Kikuchi's disease in association with DLE

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Sri Lanka Journal of Dermatology, 2007, **11**, 28-29

Introduction

Kikuchi's disease (KD), also called histiocytic necrotizing lymphadenitis or Kikuchi-Fujimoto disease is an uncommon, idiopathic, generally self-limited cause of lymphadenitis, which affects mainly young women. The most common clinical manifestation is cervical lymphadenopathy, with or without systemic symptoms and signs[1,2].

Several associated conditions were reported with KD. In only rare instances has its associations with cutaneous lupus erythematosus without systemic involvement been reported[2,3]. We report a case of KD associated with discoid lupus erythematosus (DLE).

Case report

A 28-year old female presented to the Dermatology Clinic, Teaching Hospital, Kandy, with a history of patches of hair loss over the scalp for 1 year and she had mild itching and pain over the lesions. She denied any associated fever, night sweats, weight loss or photosensitivity. She also had painful swellings over the lateral aspects of the neck for 2 months.

Her past history was uneventful except laparotomy in 1992 for dysgerminoma.

Examination revealed multiple patches of scarring alopecia with scaling over the scalp suggestive of DLE. There were no other skin lesions suggestive of DLE or KD. No oral ulcers were found. She had bilateral tender firm cervical lymphadenopathy. Other groups of lymphnodes were not enlarged. There was no hepatosplenomegaly. Systemic examination was normal.

A diagnostic lymph node biopsy was performed when her lymphadenopathy failed to improve with antibiotic treatment.

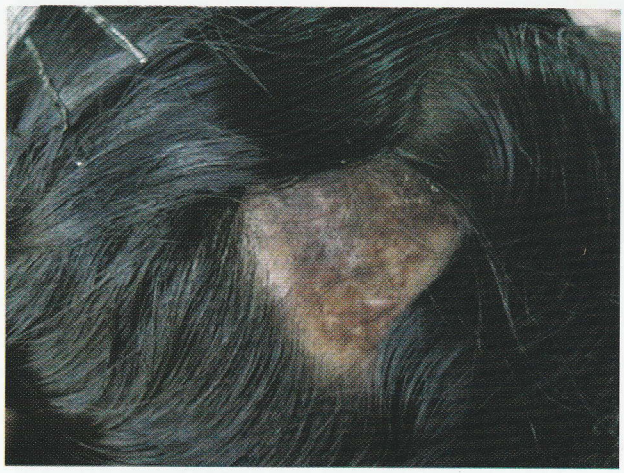

Figure 1. *Patches of scarring alopecia.*

On investigation, ESR 10mm/1st hr, WBC 5.2x109/l, N-36, L-64, Hb% 11.7g/dl, platelet count 301x109/l, blood picture - rouleaux formation with neutropenia, ANF- negative, chest x-ray - NAD, Mantoux test - negative.

Biopsy from scalp showed epidermal atrophy, basal cell degeneration, marked follicular plugging and dermal perivascular and periadnexal lymphocytic infiltrate consistent with DLE.

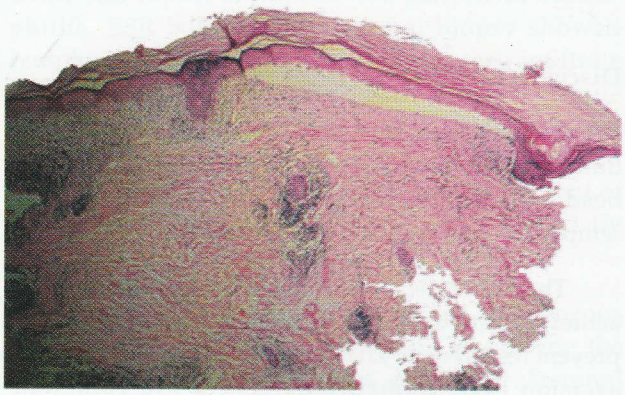

Figure 2. *Histopathology of scarring alopecia.*

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Cervical lymph node biopsy showed preserved nodal architecture with focal, well-circumscribed paracortical necrotizing lesions. Focus of coagulative necrosis surrounded by histiocyte proliferation. There are abundant karyorrhectic debris, scattered fibrin deposits and collection of large mononuclear cells.

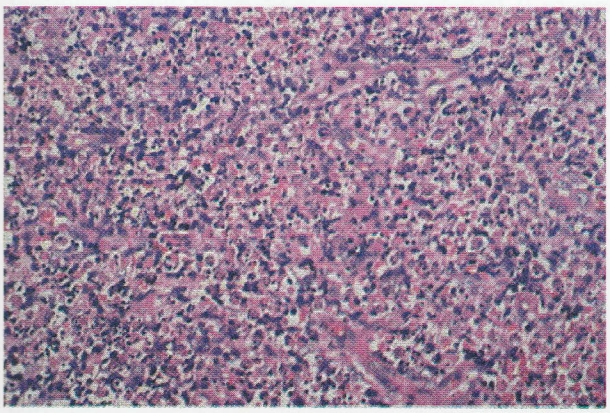

Figure 3. *Histiocytic necrotizing lymphadenitis lymph node.*

The diagnosis of KD was made from the histological findings of the lymph node. She responded to a course of prednisolone over 6-8 weeks (cervical lymphadenopathy and pain disappeared).

Discussion

Kikuchi's disease is a histiocytic necrotizing lymphadenitis that is prevalent in Asia and is being increasingly recognized in other areas of the world. In 1972 both Kikuchi and Fujimoto et al described this disease in the Japanese literature[1,4,5,6].

It presents with localized lymphadenopathy, predominantly in the cervical region, accompanied by fever and leucopenia in upto 50% of the cases[1]. Skin changes occur in about one third of patients (16-40%) and are often of a non specific nature[1-6]. Most cutaneous lesions present as morbilliform rash, papules, indurated plaques, nodules or urticaria in the upper part of body[1,3,4,6]. Our patient had posterior cervical lymphadenopathy and leucopenia but she didn't have fever and skin lesions suggestive of KD.

The aetiology of KD is not known, but various triggers have been incriminated, including the human herpesvirus 6, parvovirus B19, Epstein-Barr virus, dengue virus, cytomegalovirus, human immunodeficiency virus and toxoplasma. Other associated conditions include Still's disease, Sweet's syndrome and leucocytoclastic vasculitis[1,3]. In only rare instances has its associations with cutaneous lupus erythematosus without systemic involvement been reported[2,3].

The diagnosis is primarily made by tissue biopsy[1,6]. KD almost always runs a benign course and resolves in several weeks to months[6]. Disease recurrence is unusual and fatalities are rare[1].

Clinically and histologically, the disease can be mistaken for lymphoma or SLE[1]. Although the presence of lymphadenopathy is not uncommon in SLE patients, particularly in the phases of disease activity, the concomitance with KD has been reported in the literature[1,3]. Its recognition is necessary because one can avoid laborious investigations for infectious and lymphoproliferative diseases. KD has been reported to precede, coexist or follow the diagnosis of systemic lupus erythematosus (SLE)[1,3]. In our patient no manifestations of systemic lupus erythematosus occurred after 2 years of follow up.

Once the diagnosis is established, no specific treatment is indicated because this is a self-limited condition, but a course of prednisolone may speed resolution[3].

References

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