Collagenous fibroma (desmoplastic fibroblastoma): clinical, pathological and imaging findings

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Authors: J. A. Narváez¹, X. Sanjuan², I. Santos³, J. Hernández Gañán⁴, D. Rodríguez², J. Viñals¹; ¹L’Hospitalet de Ll. (BARCELONA)/ES, ²L’Hospitalet de Llobregat/ES, ³Sant Boi (Barcelona)/ES, ⁴L’Hospitalet de Llobregat/ES
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Purpose

Our purpose is to describe the clinical, pathologic and imaging findings of collagenous fibroma, a rare benign soft-tissue tumor, first described in 1995 with the name of desmoplastic fibroblastoma.
Methods and Materials

A search of patients with collagenous fibroma (desmoplastic fibroblastoma) pathologically diagnosed in a tertiary teaching hospital retrieved 6 cases, 2 of them surgically excised without previous imaging study.

The remaining four cases constituted our study population.

Clinical charts and imaging studies were retrospectively reviewed to register clinical complaints, tumor location, size, type of margins, signal intensity, echostructure, and attenuation coefficient, as well as homogeneity/heterogeneity, contrast-enhancement pattern, presence of calcifications or cystic-necrotic changes, perilesional edema, and relationships with neurovascular and bone structures.

Pathological features were also reviewed.
Results

**OUR SERIES:**

There were three males and a female, with a mean age of 63 yrs. (range: 42-81). All cases presented painless soft tissue growing mass. The initial clinical suspicion was supraclavicular lymph adenopathy in one case with lip carcinoma treated 4 years before.

Plain films and MR imaging were performed in all cases, ultrasound (US) in 3, and angio-CT in 1 case. Pathological diagnosis was obtained by percutaneous US-guided tru-cut biopsy in 3 cases, and by incisional biopsy in 1 case.

**Radiographs** showed no calcifications or bone erosions (Fig. 1 on page 8).

On imaging studies, tumoral lesions were deeply located in the thigh (n=2), supraclavicular fossa and ankle. Close relationship with adjacent major vessels was seen in three cases, in one completely surrounding the subclavian artery, but its diameter and permeability was no affected.

Mean tumor size was 7 cm (range: 4-13).

Lesional borders were well-defined in all cases.

**Ultrasound** showed solid, hypoechoic lesions, with small hyperechoic areas (Fig. 19 on page 25 and Fig. 23 on page 29).

**Computed tomography** was performed in one case, demonstrating a solid mass with attenuation similar to the muscle (Fig. 10 on page 16, Fig. 11 on page 17 and Fig. 12 on page 18), surrounding the subclavian artery.

**Magnetic Resonance Imaging:** On T1-WI, lesions were iso-hypointense (n=3) or isointense (n=1) to muscle (Fig. 2 on page 8, Fig. 4 on page 10, Fig. 13 on page 19, Fig. 20 on page 26, Fig. 24 on page 30, Fig. 25 on page 31, and Fig. 26 on page 32). On T2-WI, all lesions were homogeneously hypointense to muscle (Fig. 14 on page 20, Fig. 15 on page 21, Fig. 25 on page 31, Fig. 26 on page 32), in 2 cases with small hyperintense foci Fig. 3 on page 9 Fig. 21 on page 27 . Fat-sat Gd-T1-WI were obtained in 2 cases (Fig. 16 on page 22 and Fig. 22 on page 28), demonstrating mild, somewhat heterogeneous enhancement. Perilesional edema was seen in 1 case.
Pathological findings

Grossly, it appears as a well-circumscribed oval, usiform-elongated, or disc-shaped mass (Fig. 6 on page 12, Fig. 7 on page 13 and Fig. 18 on page 24).

Microscopically, these lesions were hypocellular tumours with widely spaced spindle- and stellate cells with elongated nuclei in a collagenous or myxocollagenous stroma (Fig. 8 on page 14 and Fig. 9 on page 15).

The margins are variably circumscribed.

Lower cellularity, lack of fascicular pattern, predominance of amorphous collagenous stroma and inconspicuous vasculature separate it from desmoid tumour.

Treatment

In all cases of our series, the tumour was excised with wide margins.

LITERATURE REVIEW

In a literature review, we have found 142 reported cases of collagenous fibroma. Theses cases have been described in 54 papers (1-54), 9 of them not written in English (5, 14, 25, 30, 37, 40). Most cases were published in pathological journals.

Collagenous fibroma (desmoplastic fibroblastoma) is included within the fibroblastic/myofibroblastic tumors in the WHO classification. The WHO definition of this tumour is: "a rare, benign, paucicellular tumour affecting mainly adult males, characterized by densely collagenous, predominantly stellate-shaped fibroblast exhibiting bland cytological features. Myxoid stroma may be present."

Age range is wide, but approximately 70% of cases are diagnosed in men between the 5th and 7th decades.

The tumour typically presents as a painless, subcutaneous mass, but fascial involvement is common and up to 25% of cases involve skeletal muscle.

It occurs in a variety of peripheral sites, being the most common locations the arm, shoulder, lower limb, back, forearm, hand and feet.
The tumour is usually relatively small, measuring 1-4 cm in greatest dimension, but examples over 10 cm and as large as 20 cm have occurred. In our cases, the mean size was 7 cm.

The behaviour is benign, and none of the published series had recurrences (1-54).

Accurate discussion of the imaging findings, focused on MRI, was found in a minority of reports (6,13,20,26,28,36,54).

Smooth erosion into bone has been reported (6).

Imaging findings in our series are similar to the previously described.

**Differential diagnosis**

The main radiological differential diagnosis is Desmoid Tumor (Fig. 27 on page 33), a connective tissue neoformation originated in the connective tissue of the muscle, overlying fascia or aponeurosis.

Other possible radiological differential diagnoses of Collagenous Fibroma are Fibroma of the Tendon Sheath (Fig. 28 on page 34), Nucal Fibroma and Giant Cell Tumor of the Tendon Sheath (Fig. 29 on page 35).

Occasionally, malignant soft-tissue sarcomas, such as Fibrosarcoma or High Grade Pleomorphic Sarcoma (formerly named Malignant Fibrous Histiocytoma) may appear predominantly hypointense. However, depiction of necrotic/cystic or hemorrhagic areas, as well as local infiltrative signs may suggest the diagnosis.

Intra- or periarticular Collagenous Fibroma may be associated with bone erosions (6). The main differential diagnosis in this scenario is Pigmented Villonodular Synovitis (6,26). However, other entities that should be excluded are Amyloid Arthropathy, Gout, Tuberculous Arthritis and more rarely chronic Rheumatoid Arthritis (6).

Soft-tissue calcified masses such as myositis ossificans (Fig. 30 on page 36), calcinosis tumoral, extraskeletal chondrosarcoma or osteosarcoma, and synovial sarcoma may present low signal intensity on both T1- and T2-weighted MR images (26). These entities can be readily distinguished from Collagenous Fibroma with radiographs, which demonstrate the absence of calcifications in this tumor.
**Fig. 1:** CASE 1. Plain film (lateral view) of the knee shows a huge, non calcified, soft tissue mass (arrows).

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**Fig. 2:** CASE 1. Axial T1-weighted MR image demonstrates a well-defined mass in the posterior compartment of the thigh, close to the femoral artery (A) and vein (V). Signal intensity of the tumor was intermediate, with areas of very low signal intensity.

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Fig. 3: CASE 1. Axial T2-weighted MR image: signal intensity of the soft-tissue mass is predominantly low, with small foci of intermediate to high signal (arrows). A and V: popliteal artery and vein.

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**Fig. 4:** CASE 1. Coronal T1-wweighted MR image show the full extent of the tumor, which is located close to the femoral vessels.

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Fig. 5: CASE 1. Illustration of the left thigh and knee of this patient: surgical excision lines are drawn in the skin.

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Fig. 6: CASE 1. Surgical specimen of the tumour.

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Fig. 7: CASE 1. Gross specimen of the tumour.

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**Fig. 8:** CASE 1. Photomicrograph of the excised specimen shows dense collagenous matrix and spindle to stellate tumor cells.

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Fig. 9: CASE 1. Photomicrograph of the excised specimen (H&E X100) demonstrates a tumoral area of dense collagenous background.
**Fig. 10**: CASE 2. Consecutive coronal CT images show a solid mass of intermediate attenuation, surrounding the subclavian artery (A) and close to subclavian vein (V). Note normal diameter and permeability of the subclavian artery.

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**Fig. 11:** CASE 2. Consecutive coronal CT images show a solid mass of intermediate attenuation, surrounding the subclavian artery (A) and close to subclavian vein (V). Note normal diameter and permeability of the subclavian artery.

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**Fig. 12:** CASE 2. Consecutive coronal CT images show a solid mass of intermediate attenuation, surrounding the subclavian artery (A) and close to subclavian vein (V). Note normal diameter and permeability of the subclavian artery.

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Fig. 13: CASE 2. Axial T1-weighted MR image: the soft tissue mass surrounds the subclavian artery (A). Tumour margins are well defined and signal intensity is intermediate, with small areas of low signal.

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**Fig. 14:** CASE 2. Sagittal T2-weighted MR image shows a well-defined lesion, of homogenous low signal intensity. Note relationships with subclavian artery (A) and vein (V).
Fig. 15: CASE 2. Coronal T2-weighted MR image shows a well-defined mass, with homogeneous low signal intensity. Note close relationship with the subclavian artery (A), which is surrounded by the tumor, and vein (V).

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Fig. 16: CASE 2. Coronal gadolinium-enhanced fat-suppressed T1-weighted MR image: note peripheral enhancement (arrows), as well as a moderate, patchy, tumoral enhancement.

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Fig. 17: CASE 2. Picture of the left supraclavicular area of this patient taken immediately before surgery. Note the mass deforming the skin surface.

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Fig. 18: CASE 2. Macroscopic specimen of the resected tumor.

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Fig. 19: CASE 3. Ultrasound axial scan demonstrates a solid tumor at the vastus medialis muscle (arrows), with somewhat heterogeneous echogenicity, including some hyperechoic intralesion areas. Note close relationship with cortical margin of the distal femur.

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**Fig. 20:** CASE 3. Axial T1-weighted MR image demonstrates a intramuscular lesion (arrows) of intermediate signal intensity, with intralesional bands of very low signal intensity.

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**Fig. 21:** CASE 3. Axial T2-weighted MR image: the lesion (arrows) present low intensity signal, similar to muscle. Only small hypertense, peripheral areas of high signal intensity are identified.

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Fig. 22: CASE 3. Axial gadolinium-enhanced fat-suppressed T1-weighted MT image. There is moderate enhancement of the lesion, being better delineated from surrounding muscle. The intralesional bands of very low signal intensity are well-identified.

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**Fig. 23: CASE 3.** Panoramic ultrasound view in the coronal plane shows the full extent of the tumor, well demarcated, in contact with the hyperechoic cortical margin of the distal femur. This lesion are mostly hypoechoic, with smal intralesional, lineal, hyperechoic foci.

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Fig. 24: CASE 3. Coronal T1-weighted image depicts the full-extent of the lesion. Note contact of the tumor with the cortical margin of the distal femur, without signs of bone erosion or infiltration.

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Fig. 25: CASE 4. Sagittal T1-weighted and coronal fat-suppressed T2-weighted MR images show a soft-tissue mass, with well-defined margins (arrows). Note close relationship with the posterior tibial vessels (V). Signal intensity is intermediate, with small hypointense areas on T1-weighted, and homogeneously low on T2-weighted image.

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Fig. 26: CASE 4. Axial T1-weighted and T2-weighted MR images: note the proximity of the tumour (arrows) to the posterior tibial artery (A) and veins (V).

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Fig. 27: DESMOID TUMOUR. MR images show two soft-tissue masses located at the lateral region of the hip and at the abdominal wall. Margins of the tumour are partially spiculated because of infiltration of adjacent structures (arrows) Signal intensity of the tumour located at the hip is intermediate to low in both T1-and T2-weighted images, whereas abdominal wall tumour shows homogeneous low signal on T2-weighted image. This low signal intensity corresponds to hypocellular tissue and dense collagen.

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**Fig. 28:** FIBROMA OF THE TENDON SHEATH. Axial T1-weighted and FS T2-weighted MR images show a soft-tissue tumor at the dorsal aspect of the hand (arrows), surrounding the extensor tendons. Signal intensity is low in both pulse sequences, with small foci of intermediate signal. Signal intensity reflects histological composition of tightly packed spindle cells, vessels, and large amounts of dense collagenous matirz, which is markedly hyalinized.

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Fig. 29: GIANT CELL TUMOUR OF THE TENDON SHEATH. MR images demonstrate a well-defined mass at the lateral region of the 5th toe (arrows). Note close relationship of the mass with the extensor tendon (t). Most of the tumour has intermediate signal intensity, but portions of the tumour have low signal intensity in all sequences; the latter finding reflects signal attenuation due to hemosiderin deposition. After gadolinium injection, the tumour presents heterogeous enhancement.

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Fig. 30: MYOSITIS OSSIFICANS. Plain films demonstrate well-circumscribed calcifications adjacent of the lateral margin of the distal femur and fibula. Findings of more dense peripheral calcification and central lucency are compatible with the diagnosis of myositis ossificans. Axial T1-weighted and FS T2-weighted images: soft tissue mass shows low signal intensity due to calcification (great arrows). Note post-surgical changes at the subcutaneous tissue (small arrow).

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Conclusion

Similarly to described at the literature, in our series this tumour tends to appear in median-aged to older males, as a painless soft-tissue mass.

In our experience, the most distinctive imaging features are:

1. the low signal intensity on T2-weighted MR images, which corresponds pathologically with collagenous tumoral stroma, and
2. the close relationship with major vessels. This late feature, along with the well-defined borders may be useful to differentiate them from desmoid tumor.
References


