

# Adiposa Dolorosa / Dercum's Disease

Adiposis dolorosa, also known as Dercum's disease, is a rare and complex dermatologic disorder characterized by the development of painful lipomas (benign fat tissue growths) in individuals, predominantly in obese patients. These lipomas are typically associated with intermittent episodes of severe pain that may persist for hours or become chronic. The disease is also accompanied by a range of psychiatric symptoms, including fatigue, depression, anxiety, and cognitive impairment. Despite its impact, Dercum's disease remains underdiagnosed, with a significantly higher incidence observed in women compared to men, and it often presents around the age of 35-36 years, though childhood onset has been documented.

## Clinical Presentation

The hallmark of Dercum's disease is the development of lipomas, which are typically painful and are most often located on the trunk, upper arms, and upper legs. However, in some cases, lipomas may also appear on the head, hands, and feet. The size of the lipomas varies widely, ranging from small nodules, comparable to a grain of rice, to large masses that can reach the size of an apple. Histologically, these lipomas are characterized by an increased density of pain receptors within the tissue, which likely explains the chronic pain commonly experienced by affected individuals.

In addition to the lipomas, patients with Dercum's disease may experience a variety of associated symptoms, including swelling of the arms and legs, bloating, constipation, weight gain, shortness of breath, and easy bruising. Notably, the pain associated with the lipomas can be severe and is often worsened by the compression of nerves or the surrounding tissue. This chronic pain component makes the disease particularly debilitating, and it is one of the major factors contributing to the functional impairment observed in affected individuals.

## Pathophysiology

The precise etiology of Dercum's disease remains largely unclear, though several potential contributing factors have been proposed. These include fat tissue dysfunction, abnormalities in fat metabolism, and dysregulation of the nervous system. The growth of lipomas in Dercum's disease is believed to result from abnormal fat cell proliferation, with these growths displaying increased pain receptor density, which may contribute to the chronic pain observed in patients. Furthermore, nerve entrapment caused by the lipomas may exacerbate the pain through mechanical compression, leading to neuralgia.

## Diagnosis

Dercum's disease is typically diagnosed through clinical evaluation, relying on the patient's history

and physical examination. A key diagnostic feature is the presence of painful lipomas in an obese individual, with the pain persisting for at least three months. The diagnosis is further supported by the exclusion of other conditions with similar clinical features. While biopsy or imaging is not generally required for diagnosis, it may be utilized in specific cases to rule out other potential causes or confirm the presence of lipomas and their characteristics.

## Management and Treatment

Currently, there is no definitive cure for Dercum's disease, and treatment focuses primarily on symptom management and pain relief. Although weight loss may offer some benefit to individuals with obesity, it does not lead to complete resolution of the disease. A range of treatment options has been explored, although none have provided a cure for the condition.

These treatments include:

- **Pharmacological Interventions:** Medications aimed at alleviating pain and managing inflammation are commonly used. These include steroids, immune modulators, and pain-relieving agents such as opioids, anticonvulsants, and antidepressants. While these treatments may help manage symptoms, they do not address the underlying cause of the disease.
- **Liposuction:** For some patients, liposuction has been used as a method to remove lipomas, which can offer temporary relief from pain. However, liposuction does not prevent the formation of new lipomas, and the procedure may need to be repeated over time.
- **Multidisciplinary Pain Management:** Due to the complex nature of Dercum's disease, the most effective treatment often involves a multidisciplinary approach. This typically includes pain management specialists, physical therapists, and psychological support for managing both the physical and psychiatric symptoms of the disease. Treatment may also involve manual lymphatic drainage to help manage swelling and bloating, which can provide symptomatic relief.
- **Psychiatric Support:** Given the psychiatric symptoms associated with Dercum's disease, such as depression, anxiety, and cognitive impairment, addressing the psychological aspects of the disease is crucial. Cognitive behavioral therapy (CBT) and support groups can help patients cope with the mental health challenges associated with chronic pain and disease burden.

## Prognosis

The prognosis for individuals with Dercum's disease varies. While the disease is chronic and progressive, many patients can lead relatively normal lives with appropriate pain management and support. However, the severity of symptoms can vary significantly between patients, with some experiencing periods of remission and others having continuous pain. Given the lack of a cure, the focus remains on symptomatic management and improving the patient's quality of life.

## Conclusion

Adiposis dolorosa, or Dercum's disease, is a rare but debilitating condition characterized by the growth of painful lipomas in obese individuals, with associated psychiatric symptoms and significant functional impairment. Although the underlying cause remains unknown, potential factors include fat tissue dysfunction and nerve abnormalities. Treatment is symptomatic, with no single approach proving universally effective. A multidisciplinary approach involving pain management, psychological support, and liposuction for lipoma removal is currently the most successful strategy for managing the disease and improving patient quality of life.

## References

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