Morphology

Benign symmetric lipomatosis (Launois–Bensaude syndrome)

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A 37-year-old woman with a personal history of appendicectomy, cholecystectomy, left oophorectomy secondary to an ovarian cyst complication, nephritic colic with repeated episodes of pyelonephritis, alcoholic hepatopathy, Raynaud's phenomenon and bilateral exophthalmos showed an increase in volume in the root of the upper limbs and in the base of the neck over a period of 4 years, painful to the touch and of a soft consistency. She presented with a pseudo-athletic appearance (Fig. 1) produced by an increase in the volume at the root of the upper limbs, upper back and the back of the neck (Fig. 2). The lesions produced a pulling sensation and were associated with paresthesia, hyperesthesia, and a moderate loss of strength in both arms. A biopsy taken from the upper third of the right arm showed a diffuse proliferation of the subcutaneous adipose tissue, which appeared normal, and extended between the collagen fibers, reaching in some cases into the most superficial zones of the reticular dermis (Fig. 3). Laboratory evaluation revealed a chronic anemia, leukopenia with moderate lymphopenia, increased erythrocyte sedimentation rate, elevation of enzymes of hepatic function, decrease in total proteins, and increase in ferritin, all in the context of hepatopathy. Antinuclear antibodies and the hormonal profile were normal. Abdominal and gynecologic echography revealed a right ovarian cyst of no clinical relevance. Cranial nuclear magnetic resonance (NMR) revealed an increase in the periorbital fat responsible for bilateral exophthalmos.

Discussion

Benign symmetric lipomatosis, or Launois–Bensaude syndrome, can be considered as a variant of the Madelung syndrome, and is characterized by symmetric and massive fatty deposits, predominantly located in the neck and shoulders, giving the patient a pseudo-athletic appearance.[1] It often affects adults between 30 and 60 years of age, and is slightly more frequent in men.[2] The marked symmetric and disproportionate accumulation of normal subcutaneous adipose tissue in the neck, shoulders, and arms, giving rise to a pseudo-athletic appearance to the upper body, contrasts with the lower half of the body which shows an asthenic appearance. The disease may be asymptomatic, but in some cases, due to the grotesque disfigurement, can cause significant esthetic or psychologic problems.[3] In general, histologic study shows normal adipose tissue, not encapsulated, involving the overlying reticular dermis.[4] The recognition of the disease during a dermatologic examination can assist in the diagnosis of associated internal pathology, mainly alcoholic hepatopathy. Benign symmetric lipomatosis is closely associated with alcoholism, detected in 60–90% of patients in several series. The hepatopathy associated with benign symmetric lipomatosis may reach up to 60%.[5]

Figure 1 Pseudo-athletic appearance produced by the increase in volume at the root of the upper limbs

It has been suggested that the Launois–Bensaude syndrome may represent a specific primary disease of the adipose tissue that is secondary to hyperplasia of the adipocytes, caused by a functional alteration of normal adipocytes.[6] The lipolytic response, reduced by intravenous norepinephrine and by fasting of patients with benign symmetric lipomatosis, may

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Figure 2 Increased volume in the upper back and the back of the neck

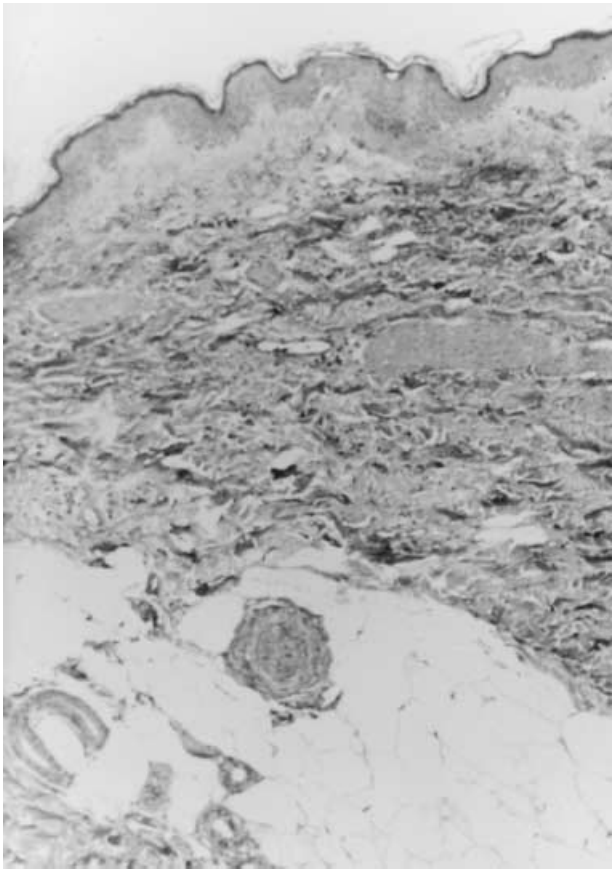

Figure 3 Diffuse proliferation of subcutaneous cellular tissue, with normal and mature cellularity, infiltrating through the collagen fibers (Hematoxiline/Eosine ×100)

indicate a "functional denervation" of certain cellular populations, and may explain the small reduction in fat cells observed in the adipose tissue after a decrease in the body weight.[2] It has been hypothesized that the lipomatous masses are formed as a result of accumulation of triglycerides in the embryonal residues of brown adipose tissue.[7]

In most patients, the process has a rapid progressive course during the initial stages and then remains constant over many years. Weight loss by dieting or the cachexia associated with neoplastic disease does not affect the extent of lipomatosis, nor does abstinence from alcohol.[5] Liposuction seems to be the best therapeutic option, although it does not prevent relapses.[8]

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