Kikuchi Fujimoto Disease

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ABSTRACT

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Kikuchi Fujimoto disease is a self limiting necrotizing lymphadenitis presenting with cervical lymphadenitis, fever, myalgia, rash & neutropenia. Rarely extra nodal involvement is seen.

The aetiology is attributed to infective or autoimmune causes. The clinical features, differential diagnosis and typical histopathological appearance is described.

Keywords: Lymphadenopathy, Necrotizing Lymphadenitis, Autoimmune, Lymphohistiocytic Infiltration

*See End Note for complete author details

INTRODUCTION

This disease was first reported from Japan by Kikuchi & Fujimoto in 1972. It is a self limiting necrotizing lymphadenitis presenting with cervical lymphadenitis, fever, myalgia, rash & neutropenia. Rarely extra nodal involvement is seen.

The descriptive term used is Histiocytic Necrotizing Lymphadenitis without granulocytic infiltration OR Sub acute necrotizing lymphadenitis.

Etiology

Unknown.But is attributed to infectious or autoimmune causes. Infections triggering the disease include Brucella, Yersinia, Toxoplasma, Viruses like Epstein-Barr virus, Herpesvirus 6, B19 parvovirus. Presence of ANA, RA factor positivity, Positive Coomb's test suggest the presence of an autoimmune process, though these patients seldom develop autoimmune dysfunction.

Pathogenesis

Plasmacytoid monocytes play a role in the pathogenesis via a cell mediated cytotoxic immune response. Histologic changes suggest a hyperimmune response of T cells, activated by an unidentified pathogen.

Pathology

Basic feature is lymphohistiocytic infiltration especially in the skin & lymph nodes. Characteristic histological features in lymph nodes are:Eosinophilic necrosis in the Para cortex/Cortex.

- Karyorrhexis with fragments of nuclear debris distributed in an irregular pattern throughout the areas of necrosis.
- Paucity of polymorphs & plasma cells in the areas of necrosis.
- Cluster of plasmacytoid monocytes in the form of nodules.

The above features will give a confident histopathological diagnosis of Kikuchi's disease. Lupus lymphadenitis may have a close resemblance to Kikuchi's disease lymphadenitis but the differentiating features are prominent plasma cell component & presence of Hematoxylin bodies. Skin biopsy also shows dense lymphohistiocytic, superficial & deep perivascular and interstitial infiltrates and absence of neutrophils; paralleling the nodal histology.

CLINICAL FEATURES

Age: Maximal incidence in the third decade. But can affect all ages from children to elderly.

Sex: Females are more commonly affected than males in the ratio of 4:1.

Clinical features are confined mostly to lymph node & skin. It presents as fever, myalgia, Subacute Posterior Cervical Lymphadenopathy. However it may also affect the anterior group, rarely other groups may be involved with generalized lymphadenopathy.

In 1/3rd of patients there is involvement of skin in the form of erythematous macules, papules, plaques,

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nodules in the upper part of the body such as face, upper limbs, trunk, occasionally the entire body.

Kikuchi's disease may be associated with cutaneous SLE.

Other features are Aseptic meningitis, Polyarthritis, Mononeuritis Multiplex, Brachial plexus neuritis, Panuveitis, Hepatocellular jaundice, Splenomegaly, Leukocytoclastic vasculitis.

Differential Diagnosis

Conditions producing lymphadenopathy such as

- Tuberculosis
- Toxoplasmosis
- Yersinia
- Cat scratch disease
- Autoimmune disorders (SLE, Kawasaki's disease)
- Lymphomas

Complications

- Hyperpyrexia
- Evolution into SLE.
- Still's disease
- Recurrence in 3 %

Laboratory Features

- Anemia, leucopenia, thrombocytopenia. Rarely (< 5% cases) Leukocytosis.
- Atypical Lymphocytes in peripheral smear.
- Coagulation abnormalities.
- ANA, RA Factor positive, Coombs test positive (in certain cases denoting autoimmunity)
- S. LDH elevated, S. Transaminases elevated, S. CRP elevated/normal.
- Flow cytometry performed on the affected lymph node, Bone marrow aspirate & peripheral blood reveals a relative expansion of mature activated T lymphocytes predominantly expressing CD8.
- Serology against EBV, CMV, Hepatitis, HIV, B19 Parvovirus, Brucella, Bartonella, Toxoplasma, Rickettsiae — are negative.

Imaging Studies

Imaging studies of lymph node show hypodense centre with peripheral ring enhancement.

MANAGEMENT

In most of the cases only supportive measures including antipyretics, are required. Use of corticosteroids are recommended in the following situations,

- Hyperpyrexia
- Aseptic Meningitis
- Uveitis
- Vasculitis
- Kikuchi's disease coexisting with SLE or presence of high titres of ANA.

Some of the patients of Kikuchi's disease are being treated as Tuberculous lymphadenitis, without any proper histological assessment.

Kikuchi's disease in a self-limiting one, usually resolves spontaneously within 2-3 months.

END NOTE

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