Low-grade fibromyxoid sarcoma of the left hand

A 17-year-old girl presented with a slow-growing soft tissue mass of the left thenar for a couple of months (Fig. 1). The mass was tender and felt warm at clinical examination. There was no recent trauma. A magnetic resonance imaging (MRI) of the left hand was performed.

**Imaging Findings:**

A MRI was performed and showed a tumour in the thenar muscle with hypointense margin (fibrous tissue) and hyperintense (myxoid tissue) centre (Fig. 2a, 2b, 3a). The lateral part of the tumour had a gyriform to lobulated shape. After contrast administration, there was no enhancement in the central myxoid and peripheral fibrous component of the tumour (Fig. 2c, 3b).

After bolus injection, the time intensity curve showed no enhancement in the myxoid and fibrous component of the tumour (Fig. 4).

A subsequent plain radiography was performed to exclude myositis ossificans (Fig. 5). A mass of the thenar was seen. There were no calcifications or ossifications.

After histopathological examination of the biopsy and the resected mass the final diagnosis of low-grade fibromyxoid sarcoma (LGFMS) was made.

Distant staging with CT of the lungs showed one aspecific flattened pulmonary nodule not suspect for metastasis. The next follow-up CT was scheduled to take place in 6 months.

**Discussion:**

Low-grade fibromyxoid sarcoma (LGFMS) was first described by Evans in 1987 and is also known as Evans tumour [1]. It occurs most commonly in young adults and in deep tissues of the lower extremities and trunk.

LGFMS is a rare subtype of fibrosarcoma with a benign histological appearance but with a propensity for local recurrence (in 64 %) and metastasis (in 45 %). Long-term clinical follow-up and imaging surveillance is important because of the long latency periods between diagnosis and recurrence (ranging up to 15 years) and slow tumour growth. The most common distant metastatic site is the lung with the interval period to metastases ranging up to 45 years [2]. Treatment for patients with LGFMS is surgical resection, but even with successful resection, recurrence or metastases are possible. Up to now, there is no dedicated protocol regarding follow-up recommendations. In this case, clinical follow-up with CT of the chest for an extensive, if not lifelong period was planned. However, it is still unclear how regularly imaging of the chest should be repeated [3]. In our case, the interval was 6 months.

In 2012, Hwang et al. described the imaging features of LGFMS. Calcifications and bone erosions are uncommon
on radiographs. On ultrasound, LGFMS was solid with heterogeneous echogenicity. On CT, LGFMS tends to be hypodense to muscle with focal isodensity. LGFMS displays heterogeneous signal intensity on fluid-sensitive MR sequences, reflecting the histology of the tumour, which consists of fibrous (hypointense) and myxoid (hyperintense) tissue. Two patterns of signal distribution were observed: intralesional nodules with hyperintense signal and gyriform patterns [4].

Desmoid tumour is an important differential diagnosis of LGFMS. On MRI desmoid tumour typically shows linear extension along fascial planes (fascial tail sign) and hypointense bands across all sequences representing collagenized hypocellular bands. After contrast administration these collagen bands stand out in relation to the enhancing cellular areas of the tumour [5]. These MRI features of desmoid tumour are not present in our case. Histologically, the tumour contained striking bland fibroblasts in a fibromyxoid matrix. In many areas, the tumour had a swirling, whorled pattern (Fig. 6). MUC4, a highly sensitive and specific immunohistochemical marker for LGFMS, showed diffuse strong positive staining (Fig. 7) [6].

In conclusion, radiologic imaging plays an important role in the clinical management of LGFMS, from tumour detection to treatment follow-up. Certain specific patterns (intralesional nodules and gyriform pattern) on MRI can help differentiate LGFMS from other fibrous and myxoid tumours.

**Differential Diagnosis List:** Low-grade fibromyxoid sarcoma (LGFMS), Desmoid tumour (synonyms: aggressive/musculoaponeurotic fibromatosis), Ancient schwannoma with central necrosis, Myositis ossificans, Old haematoma

**Final Diagnosis:** Low-grade fibromyxoid sarcoma (LGFMS).

**References:**


Figure 1

Description: Swelling of the left thenar. Origin: Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium
Description: Coronal fat-suppressed proton density weighted image shows a tumour in the thenar muscle with hypointense margin (fibrous tissue) and hyperintense centre (myxoid tissue). The lateral part has a gyriform to lobulated shape (arrow). **Origin:** Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium
**Description:** Coronal T1 weighted image shows tumour in the thenar muscle with hypointense margin (fibrous tissue) and hyperintense centre (myxoid tissue). The lateral part has a gyriform to lobulated shape (arrows). **Origin:** Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium
Description: Coronal T1 weighted image after gadolinium contrast administration with fat suppression shows no enhancement in the central myxoid and peripheral fibrous component of the tumour. The lateral part has a gyriform to lobulated shape (arrows). Origin: Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium
**Figure 3**

**a**

**Description:** Axial fat-suppressed proton density weighted image shows the tumour in the thenar with thick peripheral fibrous (=hypointense) margin and myxoid centre. **Origin:** Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium

**b**

**Description:** Axial T1 weighted image with contrast shows the tumour in the thenar with thick peripheral fibrous (=hypointense) margin and myxoid centre. **Origin:** Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium
Description: Time intensity curve after bolus injection shows normal perfusion of artery (1), muscle (2), and no enhancement in the myxoid (3) and fibrous (4) component of the tumour. Origin: Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium
Description: Plain radiography of the left hand shows normal density of the thenar, without calcifications or ossifications. Origin: Verstraete K, Department of Radiology, UZ Gent, Gent, Belgium
Description: Histopathology of the low-grade fibromyxoid sarcoma. In a fibrous, collagenous background, bland fibroblastic tumour cells with minimal atypia are arranged in a storiform, swirling manner (Haematoxylin and Eosin staining, 200x magnification). Origin: Creytens D, Department of Pathology, UZ Gent, Gent, Belgium
Description: Diffuse and strong cytoplasmic staining of the spindled tumour cells for the immunohistochemical MUC4 staining, conforming the diagnosis of a low-grade fibromyxoid sarcoma (200x magnification). Origin: Creytens D, Department of Pathology, UZ Gent, Gent, Belgium