

Case 2: A Young Lady with Kikuchi's Disease and Lupus Erythematosus

Dr. N. M. Luk

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Investigations

Blood tests revealed a mild degree of microcytic anaemia with a low serum iron level of $3.9\mu\text{mol/l}$ (8.8 - 27) and a normal total iron binding capacity level (TIBC) of $63.6\mu\text{mol/l}$ (45 - 73). The anti-nuclear factor titre was raised at 1:120 and the anti-double strand DNA titre was 11 IU/ml (0 - 35). The white cell and platelet count, erythrocyte sedimentation rate, the liver and renal

CASE SUMMARY

History

A 35-year old chinese woman had been suffering from Kikuchi's lymphadenitis since 1995 and was followed up by the ENT surgeon. She noticed skin rash over the right side of her nose two months prior to the present consultation. The lesion was asymptomatic but was getting bigger in size. Otherwise, she had no history of photosensitivity, joint pain, Raynaud's phenomenon, fever or any constitutional upset. On further inquiry, she remembered a scaly lesion on her left upper arm that had been present for two years. Her past health was otherwise unremarkable. There was no family history of similar skin problem.

Physical examination

Physical examination revealed multiple erythematous papulonodules over the right side of her nose (Figure 1). There was also an atrophic plaque over her left upper arm with underlying induration and surface scaling (Figure 2). No lymph node was palpable. The liver and the spleen were not enlarged.

Differential diagnoses

The possible diagnoses of the lesions include discoid lupus erythematosus, Jessner's lymphocytic infiltration and morphea.



Figure 1: Erythematous papulonodules on the right side of the nose.



Figure 2: An atrophic plaque with surface scaling and underlying induration on the left upper arm.

function tests were all normal. Anti-ENA, rheumatoid factor, C3 and C4 levels were normal. No significant radiological finding was detected on chest radiography. Urinary microscopy showed the presence of one + white cell, otherwise there was no other abnormality seen.

Skin biopsy of the left upper arm lesion showed marked interface dermatitis. There was dense perivascular inflammatory infiltrates including plasma cells throughout the superficial and deep dermis. Interstitial mucin accumulation was noted. Area of hyaline necrosis was present in the deep dermis and dermo-subcutis, which was surrounded by lymphohistiocytic infiltrate with giant cells. A lymphoid follicle was seen. There was also lymphocytic vasculitis. No organism was demonstrated by special stain for fungus and acid fast bacilli. Immunofluorescence study was negative except for non-specific weak C3 staining at the dermo-epidermal junction. The diagnosis was consistent with lupus profundus.

Diagnosis

The diagnosis of this patient was lupus profundus preceded by Kikuchi's disease and iron deficiency anaemia.

Management

The patient was advised on sun protection and given clobetasol propionate (dermovate) cream for her skin lesion. She had been followed up regularly to watch out for any systemic involvement.

Discussion

To our knowledge, this is the first case report that associates Kikuchi's disease with discoid lupus erythematosus instead of systemic lupus erythematosus.

REVIEW ON KIKUCHI'S DISEASE (HISTIOCYTIC NECROTISING LYMPHADENITIS)¹

Kikuchi's disease (Histiocytic necrotising lymphadenitis) was first described in Japan in 1972 by Kikuchi.² The aetiology of Kikuchi's disease is unknown. Recently a link between Kikuchi's disease and SLE has been suggested. Up to 1997, only 17 cases

of Kikuchi's disease have been reported in association with SLE. The disease had been diagnosed after, simultaneous, or prior to the diagnosis of SLE.³ Some people speculated that both Kikuchi's disease and SLE share a common triggering factor, such as exposure to an environmental or infectious agent, which could provoke either disorder. Alternatively, Kikuchi's disease may be an autoimmune-mediated disease that can remain self-limited or develop into SLE.

Clinical Features

Kikuchi's disease primarily affects young woman of Asian or Caucasian background. The most common presentation includes fever, constitutional upset and lymphadenopathy. Majority of the patients have cervical lymph nodes enlargement whereas inguinal and mediastinal lymphadenopathy are rarely found. A minority of cases has leukopenia. ANA is usually negative.

Histology

Pathology of lymph node showed patchy paracortical necrosis with prominent histiocytes and immunoblasts. Neutrophils are usually absent, and plasma cells are rare. The pathology could be indistinguishable from that of lupus lymphadenitis. In lupus lymphadenitis, hematoxylin bodies are highly specific though not always present.

Treatment

Kikuchi's disease is self-limiting with spontaneous resolution of lymphadenopathy.

Learning points:

Patient with Kikuchi's disease must be followed up closely for lupus erythematosus.

References

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2. Kikuchi M. Lymphadenitis showing focal reticulum cell hyperplasia with nuclear debris and phagocytosis. *Nippon Ketsueki Gakkai Zasshi* 1972; 35: 379-80.
3. Martinez-Vanzquez C. Histiocytic necrotising lymphadenitis, Kikuchi-Fujimoto's disease, associated with systemic lupus erythematosus. *QJM* 1997; 90(8):531-3.