

## EXTRA-ABDOMINAL MUSCULO-APONEUROTIC FIBROMATOSIS: A CASE WITH METACHRONOUS MULTICENTRIC LESIONS OF THE RIGHT THIGH

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### Summary

Fibromatoses are a group of benign proliferations of fibrous tissue with clinical behaviour ranging from that of truly malignant tumours to that of benign reactive fibrous proliferations. Some of the superficial fibromatoses are fairly common but deep ones, also known as desmoid tumours or musculoaponeurotic fibromatoses, are rare. We describe a case of extra-abdominal musculoaponeurotic fibromatosis in a 22-years old male. He underwent 14 operations for a period of 9 years for fibromatosis of the posterior surface of the right lower limb. The histological findings of the lesions were similar consistent across surgeries and consistent with a diagnosis of aggressive extra-abdominal fibromatosis.

**Key words:** musculoaponeurotic fibromatosis, aggressive fibromatosis, deep extra-abdominal fibromatosis

### Introduction

The term aggressive fibromatosis (AF) refers to a large group of benign fibrous tissue proliferations of similar microscopic appearance that in their biological behaviour are intermediate between benign fibrous tissues and fibrosarcomas[1].

The term "desmoid" derives from the Greek word "desmos" meaning band or tendon and was coined by Mueller in 1836[2].

The first description of a desmoid tumour is, however, credited to Mac Farlane in 1832[3]. AF is a relatively rare lesion representing less than 3% of all soft tissue tumours with a reported annual incidence of 0.2–0.5 per 100,000 population [4,5].

Young adults are most commonly affected, with peak prevalence between the ages of 25 and 35 years. Lesions in patients younger than 10 years are uncommon[5].

These types of fibromatoses most frequently occur at the torso and extremities. The latter localization accounts for approximately 70% of all

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cases. Frequently affected sites include the shoulder (20%), the chest wall and the back (15%), the thigh (12%), the mesentery (10%), the neck (10%), and the knee (7%) [6, 7].

Although desmoid tumors most commonly arise from the rectus abdominis muscle in postpartum women and in scars due to abdominal surgery, they may arise in any skeletal muscle.

## Case description

A twenty-two-year-old patient presented with a history of recurrent multiple swellings of the right thigh. He underwent 14 operations for a period of 9 years for fibromatosis of posterior surface of the right lower limb.

At the time of the first surgery, the tumor was sized 20x15x15cm and was found mostly in the gluteal region, enveloping the ischiadic nerve without infiltration of its nerve sheath.

Out of the total amount of 34 nodules removed, 21 were removed from the thigh and 13-from the calf region. The myotendinous junctions were exclusively involved.

Each recurrence involved new areas, not affected on previous occasions.

On examination, there was a localized or more diffuse enlargement of the thigh muscles. The swellings were firm to bony hard in consistency but usually painless.

Soft plain films revealed a slight distortion of the outlines and fat planes of the affected muscle group. The underlying bones were normal.

On MRI the affected limb presented with multicentre superficial or deep-seated lesions, some of them well-defined, others-ill-defined, with a slight distortion of the anatomy of the postero-lateral muscle groups, due to multiple previous surgeries. The lesions (involving mostly the myotendinous junctions) were hypointense on T1WI to the affected muscles above and below the knee. On T2WI and IR (with fat suppression) the nodules were hyperintense.

Conclusion: A case of multicentre extra-abdominal desmoids recurrence.

## Grossly

The formation was found very firm and white with whorled appearance. The tumour measured 15x7x5.5 cm (Fig. 1).



Figure 1. Macroscopic appearance of the lesion

## Microscopic examination

Under the microscope the lesion was well-differentiated and infiltrated the adjacent tissue. Microscopic examination showed interlacing bundles of uniform spindle-shaped cells with pale oval nuclei and tails of eosinophilic cytoplasm arranged in streaming fascicles. Some cells had inconspicuous nucleoli but there was no hyperchromasia or mitotic figures. There was a prominent collagen component seen as broad collagen bands.

The degree of cellularity was variable – some areas demonstrating a moderate number of lesional cells in the background stroma of abundant mature collagen, while others showed minimal stroma with a large number of active mesenchymal cells (Figures 2 and 3). The cellularity was most evident at the periphery of the tumour (Fig. 4).

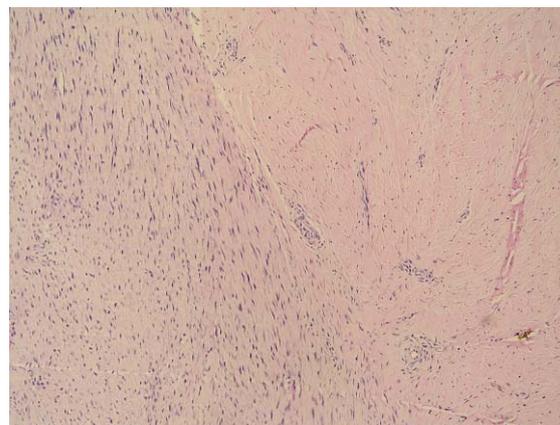
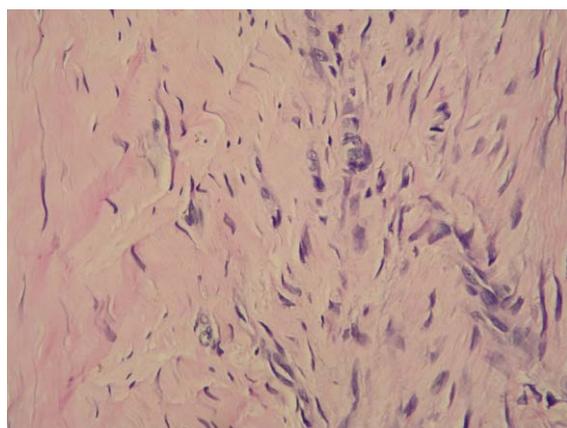
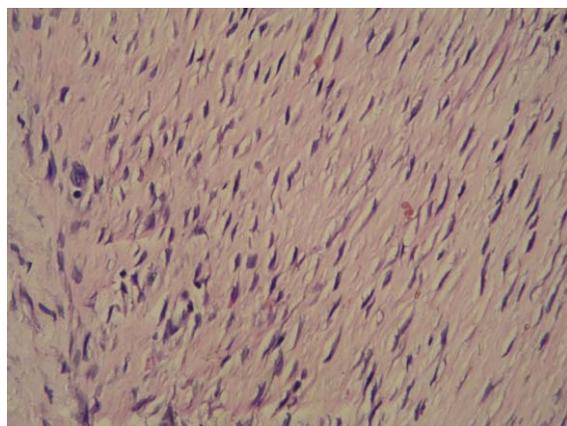


Figure 2. Microscopic appearance of the lesion- H&E staining, orig. magn. 10x20



**Figure 3.** Pleomorphic nuclei and collagen production in the stroma of the tumor. H&E staining, orig. magn. 10x40



**Figure 4.** Hypercellular zone at the periphery of the tumour. H&E staining, orig. magn. 10x40

## Discussion

Fewer than 100 cases have been reported, of which 60% have been in the metaphysis of the long bones of the appendicular skeleton[7].

AF is characterized by low mitotic activity and a strong infiltrative growth pattern along tissue planes, with an ability to invade adjacent tissues[1].

The tumours tend to infiltrate adjacent muscle bundles, frequently entrapping them and causing their degeneration. Myofibroblasts are the cells considered to be responsible for the development of desmoid tumours[8].

Musculoaponeurotic fibromatosis is histologically identical to the desmoid tumour.

Though deep or aggressive, fibromatosis is not considered a malignancy. It does not metastasize but multicentric lesions do occur – [1, 4, 5, 9-13].

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