

Adiposis Dolorosa: An Unique Case of Rare Disease with Review of Literature

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Abstract: Adiposis Dolorosa (Dercum's disease) is an unusual progressive syndrome due to unknown etiology characterized by multiple painful lipomas that arise in adult life, most often affecting post menopausal women who are obese. We hereby report Type III Dercum's disease in young, pre menopausal, non obese patient of the rarity of this condition, along with review of literature.

Keywords: Adiposis dolorosa, Dercum's disease, Obesity

INTRODUCTION

Dercum's disease was first described in 1892 by Francis Xavier Dercum [1]. It is more common in postmenopausal women and is characterized by painful subcutaneous lipomas or fat accumulations largely on the trunk and limbs [2]. There is also associated asthenia, easy bruising over the affected areas and hypercholesterolemia [3]. Dercum's disease is believed to be transmitted in an autosomal dominant manner [4, 5], however most reported cases of adiposis dolorosa are sporadic [6]. 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. Treatment consists of a combination of medical and surgical therapies along with rehabilitation maneuvers.

Different types can be identified according to spread of pain:

- Type I, or the juxta-articular type,
- Type II, or the diffuse, generalized type,
- Type III, or the lipomatosis, nodular type, with pain in and around multiple lipomas, sometimes in the absence of general obesity; lipomas are approximately 0.5-4 cm, soft, and attached to the surrounding tissue.

CASE REPORT

A 34 year old married normotensive non-obese (weight 45 kgs) woman reported to the outpatient department with 2 years history of multiple, nodular, painful sub-cutaneous swellings distributed mainly over left arm and forearm (Fig.1). These swellings first appeared in the left forearm with a single lesion which progressively increased in size with the appearance of similar lesions over the arm. Hyperalgesia was found in the fatty tissue below the skin on light pressure and touch with exacerbation on exposure to cold and on exertion. Pain did not increase in connection with menstruation and menstrual cycles were normal. There is history of paresthesias and swelling of hands. There is no history of morning stiffness, tiredness, headache, depression, loss of sleep and appetite or bruising tendency. There is no history of oral contraceptive pills use or intake of oral steroids. There is no history of painful subcutaneous swellings over body in her siblings. No other significant relevant history present.

On examination; multiple, tender, freely mobile, soft to firm in consistency, sub-cutaneous nodules of sizes varying from 0.5-15 cm were seen on left forearm, arm on both extensor and flexor aspects.

These swellings (Fig. 2) were not adherent to the overlying skin which did not show any surface changes.

Routine investigations on blood including complete blood count, liver, kidney and thyroid function tests were within normal limits. Coagulation tests and erythrocyte sedimentation rate were also within normal limits. Fine needle aspiration cytology (FNAC) revealed the lipomatous origin of these lesions with normal capsule formation.

On the basis of typical clinical features and investigations in this non-obese female s diagnosis of Type III Dercum's disease was made.

She was subjected to surgery under general anaesthesia. Multiple incisions were given to remove the tumors. Some of the tumors were intramuscular but all were smooth and encapsulated (Fig. 3). Total of twenty lipomas were removed (Fig. 4). Few swellings showed cystic degenerative changes (Fig. 5). Histopathology reveals lipomas with well developed capsule.



Fig. 1: Pre-Operative: Multiple Lipomas on Left Upper Limb



Fig. 2: Multiple Lipomas: showing no pressure effect



Fig. 3: Per-Operative: Intramuscular Lipoma being removed

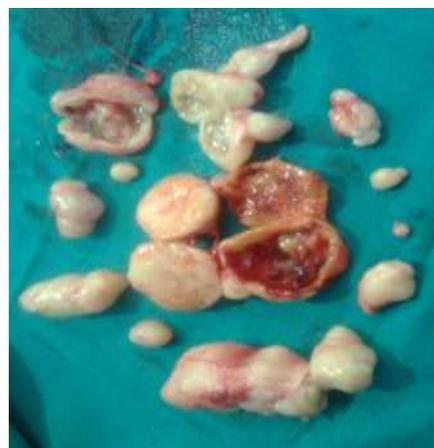


Fig. 4: Specimen of Total Lipoma Excised (no. = 20)



Fig. 5: Excised Lipomas showing solid and cystic areas

DISCUSSION

Dercum's disease discovered in 1892, is included in the rare disease category by the World Health Organisation [7]. It has been more seen in

postmenopausal women. It is characterized by multiple painful lipomas over the trunk and limbs, disproportionate weight gain and generalised obesity. Paresthesia is commonly associated. To our knowledge the case we report is the first one in our region. Depression has been reported in most of the cases along with memory lapse and sleep disturbances [8]. Our patient despite having other features, did not reveal such symptoms. Mastalgia, though not seen in our case has been reported by Trentin *et al.* [9].

Diagnosis of adiposis dolorosa is based on clinical features [9] and can be supported by other diagnostic methods like histopathology and MRI. The pathogenesis of the disease is unknown, therefore, the treatment is not very helpful [10].

Traditional management of Dercum's disease has been largely unsatisfactory relying on weight reduction and surgical excision of particularly troublesome lesions [12].

Non-pharmacological approaches for Dercum's disease may be used as adjuncts to pharmacological treatments. Some of these include acupuncture, cognitive behavioral therapy, hypnosis and biofeedback [6].

Treatment options include removal of fat by surgical means including liposuction, which has shown to improve quality of life in certain cases [11]. The reported case was managed surgically in view of large size tumors and pain was severe.

CONCLUSION

We are reporting a case of Dercum's disease managed by surgery. This is a rare case of rare disease because our patient was young, premenopausal, non-obese, no menstrual irregularity, without psychological disturbances with short history of progression.

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