Desmoid tumours, also known as extra-abdominal aggressive fibromatosis (AF), are rare benign mesenchymal tumours, which are characterised histologically by proliferation of fibroblasts and myofibroblasts with marked production of intercellular collagen (1). Although benign, these tumours are locally aggressive and may cause local symptoms (2). We present a rare case of histologically proven extra-abdominal desmoid tumour, occurring in the calf, with ultrasound, MRI and PET-CT correlation. The purpose of this report is to discuss if metabolic imaging such as PET-CT is useful as additional tool in differentiation between malignant and benign soft tissue lesions (STT).

Case report

A 64-year-old male presented to our department with a painless hard lump at the posterior aspect of his left mid-calf. There was no history of trauma to this site and previous medical history was unremarkable. Ultrasound (US) demonstrated a predominantly solid hypoechoic lesion measuring 5.0x3.0 cm arising from the lateral gastrocnemius muscle. On power Doppler, peripheral vascularity was seen (Fig. 1).

Due to the solid and vascular appearance, the lesion was suspicious for a soft tissue sarcoma and hence MRI was subsequently performed for further characterisation. Axial T1-weighted images (WI) (Fig. 2) demonstrated an infiltrative left lateral gastrocnemius lesion with central areas of intermediate signal intensity (SI) and peripheral areas of low SI. There was local lesion extension within the soleus muscle and the medial gastrocnemius. On coronal fat-suppressed (FS) T2-WI (Fig. 3), the lesion displayed lobulated margins and heterogeneous SI with intrallesional high SI and bandlike areas of low SI. Sagittal (Fig. 4) and axial FS T1-WI (Fig. 5) after intravenous administration of gadolinium contrast showed marked heterogeneous enhancement of the lesion.

The bandlike areas of low SI on non-contrast images did not enhance. The superior margin of the lesion was irregularly delineated and infiltrative. Whole body PET-CT was performed to identify distant metastases. PET-CT demonstrated no other lesion apart from the lesion in the left calf. The lesion displayed a high standard-uptake value (SUV) of up to 5.0 (Fig. 6) indicating increased metabolic activity. The lesion was suspected to be malignant and was surgically resected. The final histopathological diagnosis was a desmoid tumour.

Discussion

Desmoid tumours or deep extra-abdominal fibromatoses are rare soft-tissue tumours (STT) (0.1% of all STT), arising from connective tissue or their overlying aponeurosis or fascia (1, 2).
polyposis coli (APC) gene on chromosome 5q22 (5). Risk factors for development of desmoid tumours include oestrogen dominant states—as in pregnancy and the use of the oestrogen contraceptive pill (6). The peak age of onset of disease is between 25-35 years of age (7). In our case, the patient’s age was atypical—he was 64 years old. The most common sites affected are the upper limbs (46%), lower limbs (31%) and trunk (23%) (2).

Ultrasound findings are usually nonspecific; desmoid tumours may appear irregular and ill-defined, mainly seen as solid hypoechoic masses. If they are large, the tumour may cause muscle weakness and can compress surrounding nerves thereby causing symptoms (3, 4).

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Derived from Greek, the word “desmos” describes a tumour which is band or tendon-like (2). Desmoid tumours are characterised by proliferation of benign fibrous tissue with infiltrative growth. Despite being benign with negligible propensity for metastasis, they are locally aggressive. They have a high recurrence rate of up to 87% following surgical resection, and thus may lead to surgical amputation in the long term. Desmoid tumours may cause muscle weakness and can compress surrounding nerves thereby causing symptoms (3, 4).

Desmoid tumours occur sporadically, but may be associated with concomitant familial adenomatous polyposis and Gardner’s syndrome. These findings suggest a genetic link with mutations of the adenomatous polyposis coli (APC) gene on chromosome 5q22 (5). Risk factors for development of desmoid tumours include oestrogen dominant states—as in pregnancy and the use of the oestrogen contraceptive pill (6). The peak age of onset of disease is between 25-35 years of age (7). In our case, the patient’s age was atypical—he was 64 years old. The most common sites affected are the upper limbs (46%), lower limbs (31%) and trunk (23%) (2).

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**Fig. 2.**— Axial T1-weighted image (WI). The lesion is denoted by white arrowheads. The central part of the lesion is of intermediate signal intensity (SI) compared to muscle. Note also the presence of bandlike areas of low SI (star). The lesion is predominantly located within the left lateral gastrocnemius muscle, but invades the soleus muscle and the medial gastrocnemius.

**Fig. 3.**— Coronal fat-suppressed (FS) T2-WI. The lesion (white arrowheads) is of heterogeneous signal with hyperintense areas and low SI bandlike intraslesional areas.

**Fig. 4.**— Sagittal FS T1-WI after administration of IV gadolinium contrast. There is marked enhancement of the lesion (white arrowheads).
may cause prominent posterior acoustic shadowing. It is likely that this observation is directly proportional to the amount of dense and compact collagen within the tumour (8). Peripheral neovascularity is another recognised feature (6).

MRI is the modality of choice for precise evaluation of local lesion extent. The lesion is usually located in close relationship with the muscle fascia and may invade the adjacent muscles. The lesion may either have well-defined or irregular infiltrative margins (9) or a combination of well-defined and irregular margins. Another morphological sign consists of linear extension along the muscle fascia, also known as the fascial tail sign (10). The SI is highly variable, depending on the amount of intralesional collagen and the cellularity of the lesion (10). Most desmoid tumours show a predominantly intermediate SI compared to muscle on T1-WI, with interspersed areas of low signal intensity. Tumour heterogeneity is even more pronounced on T2-WI. Lesions or parts of the lesions with low cellularity and high collagen content are of low SI, whereas high cellular or myxoid areas are of high SI on T2-WI. Low SI areas on T2-WI are usually bandlike (10). Particularly, this bandlike morphology may be helpful in the differential diagnosis with other lesions displaying a low T2 signal (such as pigmentated villonodular synovitis, granular cell tumour, fibrosarcoma and malignant fibrous histiocytoma). Post-gadolinium sequences typically show marked and heterogeneous enhancement (9). Hypocellular and collagenized bands do not enhance and are therefore often better seen on post-contrast MR images (10).

To the best of our knowledge, this is the first case report of a histologically proven desmoid of the calf with PET-CT correlation. There are, however, case reports of histologically proven desmoid tumour in other locations depicting a spectrum of FDG uptake, ranging from low to high (11–13). Low uptake in a lesion is likely to indicate presence of collagen fibres whilst high uptake is likely to correspond to high metabolic areas and areas of high cellularity with active mitosis (12).

Because of the local aggressive nature of the lesion, the surgeon should try to ensure wide surgical margins at this site to prevent recurrence.

In conclusion, PET-CT is not helpful as an additional tool for differentiation between malignant STT and lesions with a benign biologic behaviour, such as desmoids. Precise analysis of intraskeletal bandlike areas of low signal intensity on all pulse MR sequences and intimate relationship with the muscle fascia is more useful to the diagnosis of this rare soft tissue lesion.

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