Acquired Lipodystrophies

Acquired: Generalised Lipodystrophy (AGL; Lawrence Syndrome)

This is a rare disease characterized by generalised disappearance of fat occurring during childhood and adolescence. A total of about 80 cases have been reported. Patients have normal body fat at birth thus distinguishing them from patients with congenital generalized lipodystrophy. Females are affected approximately 3 times more often than males. AGL could be due to panniculitis (inflammation in the fat) or autoimmune diseases, however, in several cases no causative factor could be identified. The onset of AGL may also occur following infections such as varicella, measles, pertussis, diphtheria, pneumonia, osteomyelitis, parotitis, infectious mononucleosis, and hepatitis. Usually, the onset of lipodystrophy is gradual over months to years. Eventually generalized and near complete loss of fat may occur in some patients resulting in muscular appearance and prominent superficial veins (veins under the skin). There is loss of fat from the palms and soles also. Dark velvety pigmentation (acanthosis nigricans) may occur in the axilla and neck, umbilicus, nipples and occasionally hands and feet. Increased linear growth may be seen in the children. Excess body hair, enlargement of genitalia (clitoromegaly) and occasional ovarian cysts (enlarged ovaries) may be seen in the females. Enlargement of liver and spleen is frequently seen. The patients commonly have moderately severe elevation of blood lipids. Diabetes occurs usually after the onset of lipodystrophy. Diabetes is often difficult to control despite requiring large doses of injectable insulin. Patients with AGL may also develop other autoimmune disorders like vitiligo (light-colored spots on skin), sicca syndrome, rheumatoid arthritis, dermatomyositis, thyroiditis and chronic active hepatitis. The precise mechanism involved in loss of body fat in this disorder however remains unknown but it is likely that autoimmune mechanisms (body defense mechanisms acting against body fat) are involved. This condition can sometimes be referred to as 'Total body lipodystrophy'.

There are 3 types of AGL:

- Panniculitis variety (type 1): The patient presents with painful and inflamed subcutaneous nodules or maculopapular lesions. Upon healing, depressed scars remain but the overlying skin is normal. New nodules appear and there is progression of subcutaneous fat loss.

- Autoimmune disease variety (type 2): In this variety, the patients have past or present evidence of autoimmune diseases.

- Idiopathic variety (type 3): This is the most common variety. The etiological factor of this variety is not known.

Acquired: Partial Lipodystrophy (Barraquer-Simons Syndrome)

The onset of acquired partial lipodystrophy usually occurs around 8-10 years of age and is usually preceded by an episode of acute viral infection. It is characterized by the loss of fat from the face, extending to involve the neck, shoulders, arms, forearms, thoracic region and upper abdomen occasionally extending to the groin or thighs. Usually legs and hips are spared. After puberty, Women may accumulate disproportionately large amount of fat in the hips and legs. Fat loss usually occurs over 18 months but can occur periodically during several years. Patients with acquired partial lipodystrophy do not have metabolic abnormalities associated with insulin resistance such as elevated lipid levels, acanthosis nigricans (dark velvety pigmentation of the skin), hirsutism (increased body hair) or menstrual abnormalities. Females are affected three times more often than males. Approximately one-third of these patients develop a kidney problem called Membranoproliferative Glomerulonephritis. It usually occurs more than 10 years after the onset of lipodystrophy. Patients have low levels of complement C3 (a factor that plays a role in immune response) in their blood (hypocomplementemia). They also have an antibody in their blood called the C3 nephritic factor. Acquired partial lipodystrophy is also associated with autoimmune disorders like systemic lupus erythematosus (SLE), dermatomyositis, hypothyroidism, pernicious anemia, celiac disease, dermatitis herpetiformis, rheumatoid arthritis, temporal arteritis and leukocytoclastic vasculitis.

Acquired: Localized

Localized Lipodystrophies are defined as a localized loss of subcutaneous fat from small areas or from parts of a limb. There may be single or multiple lesions. It is characterized by depressed areas corresponding to the loss of subcutaneous fat. In some cases, it may be associated with tender, painful nodules in the skin. Usually, it occurs in diabetic patients at the site of insulin injections. In some patients, at loss occurs from areas where pressure is applied frequently. For example, pressing thigh against a make-up table. Many others develop these due to as yet unknown mechanisms.

Acquired: Lipodystrophy in HIV-infected Patients (LD-HIV)

This type of lipodystrophy seen in Human Immunodeficency Virus (HIV)-infected patients has been associated with long duration of protease inhibitor drug therapy. Although protease inhibitors appear to be a strongest link to HIV lipodystrophy interactions among protease inhibitors and other antiretroviral drugs, and the HIV virus may also contribute to HIV lipodystrophy. The precise mechanism by which lipodystrophy occurs in HIV infected is not known. Patients lose subcutaneous fat from the face and both arms and legs including the buttocks. These subjects develop increased muscularity in the arms and legs with prominent superficial veins. At the same time excess fat may accumulate in the neck and truncal region causing "double chin","buffalo or camel hump" and increased abdominal girth commonly known as "crixivan belly". Both men and women complain of growth in breast size. The patients may also develop glucose intolerance but diabetes mellitus develops rarely. Many patients have high levels of serum cholesterol and triglycerides.

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