A Case of Benign Symmetric Lipomatosis

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Benign symmetric lipomatosis is characterized by diffuse symmetric deposits of nonencapsulated fat, generally affecting the cervical and upper dorsal regions. There is a history of alcohol abuse in nearly all the cases. It is rarely reported in non-drinkers. Herein we report a 63-year-old man presenting with one year's history of multiple ill-defined symmetric soft mass on posterior neck, upper trunk and upper extremities without the history of alcohol abuse. We administered corticosteroid orally due to osteoarthritis of the knee for a long time.
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Key Words : Benign symmetric lipomatosis, Corticosteroid

Benign symmetric lipomatosis, also called Madelung's disease or Launois-Bensaude syndrome, is an uncommon disorder of unknown etiology but may well be underdiagnosed. It is characterized by non-encapsulated accumulations of fat in a symmetrical manner around the neck and shoulders, rarely involving the lower limbs and the lower body[1]. It affects middle-aged males from the Mediterranean area, usually with alcohol abuse or chronic liver disease[2]. 13 cases have been reported in Korean literatures[3,4]. All the cases except for 4 cases were associated with alcohol abuse. We report a 63-year-old man presenting with symmetric lipomatosis on the upper torso with the history of prolonged administration of corticosteroid.

CASE REPORT

A 63-year-old man presented with gross symmetric progressive enlargement of the upper portion of his body that started 1 year ago on both shoulders and the neck. There were no associated symptoms such as tenderness and pruritus. He had been treated with oral glucocorticoids due to osteoarthritis of the knees at a local clinic during the last 3 years. He had a 40-year history of smoking and denied any alcohol abuse. He showed symmetric and ill-defined enlargement of the shoulders, neck, upper part of the back and abdomen, so he appeared to be muscular(Fig. 1). The distal extremities were spared and even looked atrophic. Palpation disclosed a moderately soft consistency similar to that of fatty tissue. The overlying skin was of normal appearance. Histological examination from the right shoulder revealed normal epidermis and dermis, and increased numbers of normal adipocytes without encapsulation in the subcutaneous tissue(Fig. 2). Results of laboratory examination revealed normal complete blood count, serum electrolyte levels, liver function test and urinalysis. Serum cholesterol was 146 mg/dl, triglycerides 54 mg/dl, and blood glucose levels were 140 mg/dl. Serum cortisol was 1μg/dl, adrenocorticotropic hormone(ACTH) 11.8 pg/ml, and urine cortisol was 3.3μg/day. Chest magnetic resonance image(MRI) showed no abnormality. A diagnosis of benign symmetric lipomatosis coexisting with iatrogenic Cushing's syndrome was made. He was treated with 10 mg prednisolone daily and regularly followed-up without any treatment for lipomatosis.

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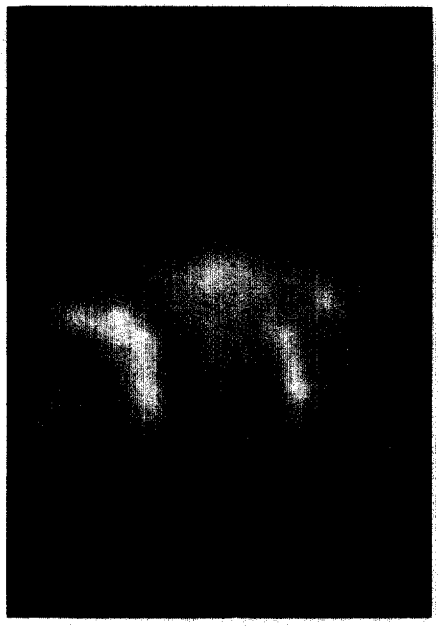

Fig. 1. Multiple symmetric subcutaneous enlargements on the upper arms, the neck and the trunk.

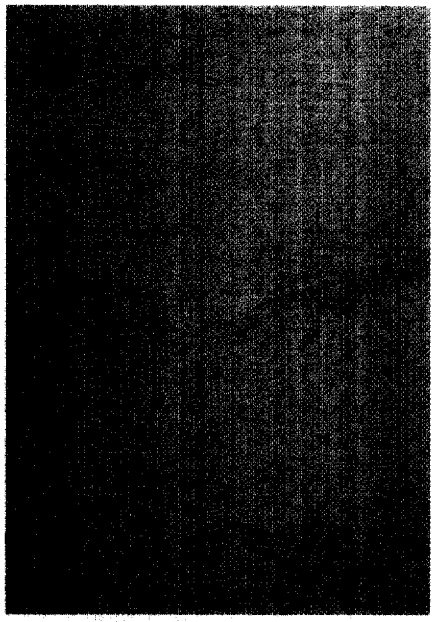

Fig. 2. Diffuse distribution of normal lipocytes in the subcutaneous tissue without encapsulation(H&E, ×40).

Table. Classification of lipomatosis

1. Multiple symmetric lipomatosis (Madelung's disease)
Type I (Madelung's collar)
Type II
2. Mediastino-abdominal lipomatosis
3. Pelvic lipomatosis
4. Epidural lipomatosis
5. Adiposis dolorosa (Dercum's disease)

(cited from ref.1)

DISCUSSION

The term 'lipomatosis' was coined to refer to a disorder characterized by multiple, non-encapsulated lipomas affecting various areas. A classification of lipomatosis is shown in Table 1[1]. Benign symmetrical lipomatosis(BSL) represents an unusual subgroup of lipomatous disease. Initially described in 1846 by Brodie, in 1888 by Madelung, and in 1898 by Launoise and Bensaude, this condition is variably known as Madelung's syndrome, Launoise-Bensaude adenolipomatosis, multiple symmetric lipomatosis[5]. Fat deposition in BSL occurs on the neck, shoulder girdle and upper arms, giving the upper part of the body a pseudoathletic appearance in contrast to the rather normal lower part[1,2,5]. The face, lower arms, hands, legs and feet are always spared[2,5]. Adults 30-60 years of age are mainly affected with a male-to-female ratio of 15:1[2]. BSL has been classified clinically into two types[5,6]. In type I, the fatty deposits are circumscribed, non-encapsulated masses protruding from the body surface and symmetrically distributed on the upper part of the body, with sparing of the distal aspects of the forearms and legs. Lipomatous tissue is frequently found in deep locations, which can include mediastinal involvement with tracheal and/or vena cava compression. In type II, the lipomatous tissue is diffusely located over the entire body surface involving the subcutaneous fat layer and gives the patient an outward appearance of simple obesity. Despite the extensive growth of lipomatous tissue, signs or symptoms of deep, space-occupying mediastinal lesions are not found in patients with type II BSL. Our patient might be classified as type I in morphologic aspects, but there were no space-occupying lesions on image work-up.

The cause of BSL is unknown, but it has been associated with alcoholism in 60% to 90% of patients[6,7]. Less common associated findings in these patients are hyperuricemia, hyperlipidemia, diabetes melitus, hypothyroidism, liver disease, hypertension, glucose intolerance, renal tubular acidosis, macrocytic anemia and polyneuropathy[2,5,6].

Ruzicka et al[8]. reported a frequent synchronous association with malignant tumors of the upper aerodigestive tract. A thorough evaluation to rule out synchronous malignancy in patients with BSL is mandatory. Its pathogenesis is unclear, although several theories have been proposed. First, it has been related to local metabolic defects in fatty tissue, leading to a proliferation of lipomatous cells. In BSL patients, a marked increase in adipose tissue lipoprotein lipase activity and a specific defect of the adrenergic-stimulated lipolysis in lipomatous tissue have been detected[5]. Alcohol might act as a cofactor, inducing changes in the number and function of (β-oxidation, and promoting lipogenesis1. Second, adipocytes of subcutaneous tissue in the neck and interscapular area could originate from brown adipose tissue. Decreased lipolysis can be related to metabolic disorders in the mitochondria of brown adipose tissue[9]. The biochemical activity of the subcutaneous fat in BSL could be transformed in brown-like adipose tissue by metabolic triggers and drugs, such as alcohol, protease inhibitors and steroid hormones as in Cushing's syndrome like our case[9]. More recently, a genetic theory involving mitochondrial DNA mutations has been suggested which may be based on the presence of mitochondrial DNA deletions in patients with multiple lipomas[10].

There has been no case report of BSL associated with iatrogenic Cushing's syndrome. However, long-term exposure to corticosteroids leads to gross obesity along the central body axis and affects the regulation of localized fat metabolism in patients with Cushing's disease[11,12]. The mechanism by which steroids alter fat metabolism remains poorly understood. Patients with Cushing's syndrome display typically altered lipid metabolism, including levels of very low-, low-, and high-density lipoproteins11. In our case, there were no abnormalities in lipid profiles. There appear to be regional differences in the ability of cellular receptors to bind corticosteroids[11]. Some authors identified the clonal cell line under the influence of glucocorticoids help in formation and proliferation of adipocytes and reported steroid-related growth of a lipoma in the popliteal fossa and adiposis dolorosa[11-13].

Management includes alcohol abstinence, weight loss, liposuction and surgical excision of adipose tissue, which give relief to patients with functional impairment[2,7]. Limited success has been reported using a β2 agonist(salbutamol) in patients who maintain normal lipolysis and camp accumulation in the absence of adrenergic stimulation[6,7]. However, there is currently no conclusive evidence that the progression of BSL will be reversed or arrested by these measures.

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