Moving towards a more conservative approach of desmoid tumours: role of MR in the assessment of their aggressiveness and in their management

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Aims and objectives

Desmoid tumours (DT) are rare mesenchymal tumours with an incidence of 5-6/1,000,000/year.

They usually affect young women between 15 and 60 years-old, with a peak age of about 30-40 years-old. Most cases are sporadic although approximately 5-10 % arises in the context of familial adenomatous polyposis (FAP) and Gardner's syndrome, suggesting a link with mutations of the APC gene on chromosome 5q22.

They may classified in three types according to their location: intraabdominal, abdominal wall and extraabdominal. These last ones are located deep in the interfascial planes of musculature of the trunk and the extremities.

Histologically, they are benign tumours characterised by a monoclonal fibroblastic proliferation with marked production of intercellular collagen but they often present an unpredictable clinical course with an infiltrative growth pattern and a tendency to local recurrence, despite they lack any metastatic potential.

Since there is not an unified evidence-based approach for their management, a multidisciplinary and individualised approach is needed. However, a consensus treatment algorithm was proposed by the Soft Tissue and Bone Sarcoma group of experts of the European Organisation for Research and Treatment of Cancer (EORTC) and Sarcoma PATients EuroNet (SPAEN) where they stand for a more conservative approach with an initial watch & wait strategy. In this scenario, imaging techniques, especially MR, have a very important role.

Objectives:

# To highlight the importance of MR in the management of DT (diagnosis, response to treatment and detection of recurrences)

# To evaluate usefulness of functional sequences of MR, such as diffusion and dynamic contrast-enhanced sequences.

# To revise the role of MR in the new algorithms proposed.
Methods and materials

45 patients with biopsy-proven DT were diagnosed in a tertiary-level healthcare hospital (Vall d’Hebron, Barcelona, Spain) from 2002 to 2017. After excluding 3 patients with intraabdominal DT, 42 patients with extraabdominal and abdominal wall DT were included in our descriptive review.

MR was performed at diagnosis and during follow-up. 10 MR performed at diagnosis (23.8%) included functional sequences, so semiquantitative analysis of tumour perfusion with SI/T curves (signal intensity/time) could be performed.

Patient age and sex, treatment and outcome were recorded. Tumour location, size, shape and margins along with T1, T2 and STIR signal intensities and heterogeneity were assessed on MR at diagnosis.

Statistical evaluation was performed using the Kruskal-Wallis test or Fisher’s exact test for continuous quantitative and qualitative variables. Multiple linear regression analysis was calculated to identify factors that were independently predictive of aggressiveness of DT. SPSS software version 21 (SPSS, Inc., Chicago, IL). A two-tailed p-value <0.05 was considered statistically significant.
Results

42 patients with biopsy-proven deep DT were included in the study. 4 patients were referred for recurrent disease. They were predominantly women (75%) with a mean age of $36 \pm 18.8$ years-old, range from 12 to 86 years-old.

DT were categorized by location in extraabdominal (35) and abdominal wall (7) The extraabdominal group was subdivided in those located at the shoulder girdle (5), the pelvic girdle (3), the trunk and thoracic wall (18) and extremities (8), including one in the stump of a subcondylar amputation. We also included 2 patients with DT in the masticatory and posterior cervical spaces. No differences between groups were found by age or sex (Table 1 on page 8)

Most of DT were solitary. Only 2 patients (4.5%) had DT in two different locations simultaneously.

**MR characteristics at diagnosis (Table 2 on page 9)**

The median tumor size was 6.9 cm in diameter (ranged from 1 to 32.5 cm) and median tumour volume, 45.6 cc (ranged from 0.1 to 1.4 mL). Abdominal wall group included the largest tumours.

Most DT were ovoid (78%) with lobulated or irregular shape. Most DT combined portions with well-defined margins with other portions characterized by tentacle-like spiculated extensions with infiltrative growth (Figure 1 to 5)

82% of DT were either isointense or slightly hyperintense to skeletal muscle in T1WI, mostly homogeneous (80%). High signal intensity on T2WI was observed in 85% of DT, mostly heterogeneous (83%) because of the combination of collagen and fibroblast portions.

A specific sign of DT such as low signal bands of collagen across the tumour was present in 35% of patients (Fig. 1 on page 10, Fig. 2 on page 11, Fig. 3 on page 12, Fig. 4 on page 13, Fig. 5 on page 14)

Bone marrow and periosteal signal was normal in every case, despite the proximity of the DT to the bone.
All DT show moderate to avid contrast-enhancement and none of them present areas of necrosis in the post-contrast sequences.

Facilitated diffusion was observed in all DT with mean values on ADC maps of 1.30 ± 0.1 (ranged from 1.15 to 1.42) (Fig. 6 on page 15). This characteristic is very helpful in differential diagnosis with other soft-tissue tumours, especially sarcomas, that show restricted diffusion in almost all cases, exceptuating myxoid sarcomas.

Dynamic contrast-enhanced (DCE) MR sequences were performed in 10 patients at diagnosis (23.8%) DCE sequences proved to be an useful tool in diagnosis because of they show two characteristical intensity/time curves (Fig. 7 on page 16). Type II curves (progressive) was observed predominantly in collagen-rich portions of DT and type V curves (washin and washout) was observed predominantly in fibroblast-rich portions of DT.

**MR characteristics at response to treatment (Fig. 8 on page 17)**

Most patients were treated with surgery (80.9%) with adjuvant radiotherapy in selected cases with microscopic or macroscopic positive margins. Medical treatments (chemotherapy or hormonal-therapy) were used in 7.1% of patients. Only 12% of patients opted for a conservative approach of "watch and wait".

Conclusions about effectiveness of treatments couldn't be drawn.

**MR characteristics at progression or recurrence (Table 3 on page 8)**

Recurrence was observed in 10 patients (23.8%) with a median disease free survival of 19 months (ranged from 5 to 43 months)

Logistic regression analysis including age, sex and tumour location, size, margins, signal intensity and diffusion was performed. None of the parameters showed a statistically major risk of recurrence. However, we observed a tendency to an earlier recurrence in larger tumours.

9 patients had local recurrence (Fig. 9 on page 18, Fig. 10 on page 19, Fig. 11 on page 20). Only 1 patient had a distal recurrence in the left calf (Fig. 12 on page 21), caused probably by direct spread of the tumour through interfascial plane rather than distant metastasis.
Role of MR in the consensus treatment algorithm proposed by EORTC and SPAEN (Fig. 13 on page 22)

DT are slow-growing tumours that tend to progress in first 1-2 years followed by a long period of stability and a late phase of involution.

Multiple treatment alternatives have been proposed, with variable results, being surgery with or without radiotherapy the classical adopted option along with medical treatment such as tamoxifen, NSAIDs... New treatments based on cryoablation, radiofrequency or immunotherapy have been proposed in recent years.

Several studies concluded that microscopic or macroscopic margins don't influence in the risk of recurrence, so a more conservative approach based on functionality and cosmetic rather than radical surgery is proposed.

Several studies concluded that "watch and wait" strategy have no differences in risk of progression with classical medical treatments in selected cases.

Based on the natural history of the DT and absence of unified evidence-based approach for their management, EORTC and SPAEN proposed a consensus treatments algorithm where imaging techniques, especially MR, has a pivotal role.

After biopsy-proven diagnosis, they propose a follow-up period of 1-2 years for the asymptomatic DT, which include a clinical evaluation and MR performed every 3 months in order to detect fast-growing or more aggressive DT.

If stability is proved, "watch and wait" strategy may continue after 2 years, increasing the time interval of MR (every 6 months between 2 to 5\textsuperscript{th} year and every 12 months after 5\textsuperscript{th} year)

Progression is defined as growth or changes in signal intensity in two consecutives MR.

No conclusions regarding the paper of DCE has been proposed, although we found that it could be an useful tool to characterised better DT and select those with higher risk of progression.
**Table 1:** DT distribution according to sex, age and localization.

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### Table 3: MR characteristics at diagnosis

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<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Size (cm)</th>
<th>Initial treatment</th>
<th>Surgical margins</th>
<th>Recurrence size (cm)</th>
<th>DFS (months)</th>
<th>Recurrence treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>29</td>
<td>Female</td>
<td>Pelvic girdle</td>
<td>1.5</td>
<td>Surgery</td>
<td>-</td>
<td>0.9</td>
<td>5</td>
<td>RT</td>
<td>Local recurrence</td>
</tr>
<tr>
<td>37</td>
<td>Female</td>
<td>Cervical</td>
<td>4</td>
<td>Surgery</td>
<td>-</td>
<td>3</td>
<td>43</td>
<td>Surgery + RT</td>
<td>CR</td>
</tr>
<tr>
<td>37</td>
<td>Female</td>
<td>Pelvic girdle</td>
<td>4</td>
<td>Surgery</td>
<td>-</td>
<td>3</td>
<td>30</td>
<td>Surgery</td>
<td>CR</td>
</tr>
<tr>
<td>41</td>
<td>Female</td>
<td>Trunk</td>
<td>3.8</td>
<td>Surgery</td>
<td>-</td>
<td>3</td>
<td>26</td>
<td>Surgery</td>
<td>CR</td>
</tr>
<tr>
<td>64</td>
<td>Female</td>
<td>Abdominal wall</td>
<td>2.5</td>
<td>Surgery</td>
<td>-</td>
<td>2.2</td>
<td>31</td>
<td>Surgery</td>
<td>CR</td>
</tr>
<tr>
<td>42</td>
<td>Female</td>
<td>Triceps</td>
<td>3</td>
<td>Surgery</td>
<td>+</td>
<td>3</td>
<td>17</td>
<td>Surgery</td>
<td>CR</td>
</tr>
<tr>
<td>28</td>
<td>Female</td>
<td>Calf</td>
<td>9</td>
<td>Surgery</td>
<td>+</td>
<td>13</td>
<td>5</td>
<td>RT</td>
<td>Distal Recurrence</td>
</tr>
<tr>
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<td>Female</td>
<td>Scapular girdle</td>
<td>13</td>
<td>Surgery</td>
<td>+</td>
<td>3</td>
<td>15</td>
<td>Surgery + RT</td>
<td>CR</td>
</tr>
<tr>
<td>48</td>
<td>Female</td>
<td>Abdominal wall</td>
<td>4</td>
<td>Surgery</td>
<td>+</td>
<td>12</td>
<td>12</td>
<td>RT</td>
<td>CR</td>
</tr>
<tr>
<td>12</td>
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<td>Surgery</td>
<td>+</td>
<td>13</td>
<td>13</td>
<td>Medical</td>
<td>CR</td>
</tr>
</tbody>
</table>

**Table 2:** Demographic and MR characteristics of recurrent DT. DFS: Disease free survival. RT: Radiotherapy

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**Fig. 1:** 26 years-old woman with abutting and painful tumour in the right calf. Axial T1WI (a), and axial (b) and sagittal (c) PD show a well-defined lesion slightly hyperintense on T1WI and heterogeneously hyperintense on PD because of the combination of collagen-rich portions (hypointense) and fibroblast-rich portions (hyperintense). Notice the split fat sign (arrow in a), the band sign (arrow in b) and the tail sign (arrow in c)

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Fig. 2: 12 years-old girl with abutting and painful tumour in the left forearm. Axial T1WI (a), PD (b) and T1WI with fat suppression (c) show an ill-defined lesion in the anterior compartment of the left forearm with infiltrative pattern and extension to the subcutaneous space. It appears slightly hyperintense lesion on T1WI, hyperintense on PD with a band sign (asterisk in b) and avid and homogeneous contrast-enhancement. On ultrasonography, it appears as a well-defined hypoechoic lesion without vascularity on colour Doppler.

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**Fig. 3:** 18 years-old woman with paravertebral DT, hypointense on T1WI (asterisk in a) and heterogeneously hyperintense on PD (asterisk in b)

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**Fig. 4:** 30 years-old woman with DT that appears as a diffuse and irregular thickening of the fascia that envolves trapezious, rhomboid and levator scapulae muscles. It shows low signal intensity both on T1WI and PD (arrows in a and b)

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Fig. 5: 44 years-old woman with an abdominal wall DT that appears as a well-defined lesion in the aponeurosis of the rectus abdominis muscle, hypointense on T1WI (a), heterogeneous on T2WI (b) with moderate contrast-enhancement (c)

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**Fig. 6:** Diffusion and ADC maps of a DT that appears with facilitated diffusion in all cases (1.30 ranged from 1.15 to 1.42). This characteristic is important in the differential diagnosis with other soft-tissue lesions, especially sarcomas.

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Fig. 7: 30 years-old woman with DT in the right lumbar space. It presents a cranial and lateral collagen-rich portion, hypointense on PD (red arrow) and a caudal and medial fibroblast-rich portion, heterogeneously hyperintense on PD (green arrow). DCE (c) and intensity/time curves (d) show different type of curves for both portions. Collagen-rich portion presents a type 2 curve (progressive) and fibroblast-rich portion presents a type 5 curve (washin and washout)

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Fig. 8: 14 years-old girl with DT treated with vinblastine and methotrexate for 6 months. After treatment, a decrease in signal intensity on PD represents the reduction of fibroblast-rich portion of the tumour, substituted by collagen and fibrosis.

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**Fig. 9:** Same patient as figure 7, presented a regression of the cranial and lateral portion of the DT (1) and a progression of the caudal and medial portion of the DT (2) with increasing the size and signal intensity overtime. DCE and intensity/time curves at the diagnosis and the progression share similar characteristics (type 5 curve) (green arrow)

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Fig. 10: 47 years-old woman with local recurrence of DT at peridiaphisary middle third of the left thigh. It appears hypointense on T1WI, heterogeneously hyperintense on PD with band sign (green arrow). Notice muscular edema of the anterior compartment muscles representing denervation.

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Fig. 11: 40 years-old man with multifocal recurrence of DT after supracondylar amputation. Recurrence was located in the stump (asterisks in a) with extra compartmental and subcutaneous extension, the popliteal fossa (asterisk in b) and the scalp (not shown)

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**Fig. 12:** Same patient as figure 1 with distal recurrence of DT because of spread through the interfascial plane, although rare hematogenious spread has been described in literature. Distal recurrence shares same characteristics on morphological sequences and DCE type of curve.

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Fig. 13: EORTC and SPAEN proposed a consensus management algorithm. Watch and wait strategy (1) after biopsy-proven diagnosis is based in a MR performed every 3 months during first 1-2 years, every 6 months between 2 and 5 years and every 12 months after the 5th year. Progression (2) is defined by the increase of size or change in signal intensity in two consecutive MR.

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Conclusion

# MR is important in management of DT: diagnosis, local staging, response to treatment and detection of recurrences.

# Signal intensity on T2WI correlates with distribution of fibroblast and collagen in anatomopathological samples.

# Diffusion sequences helps in differentiating DT from other soft-tissue tumours, especially sarcomas.

# DCE sequences are useful in better characterization of DT and selection those with higher risk of progression.

# Management of DT has evolved towards a more conservative approach ("watch and wait"), so importance of MR increases.


