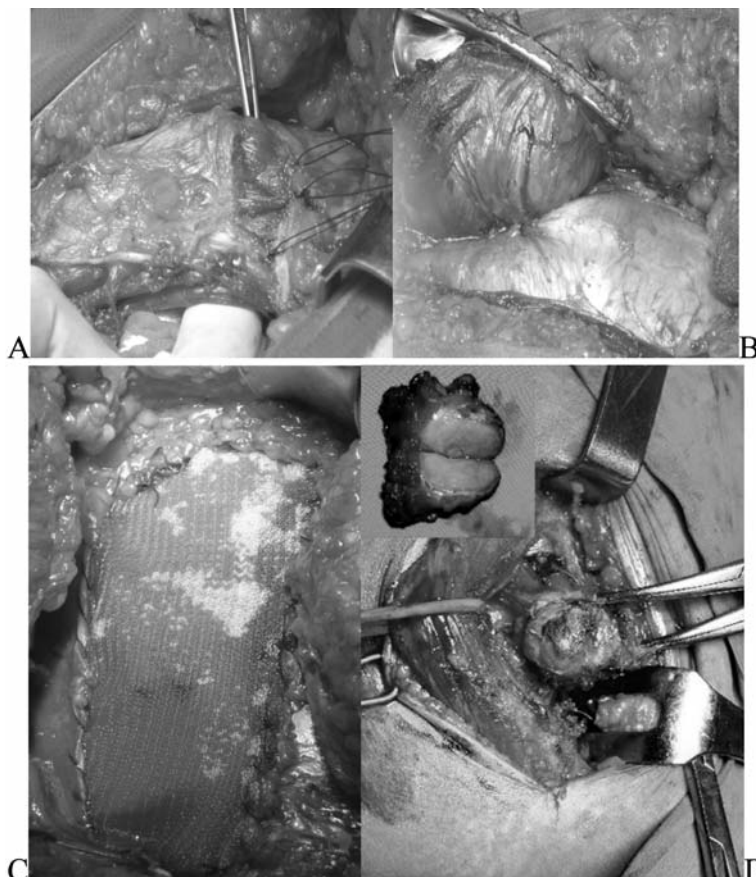


Fig. 2 - A-B) Intraoperative picture showing the integrity of the peritoneal cavity. C) Intraoperative picture: partial wall repair by a double layer of overlapped Marlex mesh. D) Intraoperative aspect of the lesion – the sartorius muscle has been di-vascularized downwards.

CT scan - performed before and after intravenous contrast injection - showed a round, hypo-isodense nonenhancing lump with well-defined margins measuring about 1,8 x 3,5 cm in the quadriceps femoris muscle. The patient refused to undergo needle biopsy and, indeed, biopsy was not required to plan surgical intervention because of small tumor size. Surgical intervention removed the mass by making attention to achieve, where possible, clear surgical margins. Cut section of the operative specimen revealed a whitish pseudocapsulated oval-shaped mass of hard elastic consistency.

Fourth case concerned a 30-year-old woman. Over the past 1 month she noticed a nodular rapidly growing lesion located deeper in the muscles of the proximal third in the medial region of the right thigh. This roundly-shaped lesion, measuring approximately 3 cm in diameter, was fairly attached to the surrounding tissues and had a hard parenchymatous consistency. This nonpulsating, painless but slightly painful mass had almost regular but not well-definable multilobulated margins. The overlying skin showed no sign of phlogosis. Echographic evaluation revealed a solid lesion, but it did not provide any further information about it. CT-scan - performed after and before intravenous contrast injection - revealed a roundly-shaped hypodense lesion in the middle third of the adductor magnus muscle of the right thigh; after intravenous contrast injection, CT showed a remarkable increase in lesion density and a small hypodense central area. This mass appeared to have well-defined margins and a regular shape referable to the neof ormation arising from the right adductor magnus muscle. Radiologist suggested an MRI for further evaluation. The MRI - performed before and after intravenous contrast injection using SE and SPIR sequence in axial and coronal planes - confirmed that the lesion observed in the adductor magnus muscle extended approximately 3 cm craniocaudally. On SE T1-weighted images the lesion appeared isointense, while it was markedly hyperintense after contrast injection. The rich vascularization of the lesion and its well-defined margins lead us to a pre-operative diagnosis of soft tissue lesion, probably benign. We couldn't exclude its angiomatous nature. Because of the rich vascularization and radiologic features of benignity, pre-operative needle biopsy was not performed. Surgery was performed to remove the mass that was almost completely indovated within the adductor magnus muscle; the sartorius muscle was isolated and cut and the underlying lesion was identified and excised. A wide area of muscular tissue from around the lesion was removed in order to achieve clear surgical margins. This tissue appeared to be adherent to the lesion so as that a neoplastic infiltration was suspected. Cut section of the lesion revealed a solid, grey-lardaceous, encapsulated mass. Postoperative course was uneventful and the patient recovered rapidly with no functional deficit. At present - more than one year after surgery - there is no evidence of recurrence.

Histologic examination - that was similar in the first three patients - showed a proliferation of fusiform cells arranged in long fascicles with infiltrative growth pattern involving the perilesional skeletal muscular tissue. There was strong evidence of atrophic phenomenon due to compression. Histology was compatible with the diagnosis of desmoid tumor (extra-abdominal aggressive fibromatosis).

On the contrary, in the fourth patient histologic examination revealed a sarcomatous-like lesion characterized by short fascicles of fusiform and star-like cells arranged in a storiform growth pattern and cheloid-like central areas surrounded by several neof ormed capillary vessels associated with haemorrhagic extravasations and inflammatory lymphocytic infiltration. Histology was compatible with the diagnosis of nodular fasciitis.

Differential diagnosis between desmoid tumor and nodular fasciitis is based upon the greater size and mainly infiltrative growth pattern of the first lesion in comparison with the second one. Desmoid tumor is characterized by a proliferation of fusiform cells arranged in long fascicles, while nodular fasciitis is characterized by short fascicles interlaced in a storiform-like pattern. Nodular fasciitis is also characterized by huge haemorrhagic extravasations and in-

flammatory lymphocytic infiltration, not so evident in desmoid tumor. Mitoses - which occur in both tumors - are usually much more numerous in nodular fasciitis.

## Discussion

Musculo-aponeurotic lesions, especially extra-abdominal ones, sometimes have very similar clinical and semeiologic features characterized by difficulty in diagnosis and doubts on therapeutic choice.

Both pre-operative needle aspiration biopsy and extemporary histologic examination are not always able to make a definite diagnosis. In fact, only the whole cytoarchitecture of the tumor provides elements for a definite diagnosis. Several therapeutic options have been proposed to treat desmoid tumors: surgical excision, radiotherapy, adjuvant radiotherapy, chemotherapy, polichemotherapy, the use of antiestrogens and FANS (26-29).

We discussed the opportunity to perform either a FNA or a pre-operative biopsy to better plan surgical intervention. Indeed, compartmental resection - when it is possible - should be the therapy of choice for sarcoma; on the contrary, it would be undoubtedly excessive in the treatment of benign lesions - such as pseudosarcomatous nodular fasciitis - as well as of low-grade tumor - such as desmoid tumors, that have a clinical behaviour completely different from that of sarcomas, do not metastasize and, at least, can recur locally - . However, a wide or, at least, a marginal excision of the lesion should be performed trying to avoid intracapsular excisions in order to lower the risk of local recurrence due to tissue residual. It is believed that desmoid tumors rarely transform into sarcoma: literature reports only 5 cases of malignant degeneration (10, 17,18), even if the true incidence could be likely to be underestimated, because the lack of systematically oriented investigations.

From a technical point of view, the only recommendation concerning the exeresis of perilesional healthy tissue of legs is to respect adjacent vascular and nervous structures. Abdominal wall lesions with no evidence of infiltration can be better treated by surgical excision involving only the peritoneal wall. It may allow the use of polypropylene meshes which represent the most appropriate solution to the problem of abdominal wall repair because of their characteristics: stability over time, plasticity, biological inertia, resistance to infection, possibility to be easily moulded and to double thickness in order to provide greater support (30).

Among the international community, the advantageousness of performing either demolitive resections (31) or radiotherapy (32) - as proposed by some Authors - is still an open issue.

Adjuvant radiotherapy seems to be the most effecti-

ve treatment to prevent recurrence of desmoid tumors after radical, marginal and even intralesional surgery. Obviously, recurrence rate depends on the kind of surgical resection performed, being lower in case of radical surgery (33). Adjuvant radiotherapy following non radical surgery should lower the chances of local recurrence (34). On the contrary, other Authors believe that adjuvant radiotherapy associated to surgery does not reduce recurrence rate but, indeed, it may double it (6)<sup>6</sup>; anyway we have no experience about it.

Some Authors – considering desmoid tumors are just locally aggressive and have a long-term good prognosis - suggest a less aggressive attitude: if radical surgery risks to be mutilant or to leave severe effects, radiotherapy (50-60 Gy) or chemotherapy with Doxorubicin and Dacarbazine may control the neoplasm for a long time (28-35). Tamoxifene seems also to control the tumor, as it has been shown by in vitro studies of inhibition of desmoid cell proliferation (11,12). From a diagnostic point

of view, pseudosarcomatous nodular fasciitis is less aggressive than desmoid tumors; in our experience the only difference concerned the size and the kind of vascularization of the lesion, which was intralesional only in this case. Local recurrence might therefore be due to diagnostic mistakes or incomplete exeresis; however, a reliable pre-operative diagnosis is seldom available, therefore the most correct surgical behaviour is similar to that of desmoid tumors.

Pseudosarcomatous nodular fasciitis and desmoid tumors have very similar clinical characteristics. Surgical excision of nodular fasciitis lesions is usually curative; on the contrary, desmoid tumors – despite they are low aggressive tumors – tend to recur locally. Therefore, primary surgery with negative surgical margins is the most successful treatment method, trying to avoid mutilant surgery and using chemotherapy, radiotherapy and hormono-therapeutic treatments to lower the chances of recurrence.

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