CORRESPONDENCE

Corticosteroid-induced Dercum's disease

Case report

A 37-year-old woman presented with progressive generalized tightening of the skin affecting the face, upper back and proximal arms for 6 years. She was initially diagnosed as scleroderma at the rheumatological department. Because of poor treatment response to cyclophosphamide and methylprednisolone, skin biopsy was performed in the dermatological department. Based on presence of irregularly oriented and separated collagen fibers with mucin deposition in the dermis, scleromyxedema was diagnosed. She received intravenous immune globulin, 24 g/day for 5 consecutive days, and intravenous methylprednisolone, 500 mg/month for 4 months. Subsequently, prednisone (20 mg/day) was given for maintenance. Her scleromyxedema improved gradually. However, during the course of 1 year there was a gradual onset of painful subcutaneous masses developing on the bilateral upper arms, and her weight increased from 54 kg to 70 kg. The pain was stabbing without radiation and persisted almost all day. The patient also complained of muscle weakness and fatigability, and the clinical impression at that time was musculoskeletal involvement of scleromyxedema. In addition, the patient developed depression, insomnia, and mood irritability, and was diagnosed with major depressive disorder. Oral quetiapine 275 mg, mirtazapine 60 mg, and zolpidem 20 mg at bedtime were added to her treatment regimen. Family history was negative for lipomatosis.

On physical examination, soft, well-circumscribed subcutaneous masses with nodular swelling were found on both upper arms (Figure 1). In addition, tachycardia (112 beats/minute) and abdominal bloating without shifting dullness were noted. Neurological examination was unremarkable. Laboratory investigations revealed a white blood cell count of 10.7 x 10^9/L (normal range = 4.0 x 10^9–10.0 x 10^9/L) with 82% segments (normal range = 42–74%). Fasting blood glucose, renal function tests, liver function tests, serum lipids (cholesterol and triglyceride), free T4, thyroid stimulating hormone, and adrenocorticotropic hormone were all within normal limits. Soft tissue ultrasound over the upper arms showed adipose tissue hypertrophy (Figure 2A).

We performed incisional biopsy on her right upper arm. The pathology revealed a lobulated lipoma composed of univacuolated mature adipocytes. The following features such as inflammation, abnormal vascularity, necrosis, fibrosis, or microthrombi were not found (Figure 2B). Based on clinical history and pathological findings, corticosteroid-induced Dercum's disease (adiposis dolorosa) was diagnosed.

Discussion

Dercum's disease is a rare disorder characterized by diffuse or localized painful fatty deposits, occurring primarily in postmenopausal, obese women of middle age.1,2 Fatty deposits can be found in almost any location. There are three subtypes of Dercum's disease: juxta-articular, diffuse/generalized, and lipomatosis (nodular). The essential features of Dercum's disease include four cardinal symptoms: (1) multiple, painful, fatty masses; (2) generalized obesity; (3) weakness and fatigability; and (4) mental disturbances, which can include emotional instability, depression, epilepsy, confusion, and dementia.3,4 Pain can be continuous or paroxysmal, and worsens with movement. Most reported cases of Dercum's disease are sporadic, but there is some evidence that it can be transmitted as an autosomal dominant trait with incomplete penetrance.4 The etiology of this disease remains unknown. Dercum's disease is a complex and underdiagnosed painful syndrome that may affect

Figure 1 (A) Nodular swelling of bilateral upper arms near shoulders. (B) Close-up lateral view.

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multiple systems, and some authors suggest that exercise induces pain in Dercum’s disease without having any effects on the size of the fatty lesions.5

Dercum’s disease is a diagnosis of exclusion. Differential diagnosis includes other lipomatoses, Cushing’s syndrome, and fibromyalgia.2,4 Madelung’s disease, familial lipomatosis syndrome, and congenital lipomatosis can be differentiated from Dercum’s disease by the lack of pain associated with the lipomas. The adrenocorticotropic hormone level is low in Cushing’s syndrome but normal in Dercum’s disease, and pain and nodular swelling are not typical signs of Cushing’s syndrome. Pain in Dercum’s disease is associated with the fatty deposits, which is not observed in fibromyalgia.

To the best of our knowledge, only one case of corticosteroid-induced Dercum’s disease has been reported in a 67-year-old female who suffered from eosinophilia–myalgia syndrome.6 Despite its rarity, the development of Dercum’s disease during the course of treatment for scleromyxedema deserves more attention. It may be misdiagnosed as Cushing’s syndrome or be mistaken for musculoskeletal involvement of scleromyxedema, as 90% of scleromyxedema patients report arthralgia, myalgia, or muscle weakness.7,8 Early diagnosis of Dercum’s disease is imperative because of the difficulties in effective treatment.

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