Subcutaneous desmoid tumors: characteristic "sun-burst" appearance on MRI and ultrasound

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Aims and objectives

Desmoid tumors, also called aggressive fibromatoses, are rare and locally aggressive soft-tissue tumors that are characterized by fibroblastic proliferation, infiltrative growth and the tendency toward local recurrence [1]. They arise from the connective tissue of muscles, overlying fascia or aponeurosis, and from scar tissue after surgery. The term "desmoid" originates from the Greek "desmos" meaning band or tendon, in order to illustrate the band-like or tendon-like consistency of the lesion [2]. Histologically they consist of uniform spindle-shaped cells surrounded and separated by abundant collagen. The extent of cellularity and collagen is variable and on gross pathological examinations the lesions display whitish bands that represent more collagenized regions [3]. Generally the desmoid tumors are divided into extraabdominal, abdominal and intraabdominal, the latter often associated with Gardner syndrome (familial polyposis syndrome) [2, 4]. The typical locations of the extraabdominal desmoid tumors include the shoulder or upper extremity, gluteal region and lower extremity, chest wall or back and neck [1, 2, 5-7].

Most of the extraabdominal desmoid tumors involve the muscles. Subcutaneous desmoids are, in comparison to their intramuscular counterparts, uncommon forms of desmoids. Their imaging appearance has not been extensively described in the literature. The purpose of this study was to identify specific imaging characteristics of subcutaneous desmoids from magnetic resonance imaging (MRI) and high-resolution ultrasound (HRUS) examinations.
Methods and materials

We retrospectively examined 12 patients (9 females and 3 males between 14 and 51 years of age with a mean of 33 years) with histologically proven primary subcutaneous desmoids. Eleven patients received MRI examinations and nine were additionally examined with HRUS. In one patient only HRUS images were available. Because of the retrospective nature of the study the MRI and sonographic examinations were performed on different equipment. The minimum required MRI protocol for inclusion in the study contained a T1- weighted and a water sensitive (either T2-weighted or a STIR) sequence. All but one patient (who had Gardner syndrome and underwent renal transplantation) with MRI examination received intravenous gadolinium administration. Sonographic examinations performed using broadband linear arrays (up to 18 MHz).

All tumors were evaluated for location, muscular fascia involved in the development of the tumor ("fascial tail" sign [3]), compartment involved (i.e. the subcutaneous fat only and possible spread within the muscular compartment), single or multiple lesions, mean size, shape (i.e., stellar and/or lobulated), margins (i.e., well defined or irregular), extensions spread, amount of regions rich in collagen, and bone involvement. Furthermore, the MRI images were assessed for the signal intensity of the tumors compared to that of the adjacent muscle tissue on T1-weighted and on water sensitive sequences, the pattern of enhancement after intravenous gadolinium administration (i.e., homogeneous or inhomogeneous) and the degree of enhancement (i.e., minor, moderate or strong). The sonographic images were evaluated for the echogenicity relative to the muscle (i.e., hypoechoic, isoechoic or hyperechoic), echotexture (i.e. homogeneous or fibrillar), vascularity on color Doppler (i.e. hypervascular when more than 2 vessels or hypovascular when less than 2 vessels were found within a large color Doppler frame over each part of the tumor).

All examinations were scored by one radiologist (RI.M.) and reviewed by one experienced musculoskeletal radiologist (G.B.).
Results

All together there were 13 subcutaneous lesions: three each around the hip, along the flanks and chest wall/breast, two each around the shoulder and along the abdominal wall. All tumors were primary tumors. Three patients, including one with familiar polyposis, had multiple lesions, and in two of these patients additional intramuscular tumors were present. In four patients the lesions showed an extracompartmental spread, with a subcutaneous and intramuscular extent. Bone involvement was present in two lesions.

In eleven patients (91%) the lesions showed a stellar configuration with irregular margins. In the remaining patient the tumor presented with a polylobular form. All lesions displayed multiple irregular sun-burst-like extensions along the fascial planes and the septa of the subcutaneous fat tissue in each of the 3 orthogonal axes (Fig.1, 2, 3, 4, and 5). These extensions showed a substantial spread from the main tumor focus. In 50% of cases these extensions reached the cutis (Fig. 4). In eleven patients (91%) the tumors showed contact to a muscular fascia forming the fascial tail sign [3] (Fig. 2B, 4 and 5). The average mean diameter of the lesions, measured on the MRI images, was 3.76 cm and varied between 0.7 and 11.5 cm.

On MRI all lesions showed a hyperintense signal intensity compared to the skeletal muscle on the water sensitive sequences (either T2-weighted or a STIR) (Fig.1 A and B). The signal intensity on T1-weighted images was predominantly isointense to skeletal muscle (in 9 patients, Fig. 1C), while in one patient each the signal intensity of the lesions was either slightly hypo- or hyperintense to skeletal muscle. In 9 patients (81%) the lesions presented bands of low signal intensities on both T1- and water sensitive sequences (asterisk in Fig.1) that were previously shown to represent more collagenized regions [3, 8, 9]. After intravenous contrast administration (in 10 patients) all lesions showed a strong enhancement, except for the collagen rich areas (Fig. 1D). The pattern of enhancement was homogeneous in 9 patients and inhomogeneous only in one patient.

Nine patients received sonographic examination. All tumors were mainly hypoechoic to the subcutaneous fat (Fig. 2A). Areas with a fibrillar hyperechoic pattern within the tumors were found in 8 patients (Fig. 2B). It was previously reported that these regions show a high amount of collagen [10]. When both MRI and HRUS examinations were performed, in 6 patients the lesions that displayed the fibrillar pattern on HRUS also presented on MRI regions of low signal intensity (Fig. 3). Color Doppler sonography was performed in 7 patients and showed rich tumor vascularity in 3 patients (Fig. 2C) and poor vascularity in the remaining 4 patients.
Fig. 1: Subcutaneous desmoid in a 33-year-old woman with a 3-month history of a painful swelling in the right flank. The MRI examination shows a stellar soft-tissue mass with contact to the thoracolumbal fascia and ill-defined infiltrative sun-burst-like extensions along the septa of the subcutaneous fat layer. The signal intensity of the tumor is on the axial STIR (A) and T2-weighted (B) sequences intermediate, higher than that of the adjacent muscle and lower than that of fat and on the coronal T1-weighted sequence (C) muscle insointense. After intravenous gadolinium administration the tumor shows a strong enhancement (D). Note extensive regions of low signal intensity on all sequences (asterisk) that correspond to collagen rich areas.

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**Fig. 2:** Superficial desmoid in a 37-year-old female with a palpable, hard, and painful mass in the right shoulder. The panoramic HRUS scan (A) and the zoomed transversal scan (18 MHz) (B) demonstrate a hypoechoic irregular mass with contact and spread along the overlaying fascia of the deltoid and lateral head of the triceps (fascial tail sign, arrows) and multiple septal extensions between the fatty lobules (arrowheads). The Color Doppler sonography (C) shows multiple vessels at the periphery of the tumor and within the extensions. Asterisks indicate areas with hyperechoic fibrillar echotexture rich in collagen components.

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Fig. 3: Subcutaneous desmoid in a 51-year-old male with a history of a palpable, hard and since 1 year growing mass in the left hip. Axial STIR (A), T1-weighted postcontrast (B) MRI and corresponding panoramic HRUS (C) images of a superficial desmoid showing central low MRI signal intensity areas and matching hyperechoic fibrillar echotexture that correspond to collagen rich tumor components.
Fig. 4: Desmoid of the left shoulder in a 27-year-old female. Consecutive axial T1-weighted MRI images (A and B) and HRUS (C) showing a typical stellar infiltrating lesion within the subcutaneous fat with spread along the overlaying fascia of the lateral head of the triceps (fascial tail sign, arrows in B and C) and radiating long distance extensions that reach the cutis (arrowhead in A).

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**Fig. 5:** Same patient as in Fig. 3. Coronal T1-weighted (A), T1-weighted postcontrast (B) MRI and corresponding panoramic HRUS (C) images of a superficial desmoid displaying a wide contact to the iliotibial tract and a spindle-shaped fascial protrusion inferiorly (arrows).

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Conclusion

This study describes the HRUS and MRI imaging features of subcutaneous desmoids, based on a series of 12 patients.

In our series of subcutaneous desmoids the predominant signal intensity (similar to that of muscle on T1-weighted images and higher than that of muscle on the water sensitive sequences) and the enhancement pattern on MRI was in accordance with the previous published data about desmoid tumors in general [5, 6, 8, 11]. Also the hypoechoic signal compared to the adjacent muscle on sonography was unspecific. One major characteristic of the subcutaneous desmoids in our series was the irregular stellar appearance in each orthogonal plane with "sun-burst" extensions spreading long distances along the underlying fascia (fascial tail sign) and along the septa of the subcutaneous fat tissue, sometimes reaching until the cutis. It is believed that the fascial tail sign is very suggestive of extraabdominal desmoids [3, 12] and it was visualized with MRI as well as with sonography, if high frequency ultrasound probes were used [3, 10, 12, 13]. It is important to detect it in its entire extent in order to achieve a complete surgical resection. The spiculated appearance of the subcutaneous desmoids may be explained by the fact that the tumors are surrounded by loose connective tissue [10]. This stellar shape, unlike the signal intensity in MRI or echogenicity in HRUS, distinguishes the subcutaneous from the intramuscular desmoids, which tend to have a fusiform shape with spindle-shaped margins at the proximal and distal ends of the tumor when imaged parallel with the course of the muscle fibers [10].
References


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