

Case Report

Dercum's Disease in a 38 Year Old White Male with Attempted Surgical Treatment: Case Report

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Abstract

Dercum's disease is a rare and poorly understood chronic process where pain and increased adipose tissue associated vague and highly variable symptoms are its typical presentation. In this case a 38 year old white male patient presented with a long standing history of lipomatous growths that only recently became painful. His presentation, medical history, and attempted surgical management will be detailed and compared to other case reports and surveys of this disease. Special care will be made to apply previously proposed diagnostic criteria and classification systems devised by other authors and how they apply to this patient. Ultimately this case warrants attention due to the rare nature of the disease and the unusual nature of the patient's symptoms and presentation.

Introduction

Dercum's Disease (also known as Adiposis Dolorosa, adipose tissue rheumatism etc.) is a chronic and progressive condition characterized by the presence of painful adipose tissue. Because it is rare, it has a poorly understood disease process and currently lacks fully established symptoms and diagnostic criteria. As a result, there can be a great deal of variation in individual presentations of this disease. Still, some attempts have been made at a stricter definition and these will be discussed in detail. Ultimately this disease is insidious and carries a poor prognosis because the pain becomes debilitating with time and no widely effective curative treatment has been found. Instead therapy has focused on symptomatic control with medical and surgical intervention.

Due to its rarity and explicit lack of mechanism for pain, diag-

nosing this disease can be difficult. Due to the variety of clinical presentations, clinicians must learn to recognize it and differentiate it from other disorders such as fibromyalgia, lipoedema, etc. In an attempt to help other clinicians better understand the non-specific and sometimes vague symptoms, this patient's history and physical will be discussed. This case of Dercum's disease will be compared to other case reports and proposed diagnostic criteria and classification systems from other authors will be utilized to better evaluate and describe this patient.

Case Report

A 38 year old white male presented to his primary care provider with a chief complaint of right lower quadrant pain and a 20 year history of multiple fatty tissue growths around his abdomen, back, upper and lower extremities. His pain was primarily located in the right lower quadrant and contained entirely in an apparent lipoma in that area. He described it as a 3/10 at the time, constant, crampy, worsened with palpation and light touch of the area, and without palliating factors. He also admitted to having similar pain on his left anterior thigh which also contained another apparent growth.

The patient first noticed the lipomas beginning to grow on his proximal arms and legs in 1997 after completing boot camp for the navy. At the time he did not have any pain and denies any specific event, trauma, or exposure that may have been influential in causing this. Steadily over the years he began to accumulate additional lipomas over his arms, legs, abdomen, and back. The above mentioned pain first began shortly after he had a laparoscopic cholecystectomy in 2014. Since then it steadily increased in intensity and frequency until it was hindrance in daily

activities.



Figure 1:

When questioned, the patient also admits to a myriad of other symptoms. He complains of chronically “cold feet” since 1997 and has had some transient numbness and tingling in his hands for 15 years. He has chronic lumbar pain for 4 years and is attributed to the degenerative disc disease diagnosed at that time. He also complains of persistent asthenia, having to take two naps daily and shortness of breath at night. But the patient states that he has no snoring and he denies any history of obstructive sleep apnea. The patient is also experiencing persistent non-bloody diarrhea and loose stools.

Also, noteworthy the patient admits to feeling depressed since 1998 secondary to worries about his appearance due to the lipomas. He also admits to three cases of disorientation and confusion which have occurred over the past two decades. He describes them as being “blanked out for a day or two at a time.” Patient reported that his family has observed him to be extremely “groggy” and not oriented to place or time. He denies any history of seizure like activity. These episodes then spontaneously resolve and the patient claims to have no memory of any events. Patient states that prior work ups failed to explain any apparent mechanism or cause for these symptoms.

The patient's medical history was significant for degenerative disc disease in the lumbar spine, previous laparoscopic cholecystectomy in 2014, and benign recurrent intrahepatic cholestasis (BRIC) with associated abnormal liver function tests. He denies history of other medical conditions and does not currently take any daily medications. His family and social history were non-significant

apart from his brother having lupus from a young age and his father being recently diagnosed with Crohn's disease.

On physical exam, the patient is fairly large and muscular at 6 ft with a stable BMI of 35.3 over the past decade. All vital signs were within normal limits. He had apparent numerous lipomas diffusely growing in the subcutaneous tissues. The remaining cardiac, respiratory, musculoskeletal, neurologic, abdominal, HEENT, and dermatologic exams failed to reveal any other significant findings apart from those mentioned above.



Figure 2:

After discussing options with the patient, he consented to surgical removal of the most problematic lipomas. Prior to surgery, the patient had had basic lab work done including a CMP and CBC. The CMP showed only mildly elevated bilirubin at 1.6 mg/dL, AST at 48 U/L, and ALT 119 U/L which were consistent with his diagnosis of BRIC. The CBC had only a minimally elevated RDW at 16.8%. Surgical removal of two 5x6 cm masses in the right flank and abdomen as well as a 6x6 cm mass from the left anterior thigh was completed without complications. Pathological reports showed the specimens contained only lobules of normal adipose tissue consistent with lipomas. At follow up one week later the patient stated that the pain from those lipomas had resolved, but others were becoming more painful. At that time he ranked his pain as 5/10 with no change in other described symptoms.

The patient then underwent surgical removal of four more masses several days later. 7x3 and 7x2.5 cm masses were removed

from the anterior and posterior right forearm, 4x2.5 cm on the right flank and 2x2.5 cm mass from the left arm were also removed. As before, pathological examination showed lobulated adipose tissue consistent with lipomas and appeared normal under microscopic examination. At 1 month follow up, the patient's presentation was similar to what was previously described. The pain at the area of surgical removal had resolved, but other masses were steadily becoming more painful. It is interesting to note that the transient numbness in his right hand did greatly improve as a result of the surgery. Otherwise there were no significant changes in described symptoms.

Discussion

Currently, all attempts at establishing a mechanism behind the pathogenesis of Dercum's disease have been inconclusive [1]. It has been previously suggested that a portion of the pain and symptoms may be due to mechanical pressure of growths on nearby nerves. In this patient's case, the localized pain of the lipomas had completely resolved after surgical removal as well as significant reduction in right sided hand paresthesia. Nerve impingement may play a part in some areas of this patient's pain, but it does not account for the entirety of his present complaints. The patient's description of his pain and its distribution elsewhere in his body are not consistent with nerve impingement and, as with other cases, no exact pathology or mechanism could be identified as the causative mechanism.

The exact prevalence rate (like the etiology) has not yet been established but analysis has shown that there are higher incidences among women compared to men at ~ 5:1 and an average age of onset at 34.5 ± 12.8 years [2]. Like in the above patient's case, early tumors occur primarily on the arms and legs [3]. The patient in this case had a fairly early onset at the age of 18. Still it is important to note that only recently in the course of his disease did he start to experience actual pain. If one were to use that as the exact qualifier behind his diagnosis, it is reasonable to state that his age at onset was 35 yo. This is particularly interesting when compared to other cases of Dercum's because of the extended latent period where this patient was pain free.

Exact symptoms and diagnostic criteria are still lacking, but there has been some head way. Recently Hansson, Svensson, and Broson expanded upon previous research and proposed the following criteria for diagnosis [1]:

- I. Generalized overweight or obesity
- II. Chronic pain (>3 months) in the adipose tissues. It should be noted that Dercum's disease has also been commonly described as having two additional criteria for diagnosis (as proposed by Roux and Vitaut in 1901), but there is a question as to whether these symptoms are directly caused by the condition versus associated with it.
- III. Weakness and susceptibility to fatigue (asthenia)

IV. Psychiatric manifestations, including emotional instability, depression, epilepsy, confusion, and dementia.

Generally speaking, obesity combined with chronic pain can easily lead to the above latter two points manifesting themselves due to the great stress they induce. His long standing fatigue is present and sufficient enough to impede daily activities. While this is due in some part to his nocturnal dyspnea, the patient felt that there was more at work here and that his shortness of breath was not significant enough to explain his symptoms.

There has also been evidence to show that there is a great correlation between Dercum's and depression. In fact, nearly half of patients diagnosed with Dercum's admit to depressive symptoms to some degree [4]. In our patient's case, he admits to a history of depressed mood as well as the episodes of confusion and memory loss. This patient's presentation should be more than enough to satisfy the minimal diagnostic criteria in points I and II as well as the additional points in III and IV.

As for the diagnosis of Dercum's disease, it in and of itself has a varied clinical presentation. To help account for this Hansson, Svensson, and Broson proposed the following classification system [1]:

- I. Generalized diffuse form: A form with diffusely widespread painful adipose tissue without clear lipomas.
- II. Generalized nodular form: A form with general pain in adipose tissue and intense pain in and around multiple lipomas.
- III. Localized nodular form: A form with pain in and around multiple lipomas
- IV. Juxta-articular form: A form with solitary deposits of excess fat for example at the medial aspect of the knee
- V. Given the patients description of pain and his presentation his clinical picture is most consistent with class III localized nodular form.

Treatment

For this patient, surgical management via lipectomy was attempted for the most problematic lipomas as they presented. This was also done to obtain specimens for pathologic review to aid in diagnosing the patient's symptoms. Though pain in the symptomatic lipomas had total improvement, other areas are persistently and increasingly becoming symptomatic. It is unfortunate, but this is similar to other cases of attempted surgical management with local excision [5, 6]. Liposuction has shown some efficacy when compared to control groups, but the increase in quality of life was deemed so insignificant as to not warrant the procedure [7]. In this patient's case, surgical excision offers relief for short periods of time but the rapid development of pain in other lipomas diminishes its efficacy as a primary treatment. Thusly other options must be explored.

The recent survey from Herbst of over 100 individuals diagnosed with Dercum's has shown some efficacy for pain relief in the form of analgesics and NSAIDS [3]. In one case report, intravenous lidocaine was particularly effective and offered significant pain relief for 2-12 months per treatment [8]. Intralesional Lidocaine has also shown some efficacy for lasting pain relief [9]. This treatment method, so far, has been regarded as one of the more effective options for symptom control.

Many other alternative treatments have been documented but have failed to have the reproducibility and relative lack of adverse effects of surgical intervention and the use of anesthetics such as lidocaine. A brief summary of such treatments is cited below from Kosseifi and are best used on a case by case basis [9]:

Systemic analgesics and opioids

Intravenous lidocaine injection (systemic toxicities)

Oral mexiletine (drugs interaction)

Local and systemic corticosteroids

Liposuction (refractory cases)

Methotrexate

Azathioprine

Cyclosporine

Hydroxychloroquin

Conclusion

Dercum's Disease is a particularly rare and poorly understood disease process that can be quite debilitating, thus it is important to document and share informative cases as well as effective means of managing symptoms. In this case report, the patient's

presentation does meet the qualifications for the commonly proposed diagnostic criteria of increased adipose tissue and chronic pain >3 months in duration (points I and II) as well as other associated symptoms of asthenia and psychological/neurologic manifestations (points III and IV respectively). Likewise, this supports the usage of points I and II as diagnostic criteria with points III and IV serving as associated or common symptoms to help differentiate this diagnosis from other diseases. Likewise, the proposed classification system does help describe this patient's case and likely warrants application in other instances of Dercum's in order to help standardize the approach to diagnosis.

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