Adiposis dolorosa (Dercum’s disease): MRI and ultrasound appearances

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AIM: To describe ultrasound and magnetic resonance imaging (MRI) features of adiposis dolorosa, Dercum’s disease, and to evaluate the MRI features prospectively against a large number of MRI examinations.

MATERIALS AND METHODS: Institutional review board approval for this study was obtained. The imaging features at MRI and ultrasound of 13 cases of adiposis dolorosa (nine female, four male; age range 32–72 years) were reviewed. MRI findings typical for adiposis dolorosa were proposed and prospectively evaluated on 6247 MRI examinations performed over a period of 8 months.

RESULTS: Adiposis dolorosa demonstrates multiple, oblong, fatty lesions in the superficial subcutaneous fatty tissue. They are mostly <2 cm in long axis diameter. They demonstrate nodular (“blush-like”) increased fluid signal at unenhanced MRI and are markedly hyperechoic at ultrasound. There is no contrast medium enhancement at MRI and no increased Doppler signal at ultrasound. Most lesions were clinically asymptomatic, some were painful/tender. There was no imaging evidence of oedema or inflammation. During prospective validation of these MRI features on 6247 MRI examinations, two cases with typical imaging features were encountered; both were diagnosed as adiposis dolorosa on clinical review. All cases of adiposis dolorosa showed these imaging findings. This results in a very low likelihood that a nodular, blush-like appearance of subcutaneous fat on MRI is not due to adiposis dolorosa.

DISCUSSION: Adiposis dolorosa, Dercum’s disease, should be suggested in the presence of multiple (many) small, oblong, fatty lesions in the subcutaneous fatty tissue in adult patients if they are hyperechoic on ultrasound imaging or blush-like at unenhanced MRI; typically a small number of these lesions are tender/painful. Imaging does not demonstrate inflammation or oedema in relation to these lesions. These MRI features should suggest the diagnosis and are likely to be pathognomonic. The radiologist is often the first to suggest the diagnosis based on the imaging features.

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Introduction

Adiposis dolorosa, Dercum’s disease, is a complex, incompletely understood disorder of subcutaneous fat. The World Health Organization has recognized it as a distinct disease entity and classes it as lipomatosis not elsewhere classified (ICD-10, IV, E88.2). Adiposis dolorosa is also...
referred to as Dercum’s syndrome, reflecting the lack of clear definition and aetiology.

Patients typically present with painful, subcutaneous lumps; further investigations show these to be fatty lesions. The pain can be severe and there is often associated obesity, generalized aches and fatigability, and a wide range of mental disturbance. There is symptomatic overlap to the fibromyalgia syndrome, although the aetiology is thought to be different.\(^1\)\(^–\)\(^3\) It is unclear whether psychological problems are primary symptoms of this disorder or secondary to chronic pain and obesity.\(^3\)\(^–\)\(^5\)

It affects women more frequently than men and often occurs perimenopausally. There is a definite hereditary element, which is presumed to be autosomal dominant with incomplete penetrance, although often the presentation is thought to be due to a new mutation, and therefore, there is no relevant family history.\(^1\)\(^–\)\(^4\)\(^,\)\(^6\)\(^–\)\(^9\)

It is unclear whether the painful lesions in adiposis dolorosa have defining histological features. Some studies find the lesions histologically identical to “normal” lipomas.\(^8\) Other studies suggest inflammatory changes and angiolipoma-like change.\(^4\) There is evidence to suggest that although there is mild inflammation, this is no more pronounced in obese controls and, therefore, not specific.\(^10\)

The painful lesions have been reported anywhere in the subcutaneous fat, although there is said to be a preponderance for the lower extremities and the lower area of the trunk of the body. Nevertheless, presentation even in the skull vault has been described.

The literature on adiposis dolorosa is, in parts, contradictory, and this disorder plays little to no role in the imaging literature. However, as the authors’ awareness of this disorder grew, it became clear that it is often the radiologist who first raises the possibility of this disorder, in particular when assessing patients with painful soft-tissue lumps. The authors have collected cases of proven Dercum’s disease to review the imaging features of this disorder. In a second step, these imaging features were prospectively evaluated on all magnetic resonance imaging (MRI) examinations performed at the authors’ institution to assess whether these findings are specific for Dercum’s disease.

### Materials and methods

For the first part of the study the authors reviewed imaging features of known cases of Dercum’s disease. These had been collected over a period of 3 years, from May 2008

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**Table 1**

Cases of clinically confirmed adiposis dolorosa listing the areas investigated with imaging.

<table>
<thead>
<tr>
<th>Age (years) at presentation</th>
<th>Examination type</th>
<th>Examination area</th>
<th>Number of lesions</th>
<th>Maximum size (mm)</th>
<th>Blush-like appearance on MRI</th>
<th>US signal echogenicity</th>
<th>Best MRI sequence</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>37</td>
<td>MRI</td>
<td>Lower calf</td>
<td>None</td>
<td>NA</td>
<td>Iso</td>
<td>hyper</td>
<td>T1, FS</td>
<td>Atypical large lesion, spindle lipoma</td>
</tr>
<tr>
<td>48</td>
<td>MRI</td>
<td>Lspine retro</td>
<td>Few</td>
<td>8</td>
<td>Yes</td>
<td>T1</td>
<td>PD FS</td>
<td>Hyper</td>
</tr>
<tr>
<td>US</td>
<td>MRI</td>
<td>Upper back</td>
<td>One</td>
<td>13</td>
<td>Yes</td>
<td>PD FS, T1</td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>38</td>
<td>US</td>
<td>Thighs, arm</td>
<td>Few in each area</td>
<td>18</td>
<td>NA</td>
<td>Iso, hyper</td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>65</td>
<td>MRI</td>
<td>Shoulder</td>
<td>18</td>
<td>Yes for typical lesions</td>
<td>Hyper</td>
<td>Hyper</td>
<td>MRI</td>
<td></td>
</tr>
<tr>
<td>32</td>
<td>US</td>
<td>Abdomen/chest wall</td>
<td>Many</td>
<td>25</td>
<td>Hyper</td>
<td></td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>42</td>
<td>MRI</td>
<td>Whole spine</td>
<td>Several</td>
<td>11</td>
<td>Yes</td>
<td>Str</td>
<td>FS</td>
<td></td>
</tr>
<tr>
<td>US</td>
<td>MRI</td>
<td>Thighs</td>
<td>Many</td>
<td>8</td>
<td>Yes</td>
<td>FS</td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>US</td>
<td>MRI</td>
<td>Thighs</td>
<td>Many</td>
<td>30</td>
<td>Hyper</td>
<td></td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>66</td>
<td>US</td>
<td>Shoulder, thighs</td>
<td>Many</td>
<td>11</td>
<td>Hyper</td>
<td></td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>72</td>
<td>US</td>
<td>Back</td>
<td>Several</td>
<td>20</td>
<td>Hyper</td>
<td></td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>72</td>
<td>US</td>
<td>Groin, flank</td>
<td>Several</td>
<td>17</td>
<td>Hyper</td>
<td>Str</td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>62</td>
<td>MRI</td>
<td>Abdomen, pelvis</td>
<td>Few</td>
<td>4</td>
<td>Yes</td>
<td>FS</td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>47</td>
<td>US</td>
<td>Chest, abdomen wall, back, arms, legs</td>
<td>Many</td>
<td>20</td>
<td>Hyper</td>
<td>Hyper</td>
<td>FS</td>
<td>Hyper</td>
</tr>
<tr>
<td>32</td>
<td>US</td>
<td>Thigh, arm</td>
<td>Many</td>
<td>20</td>
<td>Hyper</td>
<td></td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>36</td>
<td>MRI</td>
<td>Back, arm, thigh, pelvis</td>
<td>Many</td>
<td>40</td>
<td>Yes</td>
<td>Str</td>
<td>Angiolipoma, spindle cell lipoma</td>
<td></td>
</tr>
<tr>
<td>US</td>
<td>MRI</td>
<td>Lower back</td>
<td>Two</td>
<td>12</td>
<td>Yes</td>
<td>Str</td>
<td>Hyper</td>
<td></td>
</tr>
<tr>
<td>US</td>
<td>MRI</td>
<td>Lower back</td>
<td>Several</td>
<td>10</td>
<td>Hyper</td>
<td></td>
<td>Hyper</td>
<td></td>
</tr>
</tbody>
</table>

In the number of lesions section few = 2–5, several = 5–15.

FS (fat saturation) stands for proton density (PD) or T2-weighted FS or short tau inversion recovery (STIR) sequence. Where the STIR sequence was better than the PD FS or T2 FS sequence this was indicated. The lesion with histological result was larger and more atypical in appearance than the more typical lesions.

MRI, magnetic resonance imaging; ultrasound ultrasound; Hyper, hyperechogenicity.

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to June 2011. The diagnosis was based on assessment by a physician with experience in diagnosis and treatment of this disorder. It is based partly on typical clinical features, in particular, the presence of multiple, painful, fatty soft-tissue lesions and partly on exclusion of other disease entities. The presence of multiple but pain-free lipomas was not regarded as presentation of adiposis dolorosa.

The imaging features, and if present histological features, of the cases of Dercum’s disease were reviewed. Ultrasound examinations were performed on a Toshiba Aplio with a 12 MHz compound transducer. MRI was performed on a Siemens Avanto MRI system using surface coils. Routinely, T1-weighted sequences were performed in two planes, and short-tau inversion recovery (STIR) and proton-density fat-saturated (PD-FS) sequences in one plane each.

All imaging cases were initially collected prospectively, and for the study analysed retrospectively by the first author. The examinations were first reported by the following authors B.T., V.C.P., R.L., J.S., P.T., who are all subspecialty trained musculoskeletal radiologists.

For the prospective part of the study, the authors set out to test the hypothesis that the presence of multiple, largely round or oblong areas of increased water signal in the subcutaneous fat is diagnostic of Dercum’s disease by prospectively evaluating all MRI images performed at their institution for the presence of this feature. All MRIs performed in the authors’ institution are reviewed by a consultant radiologist. Before the beginning of the study, all consultant radiologists were briefed about the MRI features of notable lesions.

The presence of multiple areas of largely round areas of increased water signal in the subcutaneous fat was to be noted. These resemble an ill-defined nodular lesion on unenhanced imaging. The signal is increased in water-sensitive sequences, such as PD-FS, T2-weighted FS, or STIR images, and the signal is decreased in T1-weighted sequences.

The observations were structured in two distinct parts. In part 1 of the prospective part of the study, all MRIs performed from 1 August 2011 to 31 March 2012 performed for whatever indication and of whatever area were reviewed for the presence of the MRI features described above. In cases where the imaging features were suggestive of adiposis dolorosa, clinical review by a clinician experienced in this disorder was undertaken.

In part 2 of the prospective part of the study, all cases with an already known diagnosis of adiposis dolorosa presenting in the study period were to be reviewed for the presence of the previously described imaging features.

Institutional review board approval was obtained for this study.

Results

The results for part 1 of the study, the imaging features seen in known cases of Dercum’s disease are as follows: 13 patients with Dercum’s disease were identified in the study period. The findings are summarized in Table 1. Nine patients were female, four male. The age range was 32–72 years. The older patients (older than 40 years) had symptoms for many years, whereas the younger patients presented with newly symptomatic painful lumps. In 11 cases the diagnosis was first suggested by a radiologist. In all these cases, the patient presented with a painful soft-tissue lesion. In two cases, the diagnosis of adiposis dolorosa was known already, and the patients were sent for the assessment of painful lesions.

Nine patients underwent MRI examinations and 11 patients underwent ultrasound examinations. Two patients underwent only MRI imaging and four patients underwent only ultrasound. The other seven patients underwent MRI and ultrasound.

In all patients the lesions were centred in the superficial subcutaneous fat and not in the deep subcutaneous fat (Figs 1–4). The lesions were far more numerous than could be appreciated clinically, most of the lesions visible at imaging were clinically asymptomatic. Most lesions measured <2 cm in diameter and were oblong with the long axis parallel to the skin. All lesions were strongly hyperechoic at ultrasound and did not demonstrate flow on power or colour Doppler imaging (Fig 1). The lesions demonstrated a blush-like appearance on unenhanced MRI with decreased T1-weighted signal and increased signal in water-sensitive sequences such as STIR, T2-FS, or PD-FS-weighted imaging.
Larger lesions were more inhomogeneous in appearance at MRI (Fig 3), and generally, symptomatic lesions were larger than the majority of lesions seen. Larger and heterogeneous lesions were only seen in patients with very extensive disease, most lesions were small and more typical in appearance (Fig 3). There were no other imaging differences between symptomatic and asymptomatic lesions on ultrasound or MRI.

There was no oedema in or around any of the lesions (symptomatic or not) seen on ultrasound. In at least one case ultrasound demonstrated more lesions than MRI (Fig 4).

One lesion was surgically removed; the histological diagnosis was that of an angiolipoma. One lesion was biopsied; the histological diagnosis was spindle cell lipoma.

Based on this the presence of multiple lesions in the subcutaneous fat with blush-like nodular increased fluid signal at MRI was defined as suggestive of Dercum’s disease. For the prospective part of the study, all MRI examinations performed at the authors’ institution were assessed for these features over a period of 8 months. During this period, 6247 MRI examinations were assessed; most examinations were performed for musculoskeletal indications. Two cases of imaging features suggestive of Dercum’s disease were encountered. In both cases the diagnosis of adiposis dolorosa was first suggested by the radiologist.

Patient 1 was a 31-year-old man with multiple painful lumps in the subcutaneous fat of his arms, trunk, and lower limbs, which had typical MRI features. There was a family history of similar lesions in the patient’s mother and three brothers.

Patient 2 was a 74-year-old woman without a family history, but with multiple painful subcutaneous lesions. MRI demonstrated scattered, small, ill-defined, nodular or blush-like lesions in the subcutaneous fat of the frontal and dorsal abdominal wall. Both patients were classed as having adiposis dolorosa after clinical review.

There was no other case where the ill-defined nodular/blush-like features of the subcutaneous fat were seen. There were no cases of new clinical diagnosis of adiposis dolorosa; therefore, there were no cases in part 2 of the prospective part of the study. In combination, this makes it very likely...
that the imaging findings discussed were specific for adiposis dolorosa.

**Discussion**

Adiposis dolorosa, Dercum’s disease, is probably under-appreciated by most doctors, including radiologists. Although rare (the incidence is unclear), this disorder should be considered in the differential diagnosis when painful or tender subcutaneous fatty lumps are encountered. Dercum’s disease presents with multiple lesions, most of which are asymptomatic. The reason for the painful nature of the lipomatous lesions is not clear. Pressure on adjacent nerves, oedema, inflammation and autoimmune problems all have

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**Figure 3** A 32-year-old man with multiple lesions of the arms, legs, and trunk. Serial axial images of the upper arm (a, top: PD-FS, bottom: T1-weighted) and the thigh (b, top: PD-FS, bottom: T1-weighted) show multiple subcutaneous lesions, the larger lesions are more inhomogeneous in appearance but still positioned in the superficial subcutaneous fat. A chest wall lesion is noted on the axial images of the arm (a, arrow, top left image). Larger lesions are more likely to be clinically symptomatic. Sagittal STIR images help to appreciate the multitude of lesions (c, arm), (d, thigh, STIR and T1-weighted imaging). Very extensive disease, such as in this case, is less common than the more scattered lesions as seen in Fig 2.

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been suggested though none has been conclusively proven. It is unclear whether the painful lesions in adiposis dolorosa have defining histological features. Some studies found that the lesions were indistinguishable from "normal" lipomas.\textsuperscript{8,10} Other studies suggest inflammatory changes and angiolipoma-like change,\textsuperscript{10} and it has also been suggested that there are subtypes of adiposis dolorosa with different histopathological features.\textsuperscript{4} In the present study, there were two cases with histology demonstrating angiolipoma in one and spindle cell lipoma in the other. A proliferation of vessels and distortion of septa could explain the imaging features at MRI and ultrasound.

Figure 4 A 42-year-old woman with multiple lesions of the thigh. Axial MRI (a, top: PD-FS, bottom: T1-weighted) show typical lesions. Extended-view ultrasound (b) demonstrates a much higher lesion number than seen at MRI.

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All cases of Dercum’s disease showed very similar imaging features. The lesions were immediately subcutaneous, oblong rather than round, with the long axis parallel to the skin surface. Most lesions were <2 cm in maximum diameter. At ultrasound they were hyperechoic and did not show increased flow on Doppler imaging. At MRI they showed nodular increased fluid signal, better appreciated on water-sensitive sequences such as STIR, T2-FS or PD-FS-weighted imaging rather than on T1-weighted sequences. Larger lesions were more inhomogeneous at MRI. The appearances resembled a vascular “blush” on unenhanced images. No contrast medium enhancement was seen.

The sonographic findings in adiposis dolorosa differ from those of simple lipomas, which are often larger and not as hyperechoic, and although they can be multiple, they are not as numerous as seen in typical cases of Dercum’s disease. Simple lipomas are also not restricted to the superficial fat layer but occur anywhere in the subcutaneous fat. Simple lipomas per se are not tender or painful, although they can cause local discomfort. The sonographic appearance of the lesions in Dercum’s disease have been recently described in histologically proven cases of angiolipomas.\(^\text{11}\) In the study of Bang et al.\(^\text{10}\) most lesions were isolated. Simple lipomas demonstrate only fat signal at MRI, unlike the lesions in adiposis dolorosa disease. The MRI appearance is consistent with that of angiolipomas. Contrast medium enhancement is not part of the routine imaging protocol for soft-tissue lesions in the authors’ institution. However, contrast medium was given in one case, but the lesions did not show significant enhancement.

Only a small number of the visible lesions are clinically asymptomatic. In more severe cases, patients may have more than 100 lesions, most are clinically asymptomatic. Symptomatic lesions were larger than the majority of lesions on ultrasound, and larger lesions were more heterogeneous on MRI. There was no oedema in or around any of the lesions (symptomatic or not) seen on ultrasound.

This is contrary to the hypothesis that the lesions are associated with or the pain is due to inflammation and oedema; in the cases examined not a single lesion or perilous tissue demonstrated oedema or increased vascularity or other evidence of inflammation. This is supported by a recent histological study, which also found no undue inflammatory change.\(^\text{10}\)

Publications showing imaging examples of Dercum’s disease are very sparse and the features seen in a reported case report\(^\text{6}\) are in keeping with simple lipomas and are fundamentally different to the features seen in all cases of adiposis dolorosa presented here.

In the prospective part of the study, two cases of Dercum’s disease were suggested based on the imaging findings; the diagnosis was confirmed on clinical review. No case of Dercum’s disease not showing these imaging features was encountered. This indicates that the combination of multiple subcutaneous fatty lesions, hyperechoic on ultrasound, blush-like, nodular, increased fluid signal on unenhanced MRI, with most lesions being asymptomatic and some being tender, is very suggestive and possibly pathognomonic for adiposis dolorosa, Dercum’s disease.

In the published literature and the authors’ experience, including the cases presented here, the lesions occur mainly in the superficial subcutaneous tissue of the trunk, the upper arms, and the thighs.

In summary, adiposis dolorosa, Dercum’s disease, presents with multiple fatty subcutaneous lumps, only some of which are painful or tender. On imaging there are many small, oblong lesions in the superficial subcutaneous fatty tissue of trunk, arms, and legs. They are hyperechoic on ultrasound and ill-defined nodular/blush-like lesions on unenhanced MRI. These lesions are not associated with any inflammatory change on imaging. The radiologist is often the first to suggest the diagnosis.

References