

Dercum's Disease (adiposis dolorosa) & Associated Obesity

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Abstract

Dercum's disease also known as adiposis dolorosa is a rare condition that presents with numerous, painful fatty tissues/ subcutaneous lipomas. These lipomas are mainly spread throughout the trunk and extremities resulting in chronic pain and adipose tissue swelling. Dercum's disease has no identifiable etiology but has been linked to several endocrine disorders as well as obesity, trauma, and thought is thought of as an autoimmune disease. Mainstay treatment is focused on alleviating chronic pain whether that be with medication such as analgesics or liposuction and surgical intervention.

Presentation of Case: We present a rare case of a patient with multiple lipomas that continue to reoccur. The patient has had multiple surgeries to remove subcutaneous lipomas throughout her body. These lipomas ranged from non-painful to extremely tender with pain waxing and waning. The pain has not been resolved with prescriptions and over the counter medications. After thoroughly examining the patient's history, a probable diagnosis of Dercum's Disease was made.

Discussion: Dercum's disease is a rare condition characterized by the occurrence and recurrence of lipomas. Some other defining features include these masses being painful, varying in size, and distributed along the arms, legs, and trunk. Obesity is not uncommon in these cases as these masses are made up of fatty tissue. Various types of treatments are under scrutiny for their long-term effectiveness, namely compression therapy, pain medication, and liposuction.

Conclusion: The current mainstay treatment for Dercum's remains surgical intervention. With no clear data demonstrating the benefits of other medical interventions it is evident that liposuction along with other surgical means be used to alleviate the pain that many patients experience from lipomas.

Keywords: lipomas, obesity, connective tissue, excision, surgery

Introduction

Dercum's disease is a rare disorder characterized by multiple painful growths of loose connective tissue (fat tissue/ lipomas) which grow under the surface of the skin and can be superficial or deep. Lipomas in these diseases are found throughout the body most commonly occurring in the upper arms, upper legs, and trunk. Usually, the dorsal hands, feet, head, and neck are spared from the lipomata growth. The sizes of the lipomas vary, they can be as small as a grape or as big as a fist. Majority of the patients with Dercum's disease experience pain that can range from mild discomfort to excruciating pain if touched. The chronic pain stems from the mass effect of the lipomas pressing

on the nearby nerves as well as fascia which are inflamed connective tissue. Patients with this disease can develop swelling of various regions of the body. This is thought to be caused by inflammation slowing the lymphatic system in a painful limb and correspondingly presents with lymphedema. Histologically a biopsy taken will demonstrate macrophages and lymphocytes in a patient with Dercum's [1]. Although no epidemiology has been defined, this rare condition is thought to affect women 5-30 times more frequently than men and has an increased prevalence in obese females in the age group of 35- 50 [4].

Case Presentation

A 58-year-old Hispanic female with a past surgical history of a hysterectomy, 2002, appendectomy, 2005, colon resection, 2010, cholecystectomy, 2010, LAP band, 2014, laparoscopic removal of LAP band with exploratory laparotomy, adhesiolysis, enterolysis, enteroenterostomy, 2015 presented to the surgical outpatient clinic for consultation on the removal of a large painful lipoma on her left leg.

The patient presented with multiple lipomas throughout her body that had been growing since 2015. The patient expressed concern with multiple, painful lipomas that initially began on her left posterior thigh and have since spread to her abdomen, hands bilaterally, left leg, and scalp. These subcutaneous lipomas are tender and painful to palpation. The patient had seen multiple doctors with no definitive diagnosis.

Despite no family history of lipomatous growths, the patient's brother had a neoplastic growth removed from his back, and the patient did not know further details about the growth besides the fact it was cancerous. Multiple prescriptions and over the counter treatments had failed to improve her pain or recurrence of growths. The patient reported having used analgesics such as tramadol and oral lidocaine

Discussion

A lipoma is a growth of a mass consisting of adipose (fatty) loose connective tissue. Dercum's disease, a pathology first discussed in 1882 by an American neurologist Francis Xavier Dercum's, deals with multiple lipomas growing in various sizes in the subcutaneous fat. These growths can occur and reoccur, anywhere on the body, but the arms, legs, and trunk are especially characteristic. Although there is no direct etiology known for the disease, some cases have shown familial patterns of autosomal dominant inheritance (FML type or Angio lipomatosis type). The disease has been seen to affect women more often than men by about 5-30 times, the average age being 34 years. These lipomas are often painful, with the intensity residing all along the spectrum of pain, from mild to excruciating to the touch. Fat tends to have more pain receptors than there are in muscles, so these lipomas in a way, increase the patient's sensitivity to pain. Patients have described this sensation as all of their fat hurting, and it is known to have periods of flare-ups followed by temporary cessation. This pain can be caused by blocked lymphatic drainage, any type of inflammatory process in the body that may lead to an inflamed fascia, or from a mass effect of the lipoma compressing nearby nerves as they pass by. Lymphatic drainage has been seen to be slower in the more painful limbs, leading to possible lymphedema. This pain can correct itself, however, manual massaging to aid in lymphatic drainage has shown to help alleviate the pain. Along with this discomfort, there are other symptoms of Dercum's disease, like weight gain, fatigue, weakness, easy bruising, headaches, irritability, morning stiffness, issues with the gastrointestinal system, racing heart, and shortness of breath. Of the mentioned, significant weight gain is the most pressing to treat, to prevent other complications of obesity such as diabetes. A "brain fog" has also been noted in some patients, in which patients can suffer from depression, memory loss, loss of concentration, and anxiety. Dercum's has even been linked to other conditions such as arthritis, hypertension, congestive heart failure, sleep disorders, dry eyes, and myxedema from

Conclusion

Dercum's disease often presents with painful lipomas and various other symptoms thus making the diagnosis one of exclusion and one that is very difficult based on presentation alone. This disease mainly affects the elderly population more often and occurs sporadically with no clear early detectable signs. As in the case of our patient, this disease presents with waxing and waning pain that can be precipitated even by a light touch if the lipoma is overlying nerves. Management

patches to no resolve, as well as compression stockings and daily cleaning of the skin surrounding the lipomas. The patient had normal vitals during the office visit. The patient was scheduled for excision of a large lipoma on her left leg near her inguinal area. She will be reexamined for possible further excisions as lipomas on her arm and abdomen start to become painful.

hypothyroidism. The diagnosis is mostly clinical; a thorough patient history and recognizing the numerous lipomas in the distribution common in Dercum's disease can help distinguish this from the other similar disorders. In some cases, a biopsy may be done to ensure the masses are in fact lipomas. Currently, there is no exact treatment for Dercum's disease, and the main focus is to ease the associated symptoms. Getting the lipomas surgically removed may cause temporary relief, but there is debate as to whether or not the surgery causes more harm than good. Seeing as the body will undergo an inflammatory response to heal from the surgery, this can give rise to new lipomas. There have been efforts made to decrease the pain with liposuction, however, with time the effectiveness of this method has been seen to dissipate. Some research has been done on the effectiveness of interferon alfa-2b to reduce inflammation, and in doing so, improving the condition of patients with Dercum's disease, but more research is necessary to confirm its benefits. For now, the recommended treatment is manual compression therapy to help with lymphatic drainage and pharmacotherapy with Pregabalin [2,3]. Our patient is a textbook example of this rare disease. She inhabits a great number of complications associated with Dercum's disease. At a weight of 302 lbs, and a BMI of 44.6, she is categorized as being morbidly obese, a common result of having excessive amounts of fatty tissue associated with the lipomas. A LAP band procedure was even attempted to control the patient's weight, however, due to adhesion formation, the LAP band had to be removed. From this obesity stems diabetes, high levels of cholesterol, and hypertension, all of which the patient is victim to. She is also noted to have arthritis, yet another associated condition. Along with these, the lipomas seen in our patient are all in the distribution unique to Dercum's disease. They had started on her legs, moved up the trunk, and are now present along her arms and hands as well. Finally, after four surgeries to remove these masses, and counting, she has experienced the recurrence of these painful lipomas as well.

of this disease is focused on controlling pain through removing the lipoma via liposuction or other surgical procedures or using analgesics such as lidocaine [5]. Removal of lipomas through surgical intervention remains the treatment of choice, although the possibility of regrowth is always there. Surgical benefits include cosmetically improving the appearance by the removal of the lipomas but also removes any irritation to nerves which will result in pain reduction.

Conflict of Interest Disclosure: The authors declare that they have no financial conflict of interest about the content of this report.

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