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DERCUM'S DISEASE: Fatty tissue rheumatism caused by immune defense reaction?

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SUMMARY:

Dercum's disease, adiposis dolorosa or, more commonly, fatty tissue rheumatism, is a relatively unknown illness in spite of it being described as early as 1888 by the American neurologist Francis Xavier Dercum. It especially affects women of 20-40 years, but can even make an appearance in children. Pain in fatty tissue is the characteristic symptom. The patients, like sufferers of fibromyalgia, often imprecisely describe it saying, "all fat hurts a little". The disease is, however, classified by the WHO (World Health Organization).

Francis Xavier Dercum was born in Philadelphia on 10 August 1856 during a time of intense political unrest, which eventually resulted in the American Civil War. The family was of English descent. Dercum's father, Earnest Dercum, was a successful bookseller and grain wholesaler, which made it possible to finance his son's education.

Francis Dercum left Central High School and commenced his medical studies at the University of Pennsylvania, taking his doctor's examination in 1877. In 1878 he became associate professor in histology with Dr Richardson and worked together with Dr George A. Pierso, who later succeeded the great Leidy as professor of anatomy at the University of Pennsylvania. Dercum later became associate professor in physiology to Professor Harrison Allen. He lectured in practical biology and demonstrated in various fungi, protozoa, algae and bacteria. In this way he gained a solid background in what would later come to be called bacteriology.

He was very interested in natural history and early on became a member of the Academy of Natural Sciences, in which he was active and an enthusiastic debater for many years. His first published work was about the nervous system of fish. This, like an article about the brain, aroused a great deal of attention in the USA and Europe. He was especially interested in neuropathology and early in his career worked as a pathologist at the State Hospital for the Insane in Norristown.

Revolutionary patient records

In 1884 Dercum took over from Dr Charles K. Mills as head of the neurological clinic at the Hospital of the University of Pennsylvania. He was already in full practice as a general practitioner and a skilful diagnostician.

At that time the photographer and scientist Eadweard Muybridge commenced his study of movement. The motion film had not yet been invented and Muybridge took his pictures with the aid of a camera attached to the inside of a cylinder which he then rotated. This was the principle used by the film industry of the future.

Dercum entrusted him with photographing patients with pathological gait patterns and patients in convulsions brought about with the help of hypnosis. These pictures were the first to document neurological diseases. Muybridge published a major work consisting of eleven volumes entitled "Animal Locomotion", with 100,000 photographs, in which Volume 8 contains Dercum's patients.

Unpaid work

Dercum was one of the founders of the Philadelphia Neurological Society in 1884. He became a member of the American Neurological Society in 1885 and its president in 1886. In 1885 he also became a member of the College of Physicians of Philadelphia; in 1887 he took a post as neurologist at the Philadelphia Hospital, where there were numerous neurological patients. Although he had a large private practice, he held this unsalaried position until 1911, almost a quarter of a century. He married Elisabeth De Haven Comly, from an old Philadelphia family, in 1892.

Professor at the age of 36

In 1892 Dercum was called upon to fill the newly created post of professor of neurology at the Jefferson Medical College. A separate clinic was built in 1900 and Dercum was appointed professor of nervous and mental illnesses. He led the department until 1925 when he became an emeritus professor.

He was one of the great names at the Jefferson Medical College; a respected and popular teacher, a highly esteemed writer, a much sought after clinician and a greatly valued ethical expert in courts of law in cases concerning nervous and mental illnesses and trauma. His long and successful career makes him one of the most prominent figures in neurological history. One of his eminent patients was President Wilson, during the years 1919-1920.

Dercum's colleague, Dr Chalmers Da Costa eloquently describes him in "The Trials and Triumphs of the Surgeon and Other Literary Gems"⁽¹⁾.

"He was particularly strong in teaching insanity and his demonstrations of those borderland cases which lie near, but not in the dark continent of insanity were masterly. It was striking to hear him develop from a patient a story of an obsession, of a morbid impulse, of a morbid act, of an illusion, of an hallucination, of a delusion. In regard to that curious condition, hysteria, he actually convinced me that the condition is a reality and not clever acting. He would sum up a case like an able judge sums up from the bench. He was positive whenever it was possible to be positive, admitting doubt if doubts existed, and he always gave credit to others who deserved it. He showed that to the very basis of his nature he was a good doctor."

Dr Tom Bentley Throckmorton gives his assessment of Dr Dercum's lecture to medical students⁽²⁾:

"I shall never forget the first of Professor Decree's clinics that I attended. His ability to engage the attention of the students and then to hold it seemed almost uncanny. On this particular morning he demonstrated the difference between lesions involving the upper motor pathway and lesion involving the lower motor pathway. A patient who had suffered a cerebral haemorrhage, with the resultant hemiplegic state, was the first subject of discussion before the class. The spastic arm and leg, the scythelike swing of the palsied limb as the patient walked, the characteristic attitude of the affected arm, the weakness of the lower part of the face, the deviation of the tongue, the increased tendon reflexes, the patellar and ankle clonus and the extensor toe sign were all pointed out and then analysed with such clarity of thought and logical deductions as to hold the listeners spellbound.

"In contrast, another patient was used as a demonstration of what lesions of the lower motor pathway produced. This patient was a child who had suffered from an attack of acute poliomyelitis, which resulted in paralysis of both legs and one arm. Here again, the professor pointed out the cardinal symptoms of a lower motor neuron involvement, such as lost tendon reflexes, flaccidity and atrophy of muscles with associated electrical reactions of degeneration, and an absence of pathologic toe signs."

Adiposis dolorosa called after Dercum

Dercum is one of the few neurologists who has had a disease called after him, adiposis dolorosa. The first case was described in 1888 at a meeting in Washington of the American Neurological Association. It was published in the University Medical Magazine in the same year⁽³⁾. In 1892 he gave an account of two further cases at the same meeting⁽⁴⁾. He writes:

"Evidently the disease is not simple obesity. If so, how are we to dispose of the nervous elements present? Equally plain is it there we have not myxoedema to deal with. All of these cases lack the peculiar physiognomy, the spade-like hands, the infiltrated skin, the peculiar slowing of speech, and the host of other symptoms found in true myxoedema. It would seem, then, that we have here to deal with a connective-tissue dystrophy, a fatty metamorphosis of various stages of completeness, occurring in separate regions, or at best unevenly distributed and associated with symptoms suggestive of an irregular and fugitive irritation of the nerve-trunks - possibly a neuritis ... Inasmuch as fatty swelling and pain are the most prominent features of the disease, I propose for it the name Adiposis Dolorosa."

Productive writer

In 1895 Dercum was editor of the "Textbook on Nervous Diseases by American Authors", the first of its kind and including several writers. His other publications included "A Clinical Manual of Mental Diseases" (1913), "Hysteria and Accident Compensation" (1916), "Rest, Suggestion and Other therapeutic Measures in Nervous and Mental Diseases" (1917), "An essay on the Physiology of Mind" (1922), "The Biology of Internal Secretions" (1924) and "The Physiology of the Mind" (1925). With time he became more interested in psychology and philosophy.

Dercum was president of the American Neurological Association (1886) and of the Philadelphia Neurological Society (1892 and 1898). His membership of the Société de Neurologie de Paris was a great honor - this organization had fewer than 50 members from all over the world. In 1922 he received the Legion of Honour from the French government.

Francis Dercum's interest for philosophy also gained him a doctorate and in 1927 he was elected to the position of president of the American Philosophical Society (APS), of which he had been a member for

35 years. He died of a heart attack at the age of 74 on 23 April 1931 during a meeting and sitting in the famous "ladder library chair" made by Benjamin Franklin, who was also founder of the APS. He is buried in West Laurel Hill Cemetery, Philadelphia. His name can be added to the line of distinguished American neurologists and psychiatrists who have made a contribution to medical history. ^(1-2, 5-10)

Dercum's disease a fatty tissue rheumatism

The main symptom of the disease is generalized or local painful fat with certain inflammatory characteristics. It was described in 1888, i.e., long before the fibromyalgia syndrome was defined. In southern Sweden the name fatty tissue rheumatism has been applied, where the disease does not seem to be particularly rare. Based on the total number of patients diagnosed so far in the sample area of Lund, the total number of the case in the country may be estimated at least 10,000. Many people are probably living unaware of the illness, mainly because of having only mild symptoms or receiving the wrong diagnosis.

Up to 1871, Dercum's disease had been described in relatively few publications, covering a total of 400 cases ^(11, 12). The patients have, like fibromyalgia sufferers, often described feelings of "smarting, aching, and burning" also saying that "all fat hurts a little", the latter of course a misconception. The Swedish National Social Welfare Board has given the syndrome the classification number 272W, in accordance with ICD-9, 272.8. The new WHO classification ICD-10 gives the code E88.2.

Adiposis dolorosa affects mainly women of 25-40 years, but can make its appearance at any age. Much suggests a dominant line of inheritance ^(13, 14), particularly strong in the line great grandmother-mother-daughter. The disease is approximately twenty times more prevalent amongst women than men. The cause of the fatty deposits has been inadequately researched. The symptoms and the resistance to treatment can seriously affect quality of life. Those who seek medical treatment have usually suffered a sudden worsening of the condition (or: disease/ailment), and the symptoms then lead to permanent incapacity to work. More than half of the patients are not fit to work. In theory the disease could partly be due to an immune defense reaction. Its sometimes sudden appearance, together with the incidence of a slight increase in the number of inflamed cells in the fat, would suggest this. The sympathetic nervous system may play a role in the origin and development of the pain ^(15, 16).

Diagnosis

The diagnosis, as for fibromyalgia, is made purely clinically ⁽¹⁷⁾. Unlike fibromyalgia there is a relationship between pain and body weight. The pain is either general and is often more severe than in fibromyalgia. An insidious appearance is usual and pain increases with time. Like fibromyalgia, the symptoms of a number of associated diseases are evident (see below). The fundamental criteria here are pain in the fatty tissue and obesity.

The pain is chronic (more than three months' duration), nociceptive, usually symmetrical, but can exceptionally be one-sided for a while after its appearance. Occasionally it can be localized to the upper arm, the thigh or the knee. The pain is described as aching, stabbing, smarting or burning: "it hurts everywhere".

Hyperalgesia is found in the fatty tissue below the skin on light pressure and touch and is made worse by tightly fitting clothes or even by showering. Also massage can be uncomfortable. Often pain is also felt in the skeletal system. The pain is temperature and weather dependent and usually reduces in dry heat and when pressure is high. Hot baths have a positive, but short-term effect; some patients, however, do

not tolerate heat. Problems with sexual relations may arise as a result of the very pronounced sensitivity of soft tissue. Reduced estrogen at the menopause does not reduce the pain.

Pain increases with the increase in fatty tissue. Generally the pain increases in connection with menstruation. Before the onset of the disease, there is usually only slight obesity, but in a short time overweight develops, usually 50 per cent over the normal weight for the age. Other patients have been overweight from an early age and have probably inherited the tendency for overweight. In some there is only localized fat, without general obesity.

Different types

Different types can be identified according to the spread of pain:

Type I or juxta-articular type, with painful folds of fat on the inside of the knees and/or on the hips, in rare cases only evident in upper-arm fat.

Type II or diffuse, generalized type, where widespread pain from fatty tissue is found, apart that of type I, also often in the dorsal upper arm fat, in the axillar fat, gluteal, in the stomach wall, in dorsal fat folds and on the soles of the feet.

Type III or lipomatosis, nodular type with intense pain in and around multiple "lipomas", sometimes in the absence of general obesity. Lipomas are approx. 0.5-4 cm, soft and are attached to the surrounding tissue. Histologically, these are not always encapsulated. Some have been classified as angioliipomas⁽¹⁸⁾.

In theory, the pain should be the result of lipomas/fat masses pressing upon nerves and through an axonal reflex, perhaps released pain-inducing factors like substance P. The pain may, for example, be eliminated locally by removing the offending lipoma. It should be mentioned that the lipomas in Madelung's syndrome, or multiple symmetrical lipomatosis, are not painful and are mainly found in the upper body, often cervically^(19, 20).

Fat distribution can be diffuse with generally painful fatty tissue ("Ruben's type of woman") or merely painful "riding breech fat". Sometimes a painful stomach can dominate.

Other symptoms, with variable incidence include:

Tendency to swell up, especially in the hands. The fingers become fumbling; tingling and numbness can occur (paresthesia). Compression of the median nerve is common.

General tiredness, worsened by light physical activity and poor sleep. The tiredness can be included under the term "chronic fatigue syndrome".

Tendency to bruising, possibly secondary to formation of "delicate vessels" in fat deposits. Coagulation tests are always normal. Telangiectasia is common.

Stiffness after resting, especially in the morning.

Headaches usually a combination of types (tension headaches, classic migraine, "neck" headaches) sometimes with pain in the jaw and the eyes, which is probably originated in retrobulbary fatty tissue.

Sometimes there is *cognitive dysfunction*, with variation in concentration and lapses of memory.

Bouts of depression ("atypical depression", possibly latent). This has nothing to do with the onset of the disease.

Feeling hot affects a small percentage of patients, some with a recurring high temperature of 37.5-39°C for weeks at a time, with increased pain and incapacity to work the result. The reason is unknown; investigation provides no explanation.

Susceptibility to infection, which may have a connection with the presence of fat⁽²¹⁾. Increased pain is often reported during infections or allergen responses.

Associated conditions

Associated conditions include sleep disturbances and Pickwick syndrome; slight to moderate dryness of the eyes and mouth with a gritty feeling in the eyes in spite of normal tear production (the criteria for Sjögrens syndrome are not completely satisfied); irritable bowel; coccygodynia; vulvovaginitis; vulvodinia; carpal tunnel syndrome; Tietzes syndrome; chondromalacia patellae; thyroid malfunction, mainly hypothyreosis; trochanteritis; localised tendinitis; sometimes onset of fibromyalgia; slight to moderate raising of cholesterol.

With increased obesity it is seldom that hypertension and diabetes type II arise.

Objective findings

Certain active parametres were changed, mainly with no disease present. An increase is seen in sedimentation rate⁽¹⁷⁾, alfa-1-antitrypsin, orosomuroid, haptoglobin, compliment factors C3, C4, Cl_q and Cl_s.

The fat cells are extremely large, and even larger than in healthy people of similar weight in controls. The heat produced by the fat cells, measured with a micro-calorie meter, is approximately twice as high as that taken from extremely overweight people. The ratio of monounsaturated fatty acid (16:1, 18:1) in the fatty tissue is greater than that of saturated fatty acid (14:1, 18:0) shown by a comparison with healthy people in controls⁽²²⁾.

In the cerebrospinal fluid the amount of substance P is significantly lower compared with healthy weight-matched individuals in controls. The average, however, in both cases is above the normal level (H. Brorson, B. Fagher, R. Ekman, unpublished data). In comparing averages with the average results obtained for fibromyalgia patients⁽²³⁾, they are less than half of these. The amount of the neuropetid Y is on the lower side of normal and B-endorphin on the higher side (H. Brorson, B. Fagher, R. Ekman, unpublished data). Fatty tissue histology shows a slight accumulation of perivascular lymphocytes and plasma cells⁽²⁴⁾, H. Brorson, B. Fagher, U. Stenram, unpublished data). Since the picture is not viral it is uncertain how these findings should be interpreted in this case. Another study suggests the occurrence of a metabolic block in the synthesis of lipids⁽¹¹⁾.

Therapy

Many patients find they are misjudged and require psychological support. Information about the disease is naturally important; any possible handicapped situation should be addressed, preferably with the assistance of an occupational therapist and social worker. Various aids may be needed in the home and at work.

Traditional analgesics have a poor effect. Paracetamol and dextropropoxifen are the first choice. Lidocaine given intravenously can for certain patients give total pain relief for some days^(25, 26). Because of side effects on repeated administration this form of therapy is in our opinion impossible to use.

Some patients may, because of troublesome swelling of the fingers, require diuretics.

Experience shows that lasting weight reduction by changing the diet is difficult to achieve and does not appreciably affect the pain. A gastroplastic operation may be recommended in order to avoid metabolic complications. Unfortunately, the pain remains largely unchanged, but quality of life is usually improved after this operation.

Operation for isolated painful lipomas which are pressing and causing numbness and tingling brings relief. Localized pain may sometimes be treated with cortisone/anaesthetic injection, alternatively with sterile water given intracutaneously or more deeply.

Liposuction in order to reduce pain was recommended in a prospective study by our group in 1989^(27, 28). This method of treatment has now been tried in a research project in Lund-Malmö and has shown in objective tests a good initial and significant reduction of pain, which however reduces over time. An improved quality of life is seen for sufferers with general lower-body fat or more localized large deposits of fat at the knees, on the arms, thighs or stomach (abdominal plastic). With general diffuse pain liposuction is major and must then be considered a risky operation, requiring care for about a week in the plastic surgery department.

The method in the present case can only be regarded as supportive treatment. Any skeletal pain is not affected and over a long period of time can become more insistent in spite of reduction in pain in the fatty tissue. If fibromyalgia is also an underlying problem, liposuction is not recommended. Long-term follow-up (five years) of the operation material is ongoing, the result of which will determine how many resources will be allocated in future.

We recommend to our patients that they avoid monotonous, static work, also physical and psychological stress.

Society

There is an association, the Dercum Group, formed in 1990 in Lund. The association, part of the Association for Rheumatics, is nationwide and has approx. 300 members. It works for support and information to both individual members and other interested parties. *Address: The Dercum Group, Association for Fatty Tissue Rheumatics, P.O. Box 1251, 221 05 Staffanstorps.*

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