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Vulval skin condition

We were interested to see in the 4 July 2008 issue of the *Journal* an illustrated case reported to be genital lupus erythematosus in association with SLE and CREST syndrome (Darwish T. *Medical image. Genital lupus*. http://www.nzma.org.nz/journal/121-1277/3136). Cutaneous lupus affecting the vulva is rare. Previous case reports have described mucosal erosions,[1,2] and irregular plaques of discoid lupus on the labia majora.[3]

Did Dr Darwish consider a diagnosis of lichen sclerosus?

Vulval lichen sclerosus is a very common skin complaint that often starts around the age of 50 years. Unlike cutaneous lupus, which tends not to be itchy, lichen sclerosus often causes intense pruritus. Clinical features include often-symmetrical white, and sometimes erythematous, atrophic, sclerotic and/or hyperkeratotic plaques affecting clitoral hood, labia minora, perineum, and/or perianal skin; associated with erosions and ulceration, haemorrhages, blisters, labial resorption, and adhesions.

Histology is often reported to show prominent hyalinisation of the upper dermis in association with interface dermatitis; it may be similar to scleroderma. The pathological description of 'dystrophy' is outdated. Direct immunofluorescence is negative, unlike cutaneous lupus in which there is deposition of IgG, IgM, and C3 along the basement membrane.

Lichen sclerosus is a chronic autoimmune sclerosing skin disease. It may coexist with localised scleroderma (morphoea),[4] systemic sclerosis, and CRST.[5] These conditions all involve the extracellular matrix; an autoantibody to extracellular matrix protein I is present in up to 80% of patients with lichen sclerosus.[6]

A study of 202 New Zealand women with vulval lichen sclerosus revealed other autoimmune disorders commonly coexisted, particularly thyroid disease (19%) and psoriasis (17%). Antinuclear factor titre was greater than 1:160 in 18% of 142 tested, but none had antibodies to double stranded DNA or other evidence of lupus erythematosus.[7]

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Response from Dr Darwish

We appreciate Dr Oakley and Dr Rowan comments. We agree that these ivory-white lesions mimic lichen sclerosis (LS). However, the acuity of symptoms in our patient, in addition to the presence of systemic findings of SLE and CREST syndrome, favoured genital lupus over LS.

Although the deposition of IgG, IgM, and C3 along the basement membrane is diagnostic for cutaneous lupus, there are few reports documenting the morphological features of genital lupus.

Of note, the diagnosis can be established based on extragenital lesions and/or histopathological findings.[1]

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