CLINICAL IMAGE

Multiple symmetric lipomatosis

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A 49-year-old man was admitted for the evaluation of bilateral gynecomastia (FIGURE 1A). He had a history of heavy alcohol consumption for 10 years. Mild gynecomastia was noted 3 years prior to admission and had markedly progressed over time. On physical examination, the patient presented with obesity, large bilateral gynecomastia, and large adipose tissue in the thorax, abdomen, and the proximal parts of the upper and lower extremities (FIGURE 1B and 1C). A breast ultrasound revealed excessively fatty breasts without any definite glandular tissue. Clinical presentation

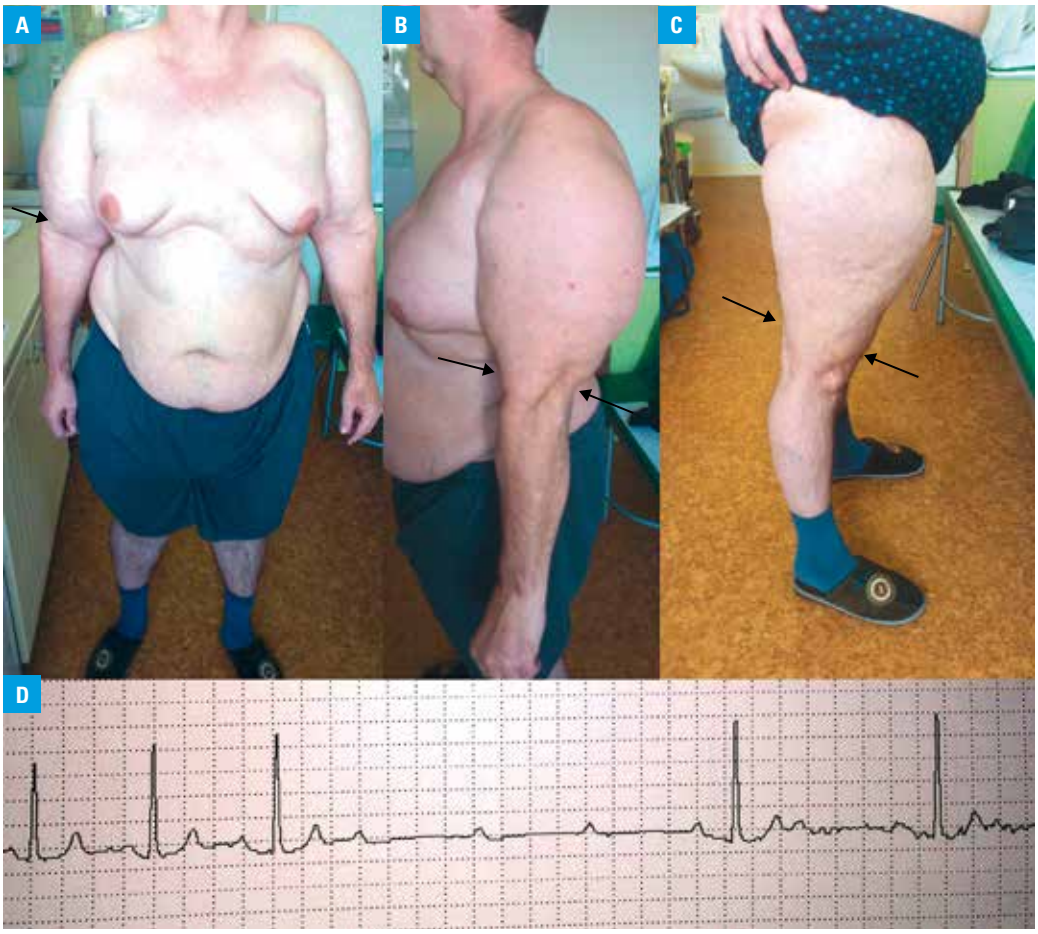

FIGURE 1 A 49-year-old man with multiple symmetric lipomatosis; **A** – bilateral gynecomastia with excessive accumulation of adipose tissue in the abdomen; **B** and **C** – excessive accumulation of adipose tissue in the proximal parts of the upper and lower extremities; **D** – advanced second-degree atrioventricular block (4:1). Arrows in panels **A**, **B**, and **C** point to the end of excessive fat on the upper and lower extremities.

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Received: April 19, 2017.
Revision accepted: May 25, 2017.
Published online: June 29, 2017.
Conflict of interests: none declared.
Pol Arch Intern Med. 2017; 127 (6): 450-451
doi:10.20452/pamw.4050

and the history of alcohol abuse were consistent with multiple symmetric lipomatosis (MSL). The diagnosis was confirmed with magnetic resonance imaging, which demonstrated an accumulation of large bilateral subcutaneous masses of nonencapsulated adipose tissue. An excision biopsy confirmed an adipose tissue mass without malignant transformation. 24-hour Holter monitoring demonstrated advanced second-degree atrioventricular block (FIGURE 1D), and the patient underwent permanent pacemaker implantation. Lipectomy of the lower abdomen with subsequent bilateral nipple-sparing mastectomy was planned, but the patient declined the surgery. He was treated conservatively with lifestyle modification and was advised to cease alcohol consumption.

MSL, also known as Madelung disease or Launois–Bensaude syndrome, is a rare adipose disorder (RAD) with an incidence of approximately 1 in 25 000. Most affected patients are men (male-to-female ratio, 15:1) with a history of chronic alcoholism.[1] Typical signs of this condition are slowly growing fatty masses around the neck, upper part of the arms, pelvis, back, and thighs. MSL type I is usually limited to the upper region of the body (head and neck), MSL type II is diffuse in nature, and MSL type III is usually limited to the thighs.[1,2] Many individuals have a combination of types II and III. The etiology of the disease remains unknown, but it seems to be associated with an alcohol-induced endocrine disorder and a mitochondrial enzyme disorder. Although lifestyle changes and bariatric surgery work effectively for the obesity component of RADs, these treatments do not routinely reduce the abnormal subcutaneous adipose tissue. Thus, people with RADs do not lose subcutaneous adipose tissue from caloric limitation and increased energy expenditure alone.[1] Alcohol withdrawal is mandatory in patients with MSL and surgical removal of lipomatous tissue is the first-line therapeutic approach to MSL.[1] Slowly progressive axonal sensory and autonomic peripheral neuropathies have been reported and impairment of autonomic function has been suggested as a cause of sudden death.[1,3] The neuropathology is a distal axonal demyelination different from that associated with alcohol intake.[4]

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