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# ADIPOSIS DOLOROSA ( DERCUM'S DISEASE)

Nursing

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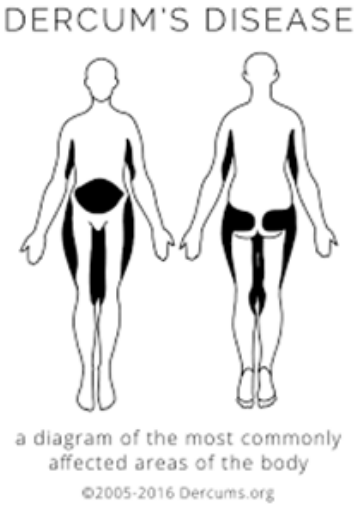
KEYWORDS

## INTRODUCTION

Dercum's disease also referred to as Anders' syndrome, Adiposis Dolorosa, or Dercum-Vitaut syndrome is one among the rarest medical conditions. It was first described in 1892 by Francis Xavier Dercum. It is more common in postmenopausal women and is characterized by painful subcutaneous lipomas or fat accumulations largely on the trunk and limbs. There is also associated asthenia, easy bruising over the affected areas and hypercholesterolemia . Dercum's disease is believed to be transmitted in an autosomal dominant manner , however most reported cases of adiposis dolorosa are sporadic . The understanding of the pathogenesis and the mechanism of Dercum's disease remain unknown. It is believed that fatty deposits cause nerve compression and result in weakness and pain.




### Definition of Dercum's disease

According to Dorland's 27th edition text in the National Library of Medicine (IGM Metathesaurus), it is defined as "A disease accompanied by painful localized fatty swellings and by various nerve lesions. It is usually seen in women and may cause death from pulmonary complications."

### According to the paraphrased version of the National Organization of Rare Diseases (NORD):

"Dercums Disease is a rare disorder in which there are fatty deposits which apply pressure to the nerves, resulting in weakness and pain. Various areas of the body may swell for no apparent reason. The swelling may disappear without treatment, leaving hardened tissue or pendulous skin folds."

### Who gets Dercum disease?

- Dercum disease affects women more frequently than men (it's reportedly, 5–30 times more common in women than in men).
- It usually appears between 35 and 50 years of age.
- It may be more common in people with obesity.
- family history of autosomal dominant pattern of inheritance

### Etiological factors:

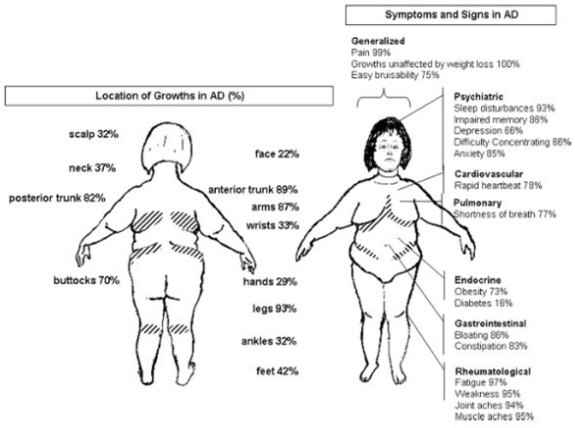
The underlying cause of adiposis dolorosa remains unknown. Possible causes are as follows

- long-term treatment with high-dose corticosteroids
- endocrine system abnormalities
- nervous system dysfunction
- mechanical pressure on nerves
- Trauma
- adipose tissue dysfunction; trauma
- Changes in fatty acid or carbohydrate metabolism

### Signs and Symptoms:

**Four cardinal symptoms have sometimes been used as diagnostic criteria:**

1. painful, fatty lipomas (benign fatty tumors) across anatomy
2. obesity, frequently in menopausal age
3. weakness and fatigue
4. emotional instability, depression, epilepsy, confusion, and dementia.




### There are also potential signs of the disease which are identified as the following:

- being bruised easily
- trouble with sleeping
- memory issues
- elevated heart rate
- difficulty with concentration
- joint aches
- shortness of breath
- vascular problems (angiolipomas)
- petechiae
- Flushing
- heavy or prolonged menstrual bleeding
- unexplained blood in the urine (hematuria)
- non-pitting edema
- in subcutaneous fat
- gastroesophageal reflux
- (GERD)
- irritable bowel syndrome
- and other gastrointestinal problems
- Migraines
- feeling of fullness
- tachycardia
- (rapid heart rate)

### Characteristics of pain

The painful lipomas have been reported to occur in any location, except in the head and the neck. The pain varies from discomfort on palpation to excruciating, paroxysmal spontaneous attacks. The pain can be aching, burning, or stabbing, often described by the patient as "it hurts everywhere."The pain is usually symmetrical however; it can become localized to the thighs, the knees, or the upper extremities. Pain can be felt in the skeletal system and in the fat.

**What are the types of Dercum's disease?**
Based on the widely spreading pain, various types of Dercum's disease can be diagnosed which are as follows:

**Type I:** Also known as the juxta-articular type of dercum's disease which is characterized with painful, fat folds over the hips and within the knees. Rarely, it is accompanied by fat growths in the upper-arm.

**Type II:** Also called a diffuse, generalized type that is characterized with widespread pain due to a fatty growth. Unlike type I variety, it causes painful fat growths in various fat bearing areas of the body such as the dorsal upper-arm fat, the stomach wall, soles of the feet, dorsal fat folds, and in the axillary and gluteal regions.

**Type III:** Also called as the nodular type of lipomatosis which is associated with intense pain around different parts that bear multiple lipomas. It may also occur in individuals without generalized obesity. Clinically, these lipomas measure between 0.5-4 cm, have soft consistency, and are attached to the adjacent tissue.

**How is Dercum disease diagnosed?**
A diagnosis of Dercum disease can be made through a systematic physical examination and identification of the characteristic triad of features (multiple lipomas, painful plaques and obesity).

**Multiple lipomas may also be features of:**
- Multiple lipomatosis (familial or sporadic)
- Fibromyalgia
- Panniculitis
- Obesity without painful plaques
- Cushingsyndrome, which may also lead to abnormal fat distribution and ecchymoses
- A biopsy may be helpful to distinguish a lipoma from an angiolipoma (another type of painful fat overgrowth).
- Other lesions that may mimic lipomas include
- Epidermoid cysts (which usually have a central punctum)
- Hibernoma (a benign growth of brown fat)
- Lipofascial hernia (a hernia in the anal or perianal region)
- Lipoedema

**How to treat the condition?**
There is no “cure” for Dercum disease. At the present time, treatment for this condition is symptomatic, meaning that it focuses on one symptom at a time rather than the whole condition. It is a very rare disease, and many clinicians are unaware of its existence. However, lipomas have been known to reoccur and even increase in number at the same site or a nearby location. Weight reduction may help with joint pain in some individuals, but has proven to be difficult to achieve and generally doesn't offer significant relief of symptoms.

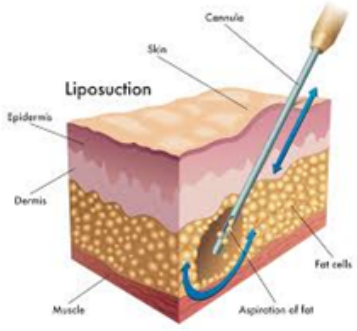
***Treatment methods include the following modalities:***
- **Symptomatic treatments that have led to some pain reduction in some affected people include:**

1. Prednisone or intravenous lidocaine for pain
2. Traditional pain medicines such nonsteroidal anti-inflammatory drugs (which are often ineffective), or acetaminophen combined with an opioid analgesic
3. Cortisone/anesthetic injection for localized pain
4. Diuretics for swelling of the fingers
5. Methotrexate
   The mechanism of action is unclear. Previously, methotrexate has been helps to reduce neuropathic pain caused by peripheral nerve.
6. Infliximab:
   Infliximab reduces neuropathic pain in patients with central nervous system sarcoidosis. The mechanism of action is tumour necrosis factor inhibition.
7. interferon α-2b:

The mechanism could be the antiviral effect of the drug, the production of endogenous substances, such as endorphins, or interference with the production of interleukin-1 and tumour necrosis factor. Interleukin-1 and tumour necrosis factor are involved in cutaneous hyperalgesia

8. Calcium-channel modulators

**Surgery**
Surgical excision of fatty tissue deposits around joints (liposuction) has been used in some cases. It may temporarily relieve symptoms although recurrences often develop. Surgical removal of particularly burdensome lesions and/or liposuction may be helpful for some people.




Liposuction is indicated for patients with general lower-body fat or more localized large deposits of fat at the knees, on the arms, on the thighs, or on the stomach as opposed to those with general diffuse pain. In those patients, liposuction is considered a risky operation, requiring about a week of care in the plastic surgery department.

**Adjunctive therapies**

- Rapid cycling hypobaric pressure.

Cyclic Variations in Adaptive Conditioning (CVAC) is a method of touch free cyclic hypobaric pneumatic compression for treatment of tissue edema and, therefore, edema-associated pain. A pressure variation accomplishes changes in both warm and cold temperature. The body constantly adapts to these changes and varied patterns of low-pressure air, accomplishing a physical challenge for every cell in the body, just as with exercise. These changes have been described to improve blood and lymphatic fluid in the body, and have been shown to reduce pain in Dercum's disease.

***Other methods includes***
acupuncture

- cognitive behavioral therapy
- Hypnosis
- biofeedback

**Consultations**
***Psychiatrist:***
Depression and other psychosomatic symptoms are associated with Dercum disease (adiposis dolorosa). Many patients find they are misjudged and require psychological support.

***Rheumatologist:***
A rheumatologic consultation is warranted to rule out osteoarthritis and fibromyalgia.

***Endocrinologist:***
An endocrinologic etiology, such as hypothyroidism and Cushing syndrome, should be ruled out.

**Diet**
Experience shows that lasting weight reduction by changing the diet is difficult to achieve and does not appreciably affect the pain.

**Activity**
Light physical activity may worsen symptoms because of the stiffness experienced after periods of rest and minimal activity. Patients should avoid monotonous, static work and physical and psychological stress.

**Patient Education**
Educating patients about the chronicity of Dercum disease (adiposis

dolorosa) and the available limited treatment modalities is important. Proper education about the aggravating and relieving factors should be explained.

Patient education about Dercum disease (adiposis dolorosa) is crucial. Addressing any possible needs of those persons with disabilities is important, preferably with the assistance of an occupational therapist and a social worker. Various aids may be needed in the home and at work.

**Prognosis**

The disease course is chronic and progressive

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