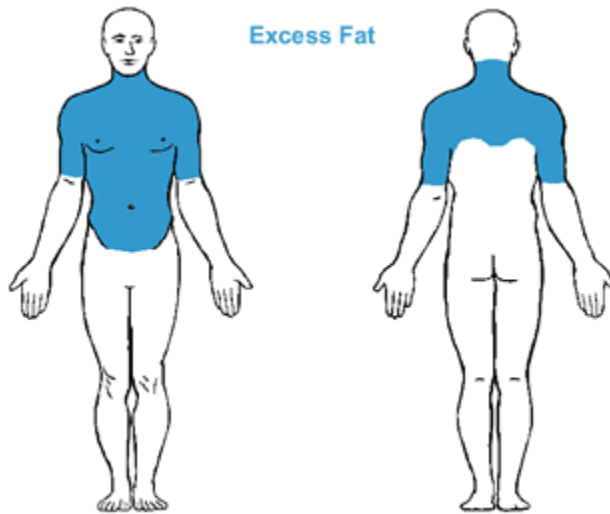


Other: Multiple Symmetric Lipomatosis (MSL, Madelung's disease)



Multiple symmetric lipomatosis (MSL, Madelung's disease, Launois-Bensaude syndrome or benign symmetric lipomatosis) is a rare disorder characterized by a diffuse, symmetrical accumulation of adipose tissue, primarily around the neck, back, shoulders and upper trunk. More than 200 cases have been reported so far. It usually affects adult males, mostly in the Mediterranean region and has a strong association with heavy alcohol intake, but a similar disease is also noted in non-alcoholics in association with mitochondrial DNA mutations.

The physical characteristics of a patient with typical Alcohol-induced MSL are the presence of diffuse lipomatous masses deposited symmetrically around the neck, back, shoulders and upper trunk. Suprascapular and supraclavicular involvement is common and occasionally it may involve the parotid and post auricular region. The face and distal extremities are usually spared, and some patients have been described to have loss of adipose tissue from the thighs and other uninvolved areas (type 1 MSL), while in others the fat accumulation is superimposed on a generalized obesity phenotype (type 2 MSL).

Female patients predominantly display the type 2 obesity phenotype of MSL, and commonly have involvement of the proximal arms with sparing of the neck and submental region. The occurrence of deep space occupying lesions leading to tracheal or esophageal compression and the superior venacaval syndrome has been reported in 15 to 20% of the patients, and accounts for a significant amount of morbidity associated with this disorder. Involvement of unusual sites such as cheeks, tongue, hands, thighs and scrotum have also been reported. Patients are noted to have peripheral neuropathy and hyperuricemia. Usually they have normal glucose tolerance, normal serum triglycerides and elevated levels of HDL – cholesterol. The molecular mechanisms by which alcohol causes lipomatous deposits in MSL is poorly understood.

Symmetrical lipomatosis has also been noted in patients with systemic mitochondrial diseases such as MERRF (myoclonic epilepsy with ragged red fiber myopathy). This is caused by point mutations in the mitochondrial DNA.

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