CASE REPORT

Unusual presentation of Hepatitis C with Dercum's disease (Adiposis dolorosa) and Lichen planus

Hafiz Abdul Moiz Fakih, Sina Aziz, Mohammad Masroor

Abstract

Dercum's disease (Adiposis dolorosa) is a rare progressive syndrome, of unknown etiology, characterized by multiple painful lipomas that arise in adult life, most often affecting postmenopausal obese women. There is a statistically significant high prevalence of positive Hepatitis C Virus RNA in patients with Lichen planus, suggestive of an etiological role of HCV in the pathogenesis of Lichen planus. Here, we report a rare case of a 50 year old postmenopausal obese woman with Hepatitis C, Dercum's disease and Lichen planus who presented in the outpatient setting of Sarwar Zuberi Liver Centre, Medical Unit 4, Civil Hospital Karachi.

Key Words: Adiposis dolorosa, Hepatitis C, Lichen Planus.

Introduction

Adiposis dolorosa, first described by Dercum in 1892, is a rare disease of subcutaneous fat characterized by localized overgrowth of fat.[1] It is commonly seen in postmenopausal obese females and is often associated with fatigue, weakness and severe emotional disturbances. The disease does not respond to routine analgesics but only to intravenous lidocaine.[2]

Lichen planus is a chronic mucocutaneous disease that affects the skin and the oral mucosa, and presents itself in the form of papules, lesions or rashes. A high prevalence of HCV infection has been found in patients with lichen planus, supporting a possible relationship between lichen planus and hepatitis C.[3,4]

Case Report

We present the case of a 50 year old, postmenopausal female, who was referred to the Sarwar Zuberi Liver Center, DUHS and CHK for the management of recently diagnosed Hepatitis C. She came to know of this diagnosis during the regular screening blood tests for a lump about 2 years back. According to her, she had consulted many General Practitioners and Dermatologists for that lump, which was in the upper part of the right forearm. She had first noticed it about 10 years back. Initially, it was soft, slightly tender, non-itchy, neither red nor warm, about the size of a pea. The lump gradually increased in size and reached the size of a lemon. She also developed similar lesions in the elbow region of limbs, left forearm, abdomen and the femoral region. Patient had also complaints of cold peripheries, discoloration, rash, itching throughout the body, dizziness, insomnia and abdominal pain. She felt that all the energy had been taken out of her body and used to fatigue easily after short activities. There was no history of epilepsy, jaundice, transfusion or surgery. Patient did not take any routine medications. Father was hypertensive and mother a diabetic. No family history of similar lumps.

On examination, middle aged lady of average height and obese built, with Body Mass Index of 25.5 kg/m^2, looking apprehensive, low tone speech, down in mood, co-operative, well ori-

Medical Unit 4, DUHS & Civil Hospital, Karachi
HAM Fakih

Sarwar Zuberi Liver Center, Medical Unit 4, DUHS & Civil Hospital, Karachi
S Aziz

Medical Unit 4, DUHS & Civil Hospital, Karachi
M Masroor

Correspondence:
Hafiz Abdul Moiz Fakih,
43, Kokan Society,
Shaheed-e-Millat Road,
Karachi-74800
4853158, 021-2010142,
0300-2933883.
caddick_1@yahoo.co.uk

Figure 1: *Two lumps in the lateral aspects of right elbow regions*

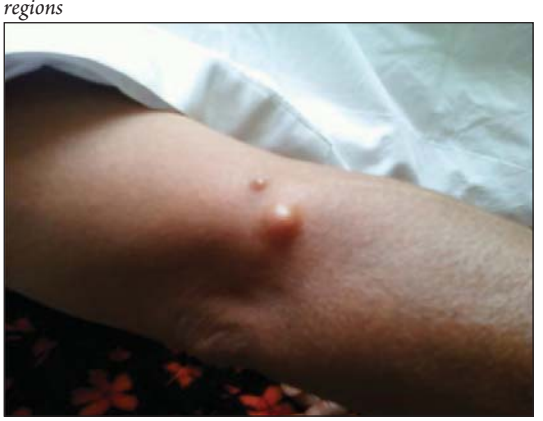


Figure 2: *A lump in the medial aspect of left ankle joint*

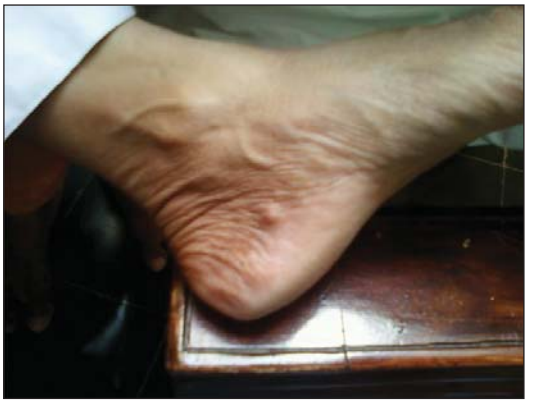


Figure 3: *Lichen planus. Histology section showing epithelial hyperkeratosis, mild acanthosis, and basement layer lichenification, also present is a layer of lymphocyte infiltrate, immediately underlying the epithelium.*

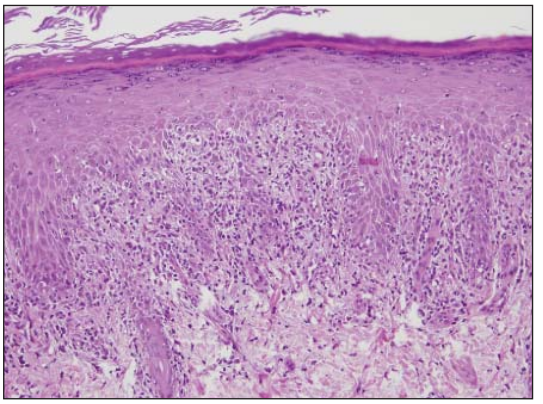
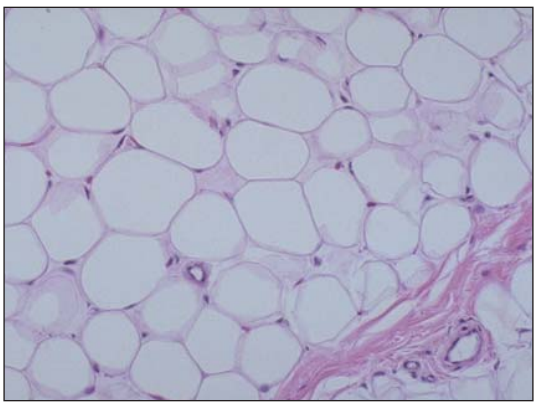


Figure 4: *Lipoma. Histology section of the lump showing normally appearing adipose tissue. There is a thin cell membrane that bounds the cytoplasmic lipid, which appears clear in this section because normal tissue processing removes lipids. The cell nucleus is pushed to one side by the lipid. Connective tissue septae divide lobules of fat and carry the vascular supply.*



ented to time, place and person. Temperature 98.6 F, respiratory rate 20/min, pulse rate 108/min, blood pressure 130/80 mmHg, mildly anemic, Thyroid examination was normal. Skin was smooth with no skin or mucosal hyper-pigmentation.

Examination showed that there were multiple lumps throughout the body. Each lump ranged form 1-3 cm in diameter, spherical, soft, smooth surface, and mobile, showing no signs of inflammation, tender on application of deep pressure, transillumination test was negative and no visible pulsation or scar was present. Slipping sign was positive.

Examination of the skin also showed multiple, red to purple, raised, polygonal rashes, around which itch-marks were present. Such rashes were present in patches throughout the body.

Laboratory workup revealed the following: Hemoglobin 11 gm/dl, Red blood cells 3.97 x 106/mm^3, White blood cells 7.1 x 103/mm^3, Hematocrit 35.3 %, Platelet count 187x10^3/mm^3, Fasting Glucose 86 mg%, Urea 22 mg%, Creatinine 0.6 mg%, Serum ALT 59 U/l (normal upto 45 U/l), Total protein 8.3 Gram%, Albumin 4.1 Gram%, PT 15/15, APTT 37/36, TSH 3.74, FT3 2.24 pgm/ml, FT4 0.90 ng/dl, Ultrasound abdomen showed moderate fatty infiltrates in the liver, HepBsAg-Non-reactive, Anti HCV-Reactive, HCV PCR positive (genotype 3.). A biopsy specimen was taken from one of the lumps . Also a skin specimen was also taken from one of the rashes. Both were subjected to histopathology. Histopathology of the lump was consistent with the findings of lipoma while the skin rash histopathology was suggestive of Li-

chen planus.

She took 3 Interferon (INF) injections per week for 3 weeks along with capsule ribavirin 400mg thrice a day for the treatment of Hepatitis C, but treatment had to be stopped it because she could not tolerate it. For Lichen planus, a dermatology consultation was done and she was given clobetasol propionate cream for local application on the rashes twice a day and tablet cetrizine hydrochloride once daily for pruritis.

Discussion

Dercum's disease (also known as Adiposis dolorosa), included in the category of rare diseases by WHO, is a clinical syndrome characterized by four cardinal symptoms: 1. multiple, painful, fatty masses; 2. generalized obesity, usually in menopausal age; 3. asthenia, weakness, and fatigability; and 4. mental disturbances, including emotional instability, depression, epilepsy, confusion, and dementia.[5] Our case fulfilled all the clinical criteria.

HCV is a pathogenetic factor in mixed cryoglobulinemia, membranoproliferative glomerulonephritis, lichen planus, autoimmune thyroididtis, porphyria cutanea tarda.[6] No association between HCV and Dercum's disease has yet been reported in literature. However, our patient, a known case of Hepatitis C presented with Dercum's disease and Lichen planus.

Dercum's disease usually occurs in females aged 45 to 60 years, the condition being 20 times more frequent in females than in males. The cause of the disease is currently unknown but an autosomal dominant pattern of inheritance has been suggested.[7] Our case had no family history of similar disease.

The presenting symptom is usually pain in the fatty lumps lasting for at least three months.[8] Our patient described the pain as mild aching but in other studies it has also been described as stabbing, smarting or burning of varying intensity. Swellings may occur in many areas of the body,[5] most commonly affecting the knees, trunk, forearms and thighs. This case had multiple swellings in upper and lower extremities and abdomen.

The disease has to be differentiated from Madelung syndrome, multiple familiar lipomatosis and Proteus' syndrome.[9] A diagnosis of Dercum's disease is clinical as the investigations are normal and the histopathology of the lumps do not reveal any significant features that might distinguish the tumors from the common sporadic lipomas. A biopsy was performed in our case which confirmed that the lumps were lipomas. Also the biopsy of the skin rash reported findings of acanthosis, epidermal colloid bodies, and lichenoid mononuclear dermal infiltrate confirming the diagnosis of Lichen planus.

In 2 reported cases, interferon (INF) alfa-2b induced long-term relief of pain in 2 patients with Dercum's disease and chronic hepatitis C. The analgesic effect of INF therapy occurred 3 weeks after treatment with 3 million units, 3 times per week, for 6 months. Whether the mechanism of pain relief with INF is related to its antiviral effect, to the production of endogenous substances (e.g., endorphins produced by INF), or to the interference of INF with interleukin 1 and tumor necrosis factor-alpha cytokine production, which are involved in cutaneous hyperalgesias, remains unclear.[10] In our case, after 3 weeks of therapy with 3 injections of INF per week, the treatment had to be stopped because according to the patient she could not tolerate the injections. Furthermore, she did not get any relief from pain.

Conclusion

Though there are many case reports in literature on Dercum's disease, but to the best of our knowledge Dercum's disease with Hepatitis C and Lichen planus, as seen in our case, has not been reported yet. Therefore, a search for HCV infection should be systematically performed in patients with Dercum's disease and chronic Lichen planus.

References:

1. Black MM, Cunliffe WJ. Dercum's disease. In: Champion RH, Burton JL, Burns DA, Breathnach SM eds. Textbook of Dermatology. Blackwell Scientific Publications, Oxford, 1998, pp2434-2435.
2. Palmar ED. Dercum's disease: adiposis dolorosa. Am Fam Physician 1981; 24: 155-157.
3. Imhof M, Popal H, Lee JH, Zeuzem S, Milbradt R. et al. Prevalence of hepatitis C virus antibodies and evaluation of hepatitis C virus genotypes in patients with lichen planus. Dermatology. 1997;195(1):1-5.
4. Ghodsi SZ, Daneshpazhooh M, Shahi M, Nikafarjam A. Lichen planus and Hepatitis C: a case-control study. BMC Dermatol. 2004 May; 4:6.
5. Brodovsky S, Westreich M, Leibowitz A, Schwartz Y. Adiposis dolorosa (Dercum's disease): 10-year follow-up. Ann Plast Surg. 1994 Dec;33(6):664-8.
6. Friedman LS. Liver, Biliary Tract & Pancreas Disorders. In: McPhee SJ, Papadakis MA, eds. Current Medical diagnosis and Treatment. The McGraw-Hill Companies Inc, 2009, pp588.
7. Campen R, Mankin H, Louis DN, Hirano M, Maccollin M. Familial occurrence of adiposis dolorosa. J. Am. Acad. Dermatolo 2001 Jan; 44(1):132-6.
8. Moi L, Canu C, Pirari P, Mura MN, Piudu G, Del Giacco GS. Dercum's disease: a case report. Ann Ital med Int. 2005 Jul-Sep; 20(3):187-91.
9. De Villiers AC, Orange AP. Treatment of pain in adiposis dolorosa (Dercum's disease) with intravenous lidocaine: a case report with a 10-year follow-up. Clin. Exp. Dermatol. 1999 May; 24(3):240-1.
10. Gonciarz Z, Mazur W, Hartleb J, Machniak M, Bednarek I, Mazurek U, et al. Interferon alfa-2b induced long-term relief of pain in two patients with adiposis dolorosa and chronic hepatitis C. J Hepatol. 1997 Dec;27(6):1141.