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Dercum's disease (DD) was described by Dr. Francis Xavier Dercum at the University of Pennsylvania over a century ago. The hallmark of DD is painful lipomas in subcutaneous adipose (fat) tissue (SAT). Women are more affected than men (5-30:1). The average age of diagnosis is 35 years.<sup>1</sup>

**Lipomas:** The presence of small lumps in SAT is a requirement for diagnosis. Types of Dercum's SAT is as follows:<sup>2</sup>

Diffuse: The lipomas can be small, the size of a pea, and diffusely affect the majority of SAT; best palpated by rolling the fingers over the fat tissue. Consider initial exam in the cubital areas and medial knee.

Nodular: Lipomas can be the size of a marble, walnut, fist, or larger, localized primarily on the arms, anterior rib cage, abdomen, low back, buttocks and thighs.

Mixed: There often is a mixed picture of diffuse and nodular lipomas.

Lipomas are non-encapsulated and may contain excess connective tissue<sup>3</sup> or are angioliipomas.

**Vasculature:** The local vasoconstrictor response to increase in venous transmural pressure<sup>4</sup> and the lymphatic architecture and flow are altered in Dercum's SAT<sup>5</sup> consistent with Dr. Dercum's description of DD as a haemolymphatic disorder.<sup>6</sup>

**Pain:** The presence of pain is a requirement for diagnosis. The pain may be in the lipomas, in skin (hyperalgesia), or sharply referred. Pain in one area one day may be gone the next and other areas may become painful. Some areas that are painful for years can become numb. Arthralgias and myalgias are common. Over time, the pain is disabling.

**Metabolism:** Oxygen consumption per kilogram body weight in women with DD is lower than in controls matched for age, weight, body mass index, muscle and fat mass.<sup>3</sup> Fatty acid desaturation in SAT is lower in people with DD.<sup>7</sup> The fat also does not respond normally to norepinephrine and insulin<sup>8</sup>, and glucose conversion to neutral glycosides is reduced.<sup>9</sup>

**Co-morbidities:** Hypothyroidism, diabetes, autoimmune disease, fibromyalgia.<sup>1</sup>

**Common symptoms:** Sleep disorder, anxiety, depression, cognitive difficulties (brain fog), tachycardia, shortness of breath, gastrointestinal disturbances.<sup>1,10</sup> DD is similar to fibromyalgia but with the addition of painful lipomatous SAT.

**Serious Rare Complications:** Blood clots, fat emboli, fatty heart,<sup>11</sup> early cardiovascular disease, and lymphedema.

**Life Expectancy:** Effect on life expectancy unknown.

### Imaging:

Magnetic resonance imaging: Best imaging of the lipomas with MRI with T1 weighted sequences in two planes, and short-tau inversion recovery (STIR) and proton-density fat-saturated (PD-FS) sequences in one plane each.<sup>12</sup>

Computed tomography: Poor visualization of lipomas.

Ultrasound: Fair to good with an experienced ultrasound technician.<sup>12</sup>

### Evidence-Based Treatment (alphabetic order)

Bariatric surgery: Improves co-morbidities but weight loss may be minimal<sup>13</sup> while the lipomas remain.

Cycling hypobaric pressure: The *Cyclic Variations in Adaptive Conditioning™ (CVAC™)* process improved pain and mental functioning in ten people with DD.<sup>14</sup>

Infliximab and methotrexate: Improved pain and induced weight loss in a single woman with DD.<sup>15</sup>

Interferon alpha-2b: Induced pain relief in two patients with hepatitis C infection.<sup>16</sup>

Lipoma resection: Reduces pain but the lipomas may recur; inhibit recurrence and seroma formation with compression after resection for 6 weeks.<sup>17</sup>

Lidocaine: Topical and intravenous preparations have been used with variable success.<sup>18-20</sup>

Liposuction: Reduces pain and improves quality of life.<sup>2,21-23</sup>

Manual lymphatic drainage combined with compression garments and pregabalin (pain medication) reduced pain and weight – case study.<sup>24</sup>

Mexilitene: After intravenous lidocaine, mexilitene was able to maintain pain relief.<sup>25</sup>

Metformin: Case study.<sup>26</sup> May either not work for pain relief, or may lose its effectiveness over time.

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