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PHOTO QUIZ

An odd looking man

T.T.M. Smit[1]*, M.J. de Jong[2], M.B. Kok[3], F. Stam[1]

[1]Department of Internal Medicine, [2]Intensive Care Unit, [3]Laboratory of Clinical Chemistry, Hematology and Immunology, Medical Center Alkmaar, Alkmaar, the Netherlands, *corresponding author: tel. + 31 (0)251-265555, e-mail: ttmsmit@hotmail.com

CASE REPORT

A 39-year-old man presented himself to the emergency department with confusion due to hepatic encephalopathy and hallucinations caused by alcoholic hepatitis. The patient had a history of alcohol abuse, which had been progressive in the last year. Laboratory blood analysis showed elevated liver enzymes and ammonia (130 µmol/l; reference value 10-45 µmol/l), renal insufficiency (eGFR 29 ml/min; reference value >60 ml/min) and elevated C-reactive protein (129 mg/l, reference value <5 mg/l). Ultrasonography of the abdomen showed an image suggestive for liver cirrhosis without ascites. The patient had a remarkable appearance with proximal adiposity (*figure 1*).

A few hours after admission the patient was transferred to the intensive care unit because of respiratory failure due to pneumonia. The patient was intubated and received inotropic medication. After extubation a symmetric paresis of both the forearms and the hand flexors became prominent.

Figure 1A. *Extreme accumulation of lipomatous tissue on the upper arms and thighs, with remarkable sparing of the shanks and forearms (figure 1B)*

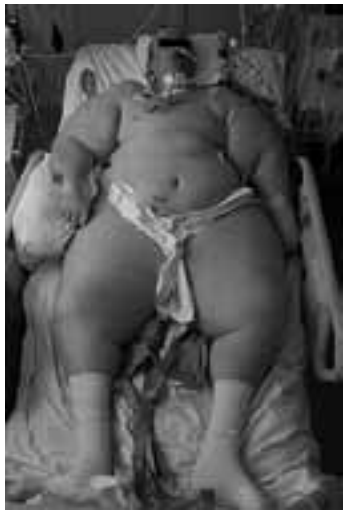
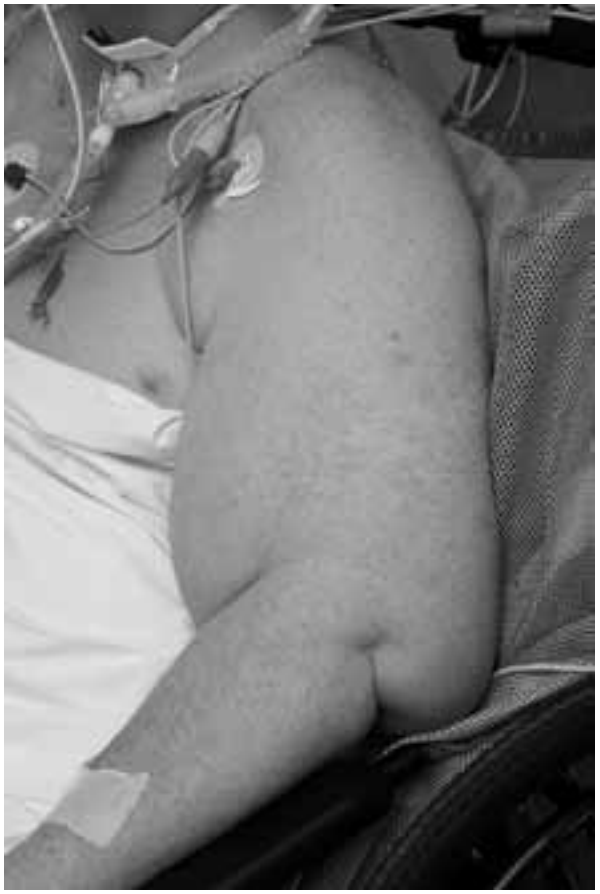


Figure 1B. *Proximal adiposity*



In addition, evident hyperesthesia of both shoulders and a paresis of the right hemidiaphragm were present. Magnetic resonance imaging of the head and neck showed no deviations. Computed tomography of the thorax and abdomen revealed diffuse atrophy of the muscles proximal in the thorax and abdomen and distal in the muscles of the legs, on the right-hand side more than the left.

WHAT IS YOUR DIAGNOSIS?

See page 328 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 324)

AN ODD LOOKING MAN

DIAGNOSIS

The remarkable appearance of our patient with proximal adiposity suggested the presence of Launois-Bensaude syndrome. Launois-Bensaude syndrome, also coined Madelung's disease or multiple symmetric lipomatosis, is a rare disease.[1]

The disease is characterised by accumulation of unencapsulated fat. In Launois-Bensaude, fat generally accumulates symmetrically around the neck, shoulders, trunk and proximal part of superior and inferior limbs. As a result, patients often have a pseudo athletic appearance. The face, forearms and shanks are usually unaffected.[1,2] The lipomatous tissue is able to infiltrate spaces between adjacent subcutaneous and muscular structures. As a result, neurological involvement is common, particularly peripheral neuropathy.[3] The neuropathy in our patient could be attributed to critical illness polyneuropathy and/or alcoholic neuropathy.

Although the aetiology of Launois-Bensaude syndrome remains unidentified, it is thought to be associated with mitochondrial respiratory chain dysfunction, which could not be demonstrated in our patient by means of an oral glucose tolerance test. Another suggested mechanism is a defect in the lipolytic pathway of the fat cell. Furthermore, most patients have or have had a history of alcohol abuse, as was the case in our patient who presented with alcohol-related hepatic encephalopathy, provoked by a community acquired pneumonia.[3,4]

Treatment is difficult and the chances of success are low. Dietetic interventions and cessation of alcohol consumption generally do not result in regression of lipomatosis. The standard treatment is surgical excision or liposuction, which has a high recurrence rate.[4]

REFERENCES

1. Verna G, Kefalas N, Boriani F, Carlucci S, Choc I, Bocchiotti MA. Launois-Bensaude Syndrome: An Unusual Localization of Obesity Disease. Obes Surg. 2008;18:1313-7.
2. Harsch IA, Bergmann T, Koebnick C, et al. Adiponectin, resistin and subclinical inflammation – the metabolic burden in Launois Bensaude syndrome, a rare form of obesity. J Physiol Pharmacol. 2007;58(Suppl 1):65-76.
3. Lee HW, Kim TH, Cho JW, Ryu BY, Kim HK, Choi CS. Multiple symmetric lipomatosis: Korean experience. Dermatol Surg. 2003;29:235-40.
4. Harsch IA, Schahin SP, Fuchs FS, et al. Insulin resistance, hyperleptinemia, and obstructive sleep apnea in Launois-Bensaude syndrome. Obes Res. 2002;10:625-32.