Cutaneous Chylous Bubbles and Lymphatic Pilar Reflux in a Patient with Juxta-articular Dercum’s Disease: A Very Rare Case

The Editor,

Sir,

Dercum’s disease is a rare disease that is characterized by generalized overweight or obesity, multiple, painful fatty masses and a number of associated symptoms. The disease can be classified as Type 1 (juxtaarticular), Type 2 (diffuse-generalized) and Type 3 (nodular). The pain is chronic (for more than three months), symmetrical and often disabling. The other cardinal symptoms are weakness and susceptibility to fatigue, and psychiatric manifestations such as emotional instability, depression, epilepsy, confusion and dementia. The prevalence of Dercum’s disease has not yet been exactly established and the condition mainly affects postmenopausal women. Diagnosis is based on clinical criteria and should be made through exclusion of differential diagnoses. Still, there is no convenient treatment for the disease (1).

On the other hand, the cutaneous lesions associated with reflux of lymphatic fluid and chyle from the skin have been reported in the dermatologic literature very rarely (2). A swollen extremity with cutaneous chylous vesicles is a common manifestation of chylous reflux (3). A triglyceride level > 110 mg/dL is diagnostic for chyle leak (4). Histopathologically, there are subepidermal blister formation and dilated lymphatics in the dermis (5).

We report the case of a 76-year old Caucasian woman with Dercum’s disease who has cutaneous lymphatic vesicles, bullae and cutaneous pilar chylous reflux in the lower extremities and suffers from severe depression. According to the history, both legs of the patient began to swell seven years ago. Thereafter, the swellings gradually increased and became painful. Three months previously, several bubbles appeared on the skin of the swollen legs and the genital region. One month ago, a milky-fluid discharge exuded from these bubbles. The patient complained of severe pain in the buttocks and legs, weakness, fatigue and constant leakage of the bubbles. There was no history of additional systemic or localized diseases (including the genitourinary system) except for the controlled hypertension (for four years) and family history of the disease. The patient did not use any drug except for furosemide therapy (40 mg/per night) for four years. Physical examination was significant for detection of excessive swellings in both lower extremities, buttocks (Fig. 1a, b) and vulva.

![Fig. 1: Clinical appearance of the lesions – (a, b)- Excessive swellings in the lower extremities; (1c-e): pendulous masses which were located around the large joints; (f): closer view of the multiple, translucent or milky-white, small vesicles and a few bullae which have leaked out of the skin.](image)

The swellings were in the form of pendulous masses and they were located around the large joints (hip, knee and ankle) [Fig. 1c-e]. On the skin overlying the swellings and vulva, there were multiple, translucent or milky-white, small vesicles and a few bullae which had leaked out of the skin (Fig. 1f). Psychiatric examination revealed severe depression and anxiety. The haematological and biochemical tests (including complete blood count, erythrocyte sedimentation rate, C-reactive protein, glucose, urea, creatinine, lipids, triglyceride, electrolytes, thyroid and liver function tests, anti-thyroid antibodies, antinuclear antibody, rheumatoid factors, tumour markers and hormones) were negative or within the normal limits.
Bio-chemical analysis of the aspiration material of the bubbles showed that its triglyceride value was 145 mg/dL. Arterial and venous Doppler angiography revealed only minimal venous insufficiency. On lymphoscintigraphy of the lower extremities and abdominopelvic computed tomography, there was no pathology. Skin punch biopsy of vulvar vesicles showed orthokeratosis of the epidermis, dilated lymphatics, moderate oedema, a few lymphocytes and macrophages in the dermis and mature lipocytes in the subcutaneous fatty tissue (Fig. 2a).

Fig. 2: Histopathology and immunohistochemistry of the vulvar vesicles; (a) A basket-style orthokeratosis at the epidermis, subepidermal vesicle immediately adjacent to the dilated lymphatic channels under the epidermis (× 200 H&E); (b) Luminal staining of lymphatic endothelium with D2-40 antibody (× 100).

Immunohistochemically, the lymphatics reacted with D2-40 monoclonal antibody that is a selective marker of lymphatic endothelium [Fig. 2b] (6). The histopathologic diagnosis of the lesion was made as cutaneous chylous vesicle. The vesiculobullous lesions and chylous pilar reflux of the patient completely improved with repeated topical astringent dressings (with 2% lead-subacetate solution, three times a day) after six weeks. However, both of them recurred after two weeks.

Although the exact aetiology of the disease is unknown, the disease is attributed to endocrine dysfunction (such as thyroid, hypophysis, sex glands, adrenal glands and pancreas), nervous system functions, mechanic pressure on the nerves, adipose tissue disfunctions, inflammation and trauma. In the differential diagnoses of Dercum’s disease, lipoedema, familial multiple lipomatosis, adipose tissue tumours, panniculitis and endocrine disorders should be considered (1). Additionally, vesiculobullous chylous reflux should be distinguished from cutaneous blistering diseases (5). We did not detect such disorders in our patient.

In conclusion, we thought that the lymphatic bubbles and pilar chylous leakage might have been due to the compression to the lymphatics of the lipomatous masses. The patient, we presented was reported because of the rarity of the disease and its coexistence with cutaneous chylous reflux and chylous bubbles. To the best of our knowledge, this patient is the first reported case of Dercum’s disease with cutaneous pilar chylous reflux and lymphatic bubbles.

Keywords: Cutaneous chylous bubbles, juxtaarticular dercum’s disease, lymphatic pilar reflux

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