

## CLINICAL VIGNETTE

# Dercum's Disease – A Mimic of Fibromyalgia

Gihyun Myung, M.D., and Meike A. Fang, M.D.

### Case Report

A 40-year-old Caucasian male with a history of depression, alcohol abuse, and traumatic splenectomy to the Rheumatology Clinic for management of chronic body pain that started about 12 years ago. He reported having sharp, intermittent shoulder, finger, knee, and hip pain not alleviated by acetaminophen, thermal modalities, or physical therapy. He also noted numbness and tingling of his left arm and left leg. He was prescribed amitriptyline and bupropion for depression; these medications did not alleviate his pain. The patient was previously told he had psychosomatic pain by a neurologist.

His vital signs were normal, and he weighed 149 lbs with a BMI of 22.0. His physical examination was remarkable for multiple small nodules on the trunk and extremities and tenderness in the right supraspinatus and right trapezius muscles as well as the bilateral C7 region. Neurological examination was normal. Laboratory evaluation including Westergren erythrocyte sedimentation rate, C-reactive protein, thyroid function tests, antinuclear antibody, and rheumatoid factor were unremarkable. The initial impression was that the patient had fibromyalgia, peripheral neuropathy, or a chronic pain syndrome. Amitriptyline was switched to nortriptyline, and an electromyogram and nerve conduction study of the left arm and left leg were performed and were normal.

At his follow-up visit to the Rheumatology Clinic three months later, the patient reported that he developed multiple lumps in his trunk and extremities about 10 years ago, which were painful to touch. He reported prior biopsy of one of the nodules revealed "fatty tumor". Repeat examination was notable for mobile, soft tender nodules on the trunk, bilateral arms extending from the antecubital fossa up to axillae and bilateral thighs. MRI of the right thigh showed tiny subcutaneous soft tissue nodular densities in the anterior medial proximal thigh.

His clinical presentation raised the possibility of Dercum's disease, so he was referred to a physician who specialized adipose tissue disorders. She confirmed that he had Dercum's disease type 3 and

prescribed a diet high in omega-3 fatty acids and recommended analgesics such as gabapentin, pregabalin, or meloxicam.

### Discussion

Dercum's disease (DD), also called adiposa dolorosa, is a rare condition characterized by painful subcutaneous lipomas.<sup>1</sup> There are three types of DD: Type 1 involves joints, classically the knees; Type 2 is diffuse and generalized; Type 3 is nodular, which causes intense pain around multiple "lipomas".<sup>2</sup>

The pain is chronic (>3 months), symmetrical, often disabling.<sup>3</sup> Even though prevalence is not known yet, DD is five to thirty times more common in women than in men.<sup>4</sup> It most commonly occurs in peri- and post-menopausal women between the ages of 40 and 60 years and is characterized by lipomas involving the trunk and extremities.<sup>5,6</sup> The presence of pain, which is required for diagnosis, is most commonly described as a burning or aching sensation.<sup>4</sup> Usually, patients complain of pain that appears out of proportion to physical findings, but pain can range from hyperalgesia, discomfort on palpation, to paroxysmal attacks of pain.<sup>4,7</sup> Even though obesity has been related with DD, it has been reported that people with normal weight can also have DD.<sup>8</sup>

In addition to pain, there are reported associations with weakness, depression, confusion, lethargy, and dementia.<sup>9</sup> The majority of the cases of DD occur sporadically, but some suggest DD might be an autosomal dominant disorder with variable expression.<sup>4</sup> The differential diagnoses of multiple lipomas include fibromyalgia, benign symmetric lipomatosis, neurofibromatosis Type 1 (NF1), diffuse lipomas, familiar multiple lipomatosis, panniculitis, and congenital lipomatosis.<sup>4,9</sup> It is important to note that DD is a clinical diagnosis and a diagnosis of exclusion.

The exact etiology of DD is unknown.<sup>9</sup> Some early reported cases suggested treating thyroid disorders decreased pain raising the possibility that thyroid dysfunction is associated with DD.<sup>10,11</sup> However, DD

generally continues to progress even if thyroid replacement treatment is started<sup>12</sup>, and recent studies have not revealed any endocrine laboratory abnormalities in patients with DD.<sup>4</sup>

Helpful imaging modalities include ultrasound and MRI. Power Doppler reveals superficial subcutaneous lesions and absence of surrounding edema or increased vascularity. Compared to lipomas, DD nodules are smaller (mostly <2cm) and more hyperechoic.<sup>13</sup> On MRI, lipomas generate high signal intensities on both T1- and T2-weighted images without enhancement after gadolinium injection. Ill-defined nodular/bluish-lesions sometimes may be seen on unenhanced MRI.<sup>13</sup> MRI provides accurate information regarding the relationship of the DD lesions to the surrounding tendons, bones, and joints and detects lesions that are not clinically obvious or symptomatic.<sup>1,13</sup> Unlike lipomas, which can occur anywhere in the subcutaneous fat, DD lesions are located in the superficial subcutaneous fat.<sup>13</sup>

DD does not have defining histological features; the biopsy of lipomas in DD is histologically identical to sporadic lipomas.<sup>9</sup> Other studies have reported inflammatory changes and angioliipoma-like features.<sup>14</sup> The cause of the pain in DD is unclear, but it may be due to pressure on nerves by the lipomas or increased vascularity, fibroblast proliferation, and fat cell necrosis around the nerve.<sup>1,9</sup>

There are standard recommendations that could be tried. Unfortunately, non-steroidal, anti-inflammatory drugs have little for no effect.<sup>1</sup> Intravenous lidocaine (400 mg over 15 mins every other day) or daily oral mexiletine have shown to provide pain relief.<sup>14</sup> However, the provider should consider drug-drug interactions with concomitant medications and potential side effects when prescribing these medications. If these medications do not adequately provide pain relief, pregabalin, amitriptyline, or SSRIs could be tried.

Another class of medications that has been suggested is corticosteroids, typically oral prednisone.<sup>1,2</sup> A single female patient's pain was improved with Infliximab and methotrexate<sup>15</sup>, and two patients with chronic hepatitis C were successfully treated with interferon alpha-2B.<sup>16</sup> Non-pharmacologic approaches such as exercise, cognitive behavioral therapy, hypnosis, biofeedback, relaxation, transcutaneous electrical nerve stimulation, and distractions can be used as adjuncts to pharmacologic therapy.<sup>12,17</sup> Liposuction can be also tried and is

considered similar in efficacy compared to the surgical excision of the lipomas.<sup>1,4</sup>

### **Conclusion**

DD is a rare condition and a diagnosis of exclusion. Although it is more common in obese women of middle age, it can occur in males and those who have normal BMI.<sup>8</sup> Due to overlapping symptoms, DD is commonly mistaken for fibromyalgia.<sup>13</sup> It is important to distinguish DD from fibromyalgia since management differs. Fibromyalgia is a condition with widespread muscle pain and a painful response to pressure. Even though distinguishing features between fibromyalgia and DD can be very subtle, DD is suspected if patient has the multiple painful nodules in subcutaneous tissue. Because pharmacotherapy alone is often unsuccessful, it is important to have a multidisciplinary approach encompassing medication, surgery, and mental health.

### **REFERENCES**

1. **Amine B, Leguilchard F, Benhamou CL.** Dercum's disease (adiposis dolorosa): a new case-report. *Joint Bone Spine.* 2004 Mar;71(2):147-9. Review. PubMed PMID:15050201.
2. **Herbst KL.** Rare adipose disorders (RADs) masquerading as obesity. *Acta Pharmacol Sin.* 2012 Feb;33(2):155-72. doi: 10.1038/aps.2011.153. Review. PubMed PMID: 22301856.
3. **Dercum FX.** A subcutaneous connective tissue dystrophy of the arms and back, associated with symptoms resembling myxedema. *University Med Mag (Philadelphia).* 1888;1:1-11.
4. **Hansson E, Svensson H, Brorson H.** Review of Dercum's disease and proposal of diagnostic criteria, diagnostic methods, classification and management. *Orphanet J Rare Dis.* 2012 Apr 30;7:23. doi: 10.1186/1750-1172-7-23. Review. PubMed PMID:22546240; PubMed Central PMCID: PMC3444313.
5. **Lynch HT, Harlan WL.** Hereditary Factors in Adiposis Dolorosa (Dercum's Disease). *Am J Hum Genet.* 1963 Jun;15(2):184-90. PubMed PMID: 17948485; PubMed Central PMCID: PMC1932230.
6. **Cantu JM, Ruiz-Barquin E, Jimenez M, Castillo L, Macotella-Ruiz E.** Autosomal dominant inheritance in adiposis dolorosa (Dercum's disease). *Humangenetik.* 1973 Mar 23;18(1):89-91. PubMed PMID: 4721343.

7. **Brorson H, Fagher B.** [Dercum's disease. Fatty tissue rheumatism caused by immune defense reaction?]. *Lakartidningen*. 1996 Apr 10;93(15):1430, 1433-6. Swedish. PubMed PMID: 8667732.
8. "Learning about Dercum Disease." National Human Genome Research Institute. June 27, 2012. Web. April 28, 2014.
9. **Campen R, Mankin H, Louis DN, Hirano M, Maccollin M.** Familial occurrence of adiposis dolorosa. *J Am Acad Dermatol*. 2001 Jan;44(1):132-6. PubMed PMID: 11148491.
10. **Frankenheimer JB.** Adiposis dolorosa. *J Am Med Assoc*. 1908;L:1012-3.
11. **Bergeron PN.** A case of adiposis dolorosa with involvement of the large nerve trunks. *J Nerv Mental Disease*. 1918; 36:159.
12. **Herbst KL, Asare-Bediako S.** Adiposis Dolorosa is More than Painful Fat. *The Endocrinologist*. 2007 Dec;17(6):326-44.
13. **Tins BJ, Matthews C, Haddaway M, Cassar-Pullicino VN, Lalam R, Singh J, Tyrrell PN.** Adiposis dolorosa (Dercum's disease): MRI and ultrasound appearances. *Clin Radiol*. 2013 Oct;68(10):1047-53. doi: 10.1016/j.crad.2013.05.004. Epub 2013 Jun 25. PubMed PMID: 23809264.
14. **Lange U, Oelzner P, Uhlemann C.** Dercum's disease (Lipomatosis dolorosa): successful therapy with pregabalin and manual lymphatic drainage and a current overview. *Rheumatol Int*. 2008 Nov;29(1):17-22. doi: 10.1007/s00296-008-0635-3. Epub 2008 Jul 5. PubMed PMID: 18604537.
15. **Singal A, Janiga JJ, Bossenbroek NM, Lim HW.** Dercum's disease (adiposis dolorosa): a report of improvement with infliximab and methotrexate. *J Eur Acad Dermatol Venereol*. 2007 May;21(5):717. PubMed PMID: 17448013.
16. **Gonciarz Z, Mazur W, Hartleb J, Machniak M, Bednarek I, Mazurek U, Wilczok T.** Interferon alfa-2b induced long-term relief of pain in two patients with adiposis dolorosa and chronic hepatitis C. *J Hepatol*. 1997 Dec;27(6):1141. PubMed PMID:9453446.
17. **Campen RB, Sang CN, Duncan LM.** Case records of the Massachusetts General Hospital. Case 25-2006. A 41-year-old woman with painful subcutaneous nodules. *N Engl J Med*. 2006 Aug 17;355(7):714-22. PubMed PMID: 16914708.