

P94

Acquired partial lipodystrophy (Barraquer-Simons Syndrome) associated with chronic discoid lupus erythematosus

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We report the case of a 44-year-old woman suffering from progressive loss of subcutaneous fat tissue. The symptoms started at age of 24 in the face spreading in the next years symmetrically to thorax, mammae, abdomen and the inguinal region. Seven years after onset of the disease the patient developed erythematous hyperkeratotic plaques with atrophy and scarring on the capillitium and shoulder area. Laboratory tests were negative for autoantibodies, HIV, hepatitis, borreliosis and lues. Complement status showed no abnormalities. Urine sediment remained without pathological findings. The clinical presentation together with the laboratory findings and the results of the histological analysis lead to the diagnosis of an acquired partial lipodystrophy (APL) associated with chronic discoid lupus erythematosus (CDLE). APL also known as Barraquer-Simons syndrome is a rare condition with progressive loss of subcutaneous fatty tissue with onset in childhood or early adolescence. It is characterized by progressive and symmetrical loss of subcutaneous fat beginning in the face and further spreading in a craniocaudal pattern involving trunk and upper extremities, not affecting the lower extremities. So far 220 cases of APL have been described in the literature. Most of the patients were female. Several autoimmune diseases, in particular systemic lupus erythematosus and dermatomyositis, are associated with APL. The etiology of APL remains unknown, but activation of an alternate complement pathway has been discussed. In most APL patients low levels of complement C3 and the presence of polyclonal immunoglobulin C3 nephritic factor (C3NeF) could be detected. Recently, the disease has been linked to mutations in the gene encoding the nuclear lamina protein (lamin B2- LMNB2). Approximately twenty percent of the patients develop membrano-proliferative glomerulonephritis (MPGN), which is also the crucial factor for the prognosis of the disease. Treatment options are mostly limited to cosmetic reconstruction of facial lipodystrophy. The association of CDLE with APL as found in our case further supports the notion of an autoimmune pathogenesis of APL.

P95

Double island flap for reconstruction of excision defects larger than 5 cm²

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An excision larger than 5 cm² on the extremities is often challenging to close for the dermatologist. If the skin tension is too high a closure with local

flaps, full thickness skin grafts or Thiersch graft has to be considered. Skin grafts have the disadvantage to show an important texture and colour mismatch with – in deep excisional defects - a visible depression on the former defect. Therefore for reconstruction on the body and extremities, this option is only a second choice. We present an easy approach of covering extended defects with a double island pedicle flap.

Technique: The double island flap is a good solution for defects which cannot be closed side to side. On each opposite sides of the round wound defect two triangles are marked similar to the wedge excision or lancet form excision with an angle of not more than 60°. The lancet form excision should be placed according to common rules in the sense of the hair growth as published in Sau rat et al. The island pedicle flap is prepared according to our former publication in Dermatologic Surgery. The skin is incised until the subcutaneous fat tissue. The subcutaneous fat tissue is bluntly separated so that deep vessels can be defined and coagulated thoroughly. To preserve a sufficient blood supply, the last third of the subcutaneous tissue should not be severed. In each triangle the skin between dermis and hypodermis is undermined in the first third adjacent to the defect. On the lateral angle of the triangle the first third of the pedicle in the sub cutis should be undermined bluntly to allow optimal advancement of the island flap. The operator should try to advance both triangled pedicle flaps one to each other as far as they can be joined without tension. If the flap is too much under tension the operator should try again to undermine thoroughly by tearing the flap to the defect. When each flap can be positioned to one each other without much tension, each triangles are sutured together.

This procedure is easy to perform and provides better aesthetic and functional results than a skin graft, especially in deep excision defects. Disadvantages of the double island flap is the unnatural geometric shape of the scar and a certain fragility if the flap was traumatized during the operation. Furthermore, the pedicle is relatively small, which can lead to postoperative edema of the flap (trapdoor deformity, pincushening).

P96

Screening of different stages of cutaneous squamous cell carcinoma for changes in miRNA expression

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Development of cutaneous squamous cell carcinoma (cSCC) is promoted by an accumulation of UV-light exposure through lifetime, but can also be part of inherited genetic skin diseases such as epidermodysplasia verruciformis (EV). The expression pattern of mRNA alters during the formation of cSCC. MicroRNAs (miRNA) are known to regulate the expression of mRNA, but data addressing the miRNA expression pattern in cSCC in the general popula-