Dercum’s disease, also known as Adiposis Dolorosa, Anders’ syndrome and Dercum-Vitaut syndrome, is an extremely rare disorder characterized by multiple painful fatty tissue growths known as lipomas. These fatty growths will form mainly over the trunk, upper arms and legs. Dercum lipomas are extremely painful thus differentiating themselves from normally fatty tissue lipomas. This disease affects more women than men and is reported occurring between the ages of 45-60, especially in post menopausal overweight women. Some reports cite the disease is 20 times more likely to occur in women than in men. The current prevalence of the disease is unknown as the disorder is under-diagnosed, thus making it difficult to report the correct number of cases in the population. Dercum’s disease was first described in 1882 by an American neurologist named Francis Xavier Dercum.

Pain caused by these growths is thought to be associated with the tissue pressing on nearby nerves. The fatty deposits (lipomas) cause nerve compression and can also result in weakness and pain. Pain may vary from mild discomfort when touched to severe pain that is not in proportion to the physical manifestation. Some patients have been reported saying “all fat hurts.” Pain from the lipomas can last for hours or wax and wane continuously.

Sleep professionals are familiar with fibromyalgia and chronic pain syndromes. Most of the chronic pain syndromes are characterized by pain throughout the muscles with pain being sudden or beginning gradually. Chronic pain syndromes are more likely to affect the back of the neck, shoulders, elbows, hips, knees or lower back. Small specific areas are more painful and patients commonly refer to them as “tender points.” This differs from the symptoms and location of the growths associated with Dercum’s disease.

Rare Dercum disease cases have reported swelling of other body parts, especially the hands. Swelling can occur for no apparent reason and disappears without treatment. Significant weight gain is a common occurrence for most patients affected by Dercum’s disease. Medical literature has linked Dercum’s disease to other medical conditions such as arthritis, hypertension, congestive heart failure, sleep disorders, dry eyes and myxedema (a condition due to underactive thyroid causing dry skin, swelling around the lips and nose and mental deterioration).

So what causes Dercum’s disease? The exact cause is unknown and the disease appears to occur spontaneously and sporadically for no apparent reason. Some literature reports have suggested Dercum’s disease may be linked to an autoimmune disorder in which the body’s immune system mistakenly begins to attack healthy tissue. Disturbances in the endocrine and metabolic systems have been proposed as playing a potential role in the development of the disorder.

Cases of Dercum’s disease have been reported to run in families and some reports cite the possibility the disorder may be inherited as an autosomal dominant trait. Genetic diseases are passed on through a gene for a particular trait on a chromosome received by either the mother or father. With Dercum's disease, a single copy of an abnormal gene can result in an affected individual. The risk of passing the abnormal gene to potential offspring is 50 percent for each pregnancy regardless of the sex of the child.

The diagnosis of Dercum's disease is based on a very detailed patient history with thorough identification of the multiple fatty growths. To make a definitive diagnosis, four major symptoms should be present: (1) multiple, painful, fatty masses; (2) generalized obesity; (3) weakness and fatigue; and (4) mental disturbances, including emotional instability, depression, epilepsy, confusion and dementia. Additional symptoms may include cyanosis (a bluish discoloration of the skin), hypertension, headaches, and epistaxis (nosebleeds).

Treatment of this disease can vary from the use of painkillers (analgesics) to surgical options. Surgical removal and biopsy of the tissue will confirm the growths to be lipomas. Surgical excision of the fatty deposits around the joints may temporarily relieve pain although recurrences often happen. Liposuction has been useful as it provides an initial reduction of the pain with improvement of quality of life.

Psychotherapy and pain management are treatment choices for patients dealing with long-term intense pain. Researchers have reported the use of interferon alfa-2b as a potential drug to treat this disease. Patients receiving interferon alfa-2b for the treatment of hepatitis C also reported improvement with Dercum’s disease.
Alternative approaches may also be pursued such as acupuncture, biofeedback, cognitive behavioral therapy and hypnosis. Clinical trials on Dercum's disease are ongoing and being conducted by the National Institutes of Health (NIH).

Dercum's disease is a rare occurrence in the sleep center. Technologists who are aware of a reported pain syndrome patient may encounter this disorder. Care with the placement of electrodes and belts should be taken if the disease process has been identified.

REFERENCES


