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AGGRESSIVE FIBROMATOSIS: IS PET-CT USEFUL IN LESION CHARACTERIZATION?

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Deep extra-abdominal fibromatoses (desmoids) are rare benign fibrous mesenchymal tumours occurring in adults, which may mimic primary malignancy on imaging. We present a case of a 64-year-old man with a hard painless lump in his left calf. The solid and partially hypervascular appearance on ultrasound, the infiltrative appearance, lesion heterogeneity and heterogeneous enhancement pattern on Magnetic Resonance Imaging (MRI) were suspicious for a soft tissue sarcoma. Moreover, PET-CT demonstrated FDG-avidity. Despite these aggressive imaging features, histopathology revealed a benign but locally aggressive desmoid tumour. The radiologist should be aware that PET-CT is not always helpful as an additional tool for differentiation between malignant and benign soft tissue lesions. Intralesional bandlike areas of low signal intensity on all pulse MR sequences and intimate relationship with the muscle fascia are more useful clues to the diagnosis of this soft tissue lesion.

Key-word: Desmoid.

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 benign desmoids from malignant soft tissue tumours (STT).

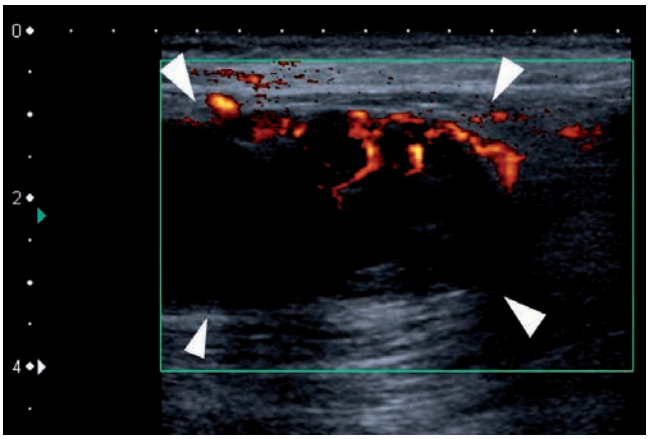

Fig. 1. — Ultrasound with power Doppler. A solid hypoechoic lesion (white arrowheads) with peripheral vascularity.

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 5.0x 3.0 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 intralesional 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 aggressive 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).

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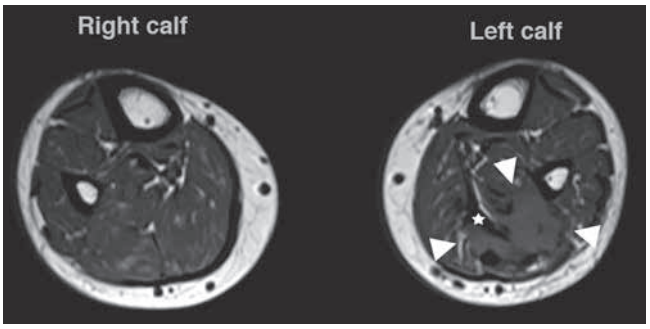

Fig. 2. — Axial T1-weighted image (WI). The lesion is denoted by white arrowheads. The central part of the lesion is of intermediate signal intensity (SI) compared to muscle. Note also the presence of bandlike areas of low SI (star). The lesion is predominantly located within the left lateral gastrocnemius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

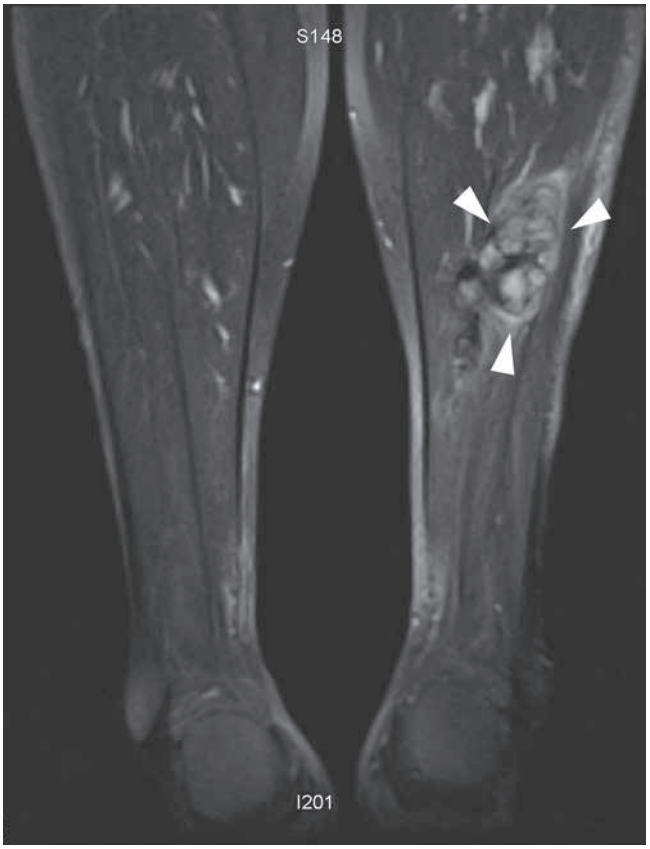

Fig. 3. — Coronal fat-suppressed (FS) T2-WI. The lesion (white arrowheads) is of heterogeneous signal with hyperintense areas and low SI bandlike intralesional areas.

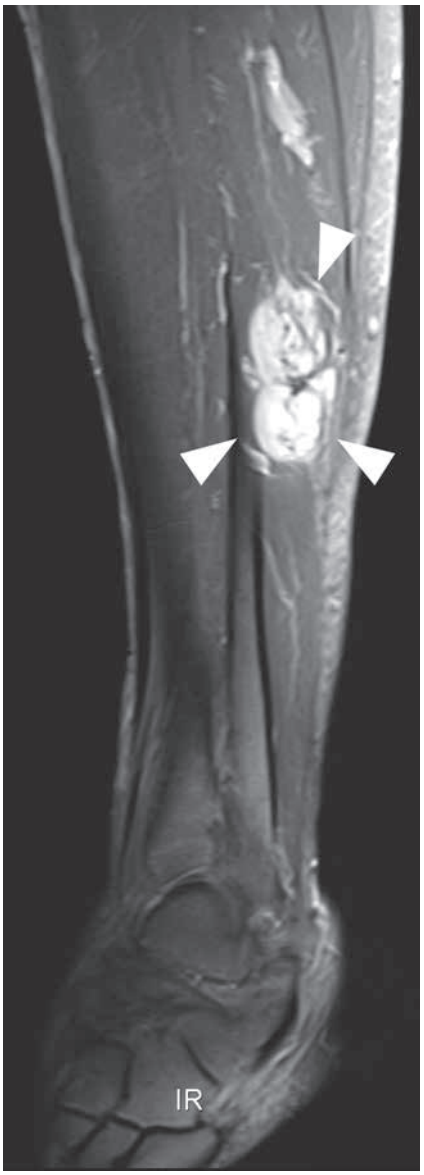

Fig. 4. — Sagittal FS T1-WI after administration of IV gadolinium contrast. There is marked enhancement of the lesion (white arrowheads).

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).

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

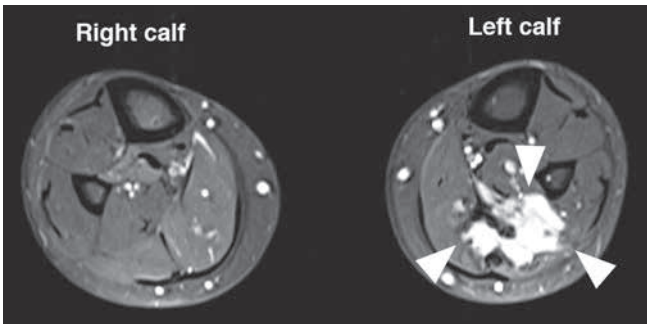

Fig. 5. — Axial FS T1-WI after administration of IV gadolinium contrast. There is marked enhancement of the infiltrative lesion (white arrowheads), except for the bandlike areas of low SI on other pulse sequences which do not enhance.

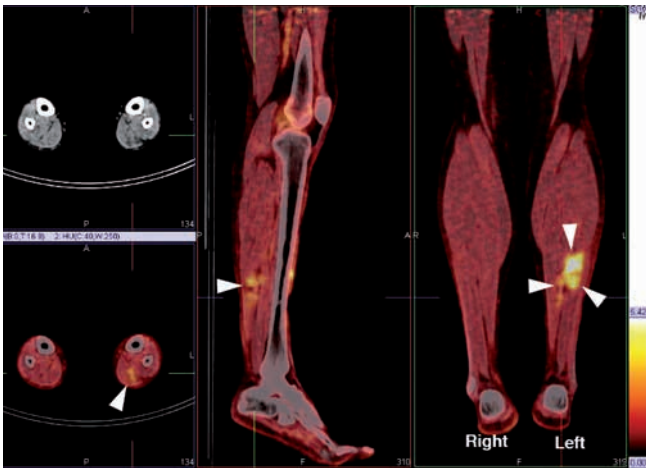

Fig. 6. — PET-CT. The focal lesion (white arrowheads) is FDG-avid with a Standard Uptake Value (SUV) of up to 5.0, indicating a metabolically active lesion.

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 pigmented 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 intralesional 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.

References

1. Weiss S.W., Goldblum J.R., Enzinger F.M.: Fibromatoses. In: *Enzinger and Weiss's soft tissue tumours*. 5th Ed. Edited by Weiss S.W., Goldblum J.R. Printed by Mosby Elsevier, Philadelphia, 2008, pp 227-256.
2. Karakousis C.P., Mayordomo J, Zografos G.C., Driscoll D.L.: Desmoid tumours of the trunk and extremity. *Cancer,* 1993, 72: 1637-1641.
3. Kabiri E.H., Al Aziz S., El Maslout A., Benosman A.: Desmoid tumours of the chest wall. *EurJ Cardiothorac Surg*, 2001, 19: 580-583.
4. Robbin M.R., Murphey M.D., Temple H.T., Kransdorf M.J., Choi J.J.: Imaging of musculoskeletal fibromatosis. *Radiographics*, 2001, 21: 585-600.
5. Clark S.K., Phillips R.K.: Desmoids in familial adenomatous polyposis. *Br J Surg,* 1996, 83: 1494-1504.
6. Erguvan-Dogan B., Dempsey P.J., Ayyar G., Gilcrease M.Z.: Primary desmoid tumour (extraabdominal fibromatosis) of the breast. *AJR Am J Roentgenol*, 2005, 185: 488-489.
7. Taylor L.J.: Musculoaponeurotic fibromatosis. A report of 28 cases and review of the literature. *Clin Orthop Relat Res*, 1987, 224: 294-302.
8. Fornage B.D.: The case for ultrasound of muscles and tendons. *Sem Musculoskel Radiol*, 2000, 4: 375-391.
9. Lee J.C., Thomas J.M., Phillips S., Fisher C., Moskovic E.: Aggressive fibromatosis: MRI features with pathologic correlation. *AJR Am J Roentgenol*, 2006, 186: 247-254.
10. Murphey M.D., Ruble C.M., Tyszko S.M., Zbojniewicz A.M., Potter B.K., Miettinen M.: From the archives of the AFIP: musculoskeletal fibromatosis: radiologic-pathologic correlation. *Radiographics*, 2009, 29: 2143-2176.
11. Souza F.F., Fennessy F.M., Yang Q., van den Abbeele A.D.: Case report. PET/CT appearance of desmoid tumour of the chest wall. *Br J Radiol*, 2010, 83(986): e39-42.
12. Basu S., Nair N., Banavali S.: Uptake characteristics of fluorodeoxyglucose (FDG) in deep fibromatosis and abdominal desmoids: potential clinical role of FDG-PET in the management. *Br J Radiol*, 2007, 80 (957): 750-756.
13. Maurer F., Horst F., Pfannenberg C., Wehrmann M.: Multifocal extra-abdominal desmoid tumour – diagnostic and therapeutic problems. *Arch Orthop Trauma Surg*, 1996, 115: 359-362.