Dermal Aplasia and Sclerocornea) syndrome. MLS/MIDAS syndrome is a rare X-linked dominant neurocutaneous disease with in utero male lethality. Besides cutaneous and ocular abnormalities, additional manifestations include developmental delay, short stature, heart, central nervous system and genitourinary tract abnormalities. The candidate gene, HCCS, encoding the mitochondrial holocytochrome c-type synthase, is involved in the mitochondrial respiratory chain and in apoptosis pathways. The mother carrying the same genetic abnormality may be completely asymptomatic (random X inactivation).

Katariina Hannula-Jouppi, Helsinki, Finland: Acrodermatitis Enteropathica. Acrodermatitis enteropathica (AE) is a rare autosomal recessive form of zinc deficiency characterized by periorificial and acral dermatitis, alopecia, and diarrhoea. Symptoms begin soon after birth in bottle fed infants and after weaning in breastfed infants. Mutations in SLC39A4 lead to deficient zinc/iron transfer by hZIP4, leading to inadequate zinc absorption from the intestine and low plasma zinc levels. Treatment of AE requires lifelong daily oral zinc supplementation 3 mg/kg/day (1–5 mg/kg). Clinical response is observed within days to a few weeks.

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Lupus

Lupus erythematosus (LE) is a complex, multifactorial autoimmune disease. Nailfold videocapillaroscopy (NVC) is a fundamental imaging technique used in the clinical examination of patients with different autoimmune diseases. It has evident diagnostic and prognostic power especially in systemic sclerosis but also in other systemic connective tissue diseases. NVC is based on light microscopy, usually using 200 × magnitude lens. It offers a good tool to distinguish between primary and secondary Raynaud's phenomenon (RP). Primary RP is quite common, occurring in 15–20% of young females. The presence of giant capillaries and microhaemorrhages are typical for early pattern changes of a systemic collagenosis like systemic scleroderma, systemic LE (SLE) or even dermatomyositis. An increase in these features and loss of capillaries (active pattern) is followed by neo-angiogenesis and fibrosis. The proceeding findings in videocapillaroscopy correlate usually with the activity of the systemic collagenosis.

Diagnosis and classification of cutaneous LE(CLE) is based on clinical features, positive serology/autoantibodies, abnormalities

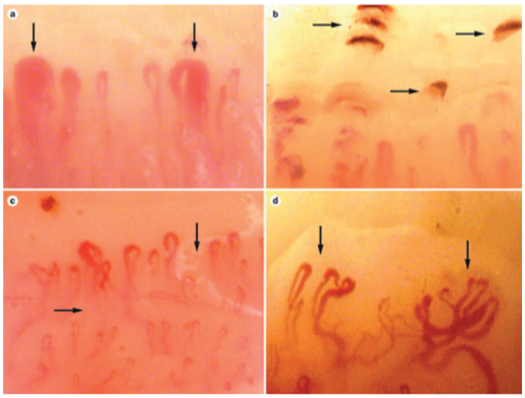

Fig. 1. Nailfold videocapillaroscopy changes: a) giant capillaries, b) microhemorrhages, c) loss of capillaries and d) neoangiogenesis.

in blood count and complements and on immune histology. ANA antibodies are positive in 5–10% of discoid LE (DLE) patients, in 60–80% of subacute CLE (SCLE) and in over 90% of SLE patients. Ro/SSA- and La/SSB antibodies are characteristic of SCLE (positivity in 70–90% and 30–50%, respectively) but they are positive also in Sjögren syndrome. Sm-antibodies are positive in 10–30% of SLE patients. DNA-antibodies refer to SLE and are seen in 40–90% of this patient group. Histone- antibodies are seen in drug-induced lupus (DILE): in up to 95% of classic DILE with systemic symptoms of lupus, only in up to 33% of drug-induced SCLE, in up to 57% of anti-TNFα-induced DILE and in up to 50% of idiopathic SLE. In the pathogenesis of lupus, the environmental triggers (hormones, viruses, UV light, drugs) and genetic factors together with either increased production and/or reduced clearance of apoptotic blebs lead to initiation of autoimmunity. Complement deficiency is also related to SLE. A number of CLE-associated risk genes have been shown like *IRF5*, *TYK2*, *ITGAM*, *CTLA4* and *STAT4*.

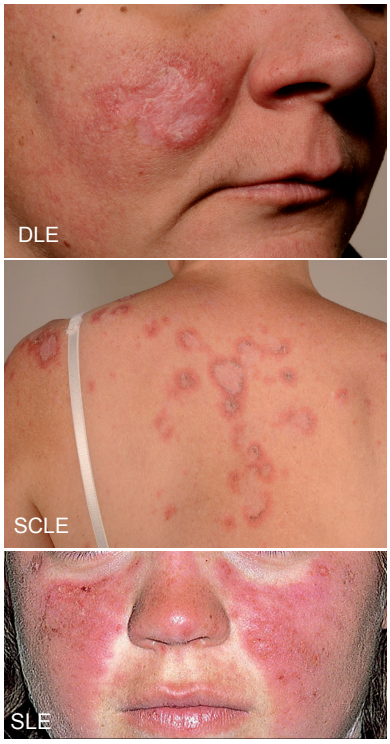

Fig. 2. Clinical appearances of discoid lupus erythematosis (DLE), subacute cutaneous LE (SCLE) and systemic LE (SLE).

Population-based epidemiological data on CLE are now reported, both from Sweden and USA. The incidence of Ro/SSA-positive SCLE in

Stockholm County Council (2 million inhabitants) has been estimated to be 0.7/100,000 persons/year and for all SCLE 1.0/100,000 with an estimated prevalence of 8.9–20/100,000 persons. The incidence of SLE in Sweden is 4.8/100,000 persons/year. In a recent study, CLE incidence in Sweden was shown to be 4.0/100,000. More than 10% of the whole group of CLE, and >20% of SCLE patients, progressed to SLE within 1 year.

CLE can be treated with local treatments by using steroid or tacrolimus creams. Often oral treatment is needed, hydroxychloroquin in the first line but quinacrine can be added if available. Prednisolone can be combined especially in the beginning to get a more rapid response. Other possibilities are immune suppressive metotrexate, atsathioprine and mycophenolate mofetil or acitretin/isotretinoin and even thalidomide. In the most complicated cases of SLE can rituximab or belimumab be used both of which have also effect on the mucocutaneous symptoms.

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Pigmented Lesions and Melanoma

Olle Larkö: New Imaging Techniques in the Diagnosis of Melanoma. The incidence of malignant melanoma is increasing rapidly in the Nordic countries. Also, mortality is high. A dermatologist is better at diagnosing melanoma than doctors from other disciplines. However, even dermatologists miss a substantial fraction of possible melanomas. Hence, new imaging devices must be developed to facilitate diagnosis. So far we have relied on dermatoscopy, but more innovative devices are just around the corner. It includes SIASCOPY where monochromatic light is reflected from the tumour surface is reflected and image analysis carried out with a special algoritm. Also confocal microscopy adds value to the diagnostic process. Several apps for smartphones have been developed but their value and safety has yet to be established.

Carin Sandberg: Mobile Teledermoscopy for Fast Track Management of Skin Cancer. The skin cancer incidence is rising at an alarming rate. For malignant melanomas the only really effective treatment is early detection and excision. This means that the referral process is critical. New technologies have emerged with smartphones, etc. In a large project it has clearly been demonstrated that the use of modern IT technology speeds up the referral process. In this study, images of suspected tumours were photographed with an iPhone and sent by e-mail from general practitioners to dermatologic clinics. Compared to the ordinary process malignant melanomas at excision was 1 mm thinner. This means a major improvement in prognosis for the patient.

Olli Saksela: Risk of New Melanomas in Multinaevus Patients. Patients with many naevi run a greater risk for developing malignant melanoma. In the current lecture, handling of this dilemma was discussed. It is important that dermatologists see patients with suspected malignant melanomas due to the better sensitivity and specificity.

Olga Tatti: MT3-MMP Controls a Proteolytic Switch Between Blood Vascular and Lymphatic Invasion of Melanoma Cells. In the process of metastasis, tumour cells invade lymphatic vessels. It clearly correlates with poor prognosis. The author found that MT3-MMP (membrane-type-3 matrix metalloproteinase) was over-expressed in nodular malignant melanomas and lymph node metastases. It seems that MT3-MMP acts as a molecular switch of vascular invasion. MT3-MMP may impair blood vascular invasion.

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Non-melanoma Skin Cancer & Actinic Keratosis

Non-melanoma skin cancer (NMSC) includes all primary skin cancers except melanoma. The most frequent are basal cell (BCC) and squamous cell carcinomas (SCC) (Fig. 1), but actinic/solar keratosis (AK) is today considered to be an initial SCC; another term is keratinocyte intraepithelial neoplasia (KIN). About 40% of metastatic SCCs start as AK. The aetiology of NMSC is UV radiation, and the incidence is increasing.

Sari Koskenmies: Aetiology and Treatment Options for Non-Melanoma Skin Cancer & Actinic Keratosis. Aetiology and treatment options for non-melanoma skin cancer (NMSC) presented. In USA in 2006 an incidence of >2 million patients with over 3 million NMSC was estimated. 70–80% of all NMSC are BCCs and 20% SCC.The risk of developing NMSC is mainly related to UV irradiation exposure, but genetics and muta-

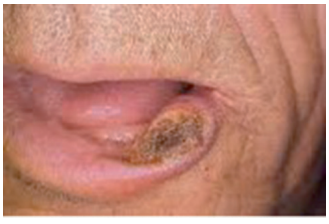

Fig. 1. Squamous cell carcinoma of the lip.