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Case Report

Dercum's disease: a case report

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Abstract Dercum's disease is a chronic progressive disorder of unknown etiology, characterized by multiple, painful, subcutaneous lipomas. We report two cases of this rare disease. The clinical, histopathological and imaging findings supported the diagnosis.

Key words
Dercum's disease, adiposis dolorosa, 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] The pathogenesis of this disease is still unknown. Though treatment consists of a combination of medical and surgical therapies along with rehabilitation maneuvers, it has not yet proved very successful.

Case reports

Case 1

A 35-year-old female presented with diffuse swelling of upper and lower limbs, as well as lower trunk, progressive for the past 12-13 years. There was also complaint of severe pain in the legs and difficulty on walking. On examination, there were multiple, soft swellings of both upper and lower limbs, as well as lower trunk (**Figure 1**). There was sparing of hands and feet (**Figure 2**). The overlying skin had a reticulate pattern at the anteromedial aspects of both calves just below the knees (**Figure 3**). There was no history of any mental disturbance. The systemic review was insignificant. There was no family history of obesity. The routine investigations were normal. The lipid profile was deranged, with serum cholesterol 390mg/dl, serum triglycerides 347mg/dl, HDL 48mg/dl and LDL 210mg/dl. Histopathology showed excess fat deposition in the subcutaneous tissue. MRI revealed excessive bilateral symmetrical fat deposition with relative enlargement of lymphatics. Arteriovenous Doppler of both upper and lower limbs was normal with focal cellulities (soft tissue) at medial aspect of right knee. There was also subcutaneous extensive symmetrical fat deposition in all the limbs.

Case 2

A 35-year-old female presented with sudden onset of weight gain over 3-4 years starting from the legs and involving the arms and the upper trunk (**Figure 4**). There was also complaint of burning sensations over the arms and legs.

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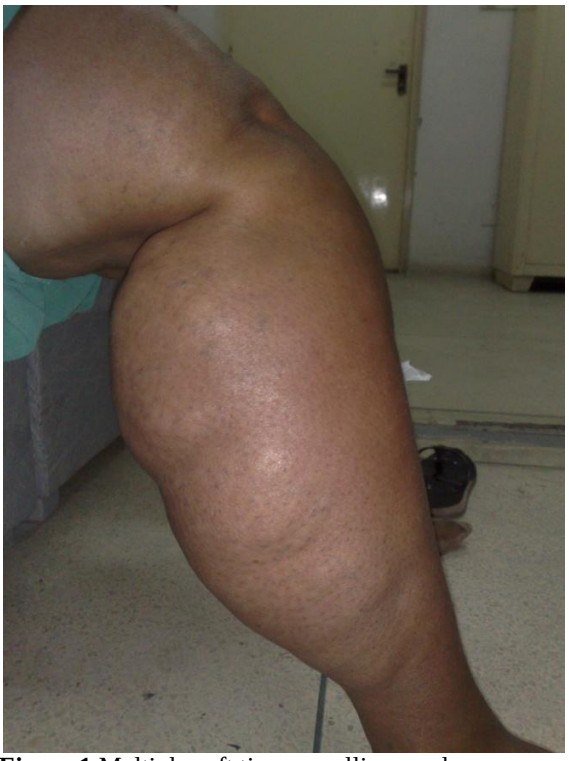

Figure 1 Multiple soft tissue swellings on leg.

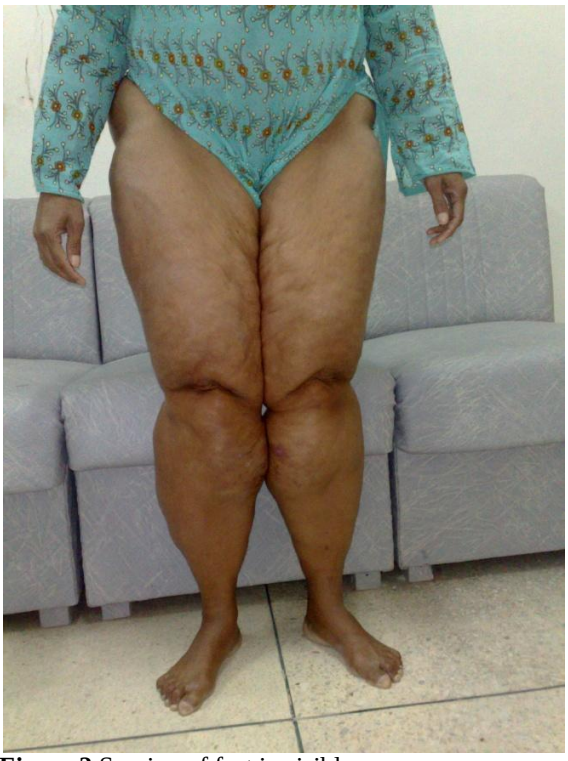

Figure 2 Sparing of feet is visible.

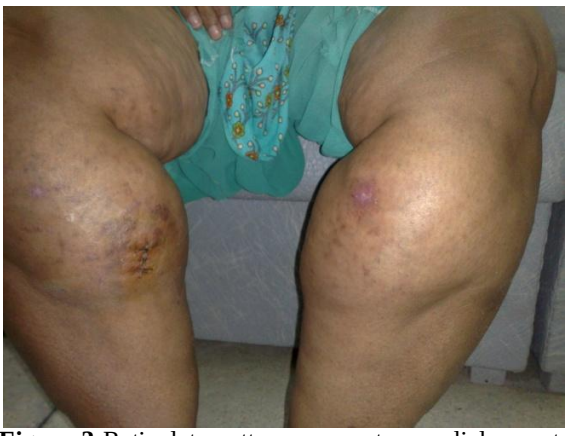

Figure 3 Reticulate pattern over anteromedial aspect of thighs.

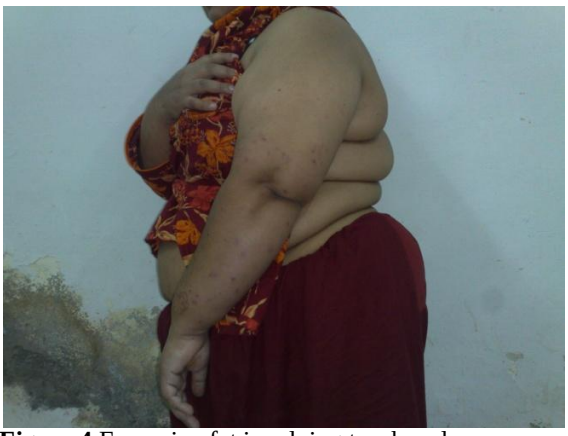

Figure 4 Excessive fat involving trunk and arm.

Along with this there was complaint of weakness, lethargy and decrease in appetite. There was no complaint of psychiatric disturbance or any other significant systemic complaint.

On examination there were firm, mobile painless lumps over legs and arms. Routine investigations were normal. Serum lipid levels were normal. Ultrasound revealed a fatty liver. Histopathology showed mild inflammation in the dermis with underlying excessive fat deposition. MRI revealed lipomatous masses in all the limbs.

Discussion

Dercum's disease discovered in 1892, is included in the rare disease category by the

World Health Organization.[4] It has been more commonly seen in postmenopausal women. It is characterized by multiple painful lipomas over the trunk and limbs, disproportionate weight gain and generalized obesity. Paresthesia is commonly associated.[4] To our knowledge the cases we report are the first ones in our setup. Depression has been reported in most of the cases along with memory lapse and sleep disturbances.[5] Our patients despite having other features, did not reveal such symptoms. Mastalgia, though not seen in our cases has been reported by Trentin *et al*.[6]

One of our cases had sparing of the ankles as reported by Amine *et al*.[4]

Diagnosis of adiposis dolorosa is based on clinical features[6] 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.[7] The patients can be advised to achieve weight loss with exercise and diet but these do not work in most cases. Patients suffering from pain can be helped with drugs such as mexiletene as well as intravenous lidocaine. Treatment options include removal of fat by surgical means including liposuction, which has shown to improve quality of life in certain cases.[8]

Our patients were managed symptomatically. The first case was also started on lipid lowering drugs. They were later referred to the plastic surgery department for further management.

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