STD & Genital Dermatology

Anne Olaug Olsen: Anal dysplasia and cancer. Anal dysplasia (anal intraepithelial neoplasia) is a precancerous condition induced by human papillomavirus (HPV), which may progress to invasive cancer. The worldwide incidence of anal cancer in the general population although low, has increased over the past three decades. The increase has been alarmingly high among HIV positive men who have sex with men. Immunocompromised individuals and those previously treated for HPV-related premalignant disease also represent high-risk groups for the development of anal dysplasia and cancer. It is however noteworthy that the majority of cases of anal and perianal cancer continuously are diagnosed in heterosexual and otherwise healthy men and women. There are numbers of challenges to be addressed as long as there is no consensus about the optimal management of HPV-induced anal intraepithelial neoplasia. Screening algorithms and follow-up routines are in demand. A wide range of treatment modalities, including topical and ablative therapy, is available. Optimal regimes for screening, intervention and follow-up of anal dysplasia in high-risk groups, is a priority.

Eija Hiltunen Back: Ulcus Vulvae Acutum Lipschütz. Lipschütz ulcer (acute genital ulcer, AGU) is an underdiagnosed disorder that presents as an acute painful necrotic vulvar ulcers in prepubertal or pubertal girls without any history of sexual contact. The onset is preceded by an acute systemic illness. Primary Ebstein-Barr virus infection is the most frequently reported aetiology. The diagnosis is established clinically after ruling out STIs, trauma, autoimmune causes, drug reactions and local manifestations of systemic illness. The histological findings are nonspecific. The management consists of symptomatic treatment like oral and topical antibiotic and corticosteroid therapy. Lesions heal spontaneously in a few weeks with no sequelae. It is important to keep AGU in mind as a differential diagnosis of vulval ulceration to avoid misdiagnoses and unnecessary invasive investigations.

Harald Moi: IUSTI Guidelines on Gonorrhoea Treatment. Neisseria gonorrhoeae has shown a remarkable capacity to develop resistance to multiple classes of antibiotics. After a steady rise in minimum inhibitory concentrations in recent years, resistance and even clinical failures to extended-spectrum cephalosporins (ceftriaxone and cefixime) have been confirmed. As a consequence, combination antimicrobial therapy is recommended. The first line treatment according to recent European IUSTI guidelines is 500 mg ceftriaxone i.m. and 2 g azithromycin p.o. as direct observed therapy. If treatment after susceptibility testing, antibiotics are given according to the test results.

Erika Wikström: Overtime Chlamydia Trachomatis Serotype Distributions in Fertile-aged Finnish females. While the occurrence of *Chlamydia trachomatis* has been high in the affluent countries for several decades little is known about the ecology of *C. trachomatis* serotypes. We studied the distribution of *C. trachomatis* serotypes in Finnish women from the 1980s to the 2000s. 1,169 healthy subjects testing positive for *C. trachomatis*-specific IgG antibodies were available from a large subcohort of 11,067 15–29-years old women belonging to the Finnish Maternity Cohort of the National Institute for Health & Welfare. The temporary *C. trachomatis* serotype replacement among females parallels changes in the sexually active population in the 1990s in Finland.

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Atopic Dermatitis

See separate Meeting Report from Louise Lönndahl on page 139–140.

Paediatric Dermatology

Katariina Hannula-Jouppi, Helsinki, Finland: Netherton Syndrome. Netherton syndrome (NS) is a rare autosomal recessive genodermatosis characterized by severe skin inflammation, erythema and scaling, multiple atopic manifestations and trichorrhexis invaginata, a hair shaft defect causing brittle "bamboo" hair. Mutations in the *SPINK5* gene cause NS by loss of LEKTI, a serine protease inhibitor in the epidermis. Loss of LEKTI disrupts normal skin homeostasis and leads to unopposed KLK activity and ELA2 activation, which initiate proinflammatory and proallergic cascades. We have studied 10 Finnish NS patients and identified a novel Finnish founder mutation in exon 8 of *SPINK5*, in 7 NS patients originating in the Ostrobothnia region. All NS patients had typical NS features and we saw a rapid increase in IgE sensitization to multiple during the first years.

Nicolas Kluger, Helsinki, Finland: Congenital Linear Streaks of the Face and Neck and Microphthalmia in an Infant Girl. A newborn girl presented with atrophic unilateral facial lesions following Blaschko's lines, aplastic nails, ipsilateral microphthalmia, aniridia and sclerocornea was diagnosed Microphthalmia with Linear Skin defects (MLS) syndrome/MIDAS (MIcrophtalmia,

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

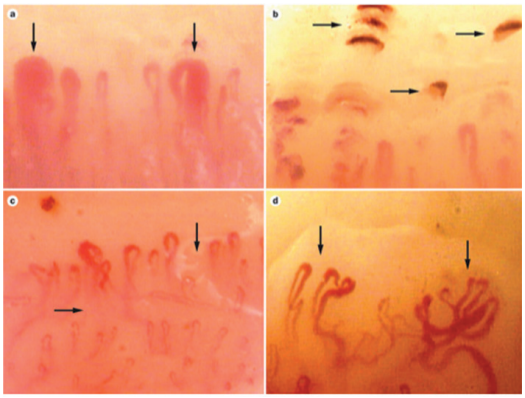

Fig. 1. Nailfold videocapillaroscopy changes: a) giant capillaries, b) microhemorrhages, c) loss of capillaries and d) neoangiogenesis.

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*.

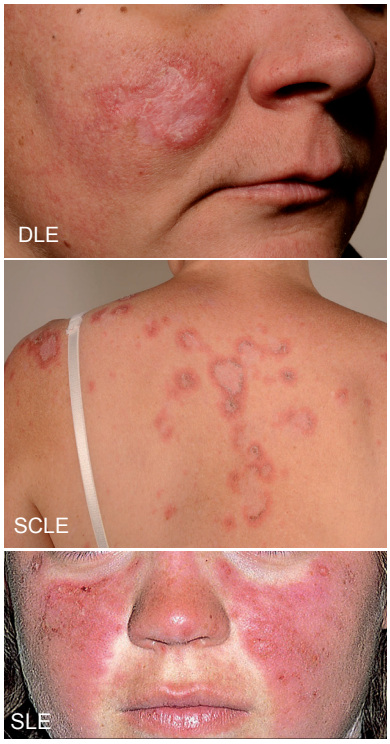

Fig. 2. Clinical appearances of discoid lupus erythematosis (DLE), subacute cutaneous LE (SCLE) and systemic LE (SLE).

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