CT and US Features of Cervical Lymphadenopathy due to Kikuchi's Disease

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Kikuchi's disease or histiocytic necrotising lymphadenitis is a distinct benign clinicopathologic entity which is to be distinguished from malignant conditions, especially lymphoma.

We report a young woman with Kikuchi's disease presenting with cervical lymphadenopathy and mild hepatomegaly. CT and US findings are presented as well as a review of the pertinent literature

Key words :

Cervical lymphadenopathy; Kikuchi's disease; US; CT.

INTRODUCTION

Histiocytic necrotising lymphadenitis or Kikuchi's disease was described in 1972 in Japan independently by Kikuchi and Fujimoto. It is an unusual benign lymphadenopathy (Asano et al., 1990; Buckley et al., 1988) affecting more commonly young women, with predominant unilateral cervical location. It is now recognised to have worldwide distribution (Chan et al., 1989; Dorfman and Berry, 1988). Its aetiology is unknown, however, the clinical course is benign and self limited. Histologically it is characterised by foci of necrosis with histiocytic cellular infiltrate, mononuclear cells and phagocytic macrophages. Large series of Kikuchi's disease were reported in the pathological literature (Chan et al., 1989; Fulcher, 1993). Only a few cases were reported in the radiological literature (Fujimoto et al., 1970; Inamura et al., 1982; Kikuchi, 1972).

Kikuchi's disease must be distinguished from

malignant lymphoma or other lymphnode malignancies, avoiding unnecessary surgery, radiotherapy or chemotherapy (Kikuchi, 1977).

We report a young patient with classical clinical presentation. The radiological findings of Kikuchi's disease are discussed.

CASE REPORT

A 33-year-old woman was referred to the Otolaryngology Department with a ten-day history of slow-growing, right cervical mass. She had intermittent low grade fever, decreased appetite and night sweats. She lost 3 kgs, over a period of two weeks. She denied diarrhoea, rashes or abdominal pain. Her past medical history was otherwise unremarkable. On examination a tender and mobile 2 cm in diameter mass was palpated in the right mid-jugular region. Multiple, sparse, small lymph nodes were found in the left posterior triangle area. The rest of her otolaryngological examination was normal, as well as the rest of her physical examination except for mild hepatomegaly. Laboratory studies showed a slight neutrophil leukocytosis (white cell count 8,300 with a shift to the left). Haemoglobin was 12 g/dl and the rest of the biochemical profile was normal. Serologic titres for infectious agents such as Epstein Barr, cytomegalovirus, human immunodeficiency virus, monospot test, toxoplasmosis and brucella, were all negative.

Immunologic studies, including total complement activity, C₃, C₄ antinuclear factor, and rheumatoid factor were negative. US examination of the neck revealed right cervical lymphadenopathy, involving the posterior

triangle, jugulodigastric and supraclavicular areas. Most nodes were oval-shaped, but some presented rather a round shape. Their size was different with homogenous echogenic structure or target-like pattern. No vascular involvement was seen (Fig.1) Abdominal US (not presented) showed mild hepatomegaly, without evidence of lymphadenopathy.

Fig-1

US of neck, longitudinal view, shows different size and shape of nodes involving the jugulodigastric area.

Contrast enhanced axial CT of the neck showed extensive right lymphadenopathy in the posterior triangle and jugulodigastric areas. Chest and abdominal contrast enhanced axial CT failed to demonstrate lymphadenopathy. Mild hepatomegaly was present.

Some lymph nodes enhanced homogenously with contrast material, others presented lower density, but without evidence of necrotic changes. The right sternocleidomastoid muscle was slightly enlarged. The vascular structures and subcutaneous fat were normal (Fig.2).

Her adenopathy failed to respond to i.v. antibiotic therapy (cefuroxime and metronidazole). A fine needle aspiration of the right cervical mass was inconclusive. Excisional biopsy of the largest cervical lymph node was performed. The pathologic examination revealed an extensive necrotising process. The presence of karyorrhexis, paracortical areas of necrosis and the absence of neutrophils, eosinophils and plasma cells were

consistent with Kikuchi's disease. On follow-up at nine months there was no evidence of lymphadenopathy or other complaints.

DISCUSSION

Kikuchi's disease, was first described in Japan, and later reported in Europe and the United States as a distinct clinico-pathological entity (Fulcher, 1993; Inamura et al., 1982; Kikuchi, 1972). This disease occurs typically in young patients. The ages range from 10 to 75 (mean 30 years) and it is four times more common in females. No race or occupational relationship was found. The symptoms present 1-24 months (mean 3 months) before the diagnosis. The largest series of 108 cases was published by Dorfman and Berry (1988).

The classical clinical manifestation is unilateral

Fig-2

Contrast enhanced CT of the neck shows right posterior triangle and jugulodigastric lymphadenopathy with a different appearance. Some present homogenous contrast enhancement., No necrotic changes were present.

cervical lymphadenopathy, with predominance of the posterior triangle group. Other possible affected lymph node groups are axillary, supraclavicular, mediastinal, retroperitoneal, inguinal, iliac, intraparotid or generalised. Tenderness of lymph nodes or pain may be present (Fulcher, 1993; Kikuchi, 1972). Fever occurs in half of the patients. Hepatomegaly or splenomegaly are rare. Other reported symptoms include; weight loss, nausea, vomiting, diarrhoea, chills, sweats, sore throat, chest pain, lower abdominal quadrant pain, myalgia, and arthralgia. Rash or cutaneous plaques are only rarely found (Kikuchi, 1978).

The laboratory data may be normal. Neutropenia or lymphocytosis with atypical lymphocytes may be present. The serologic tests for Epstein-Barr virus, toxoplasmosis, cytomegalic virus, monospot test, influenza, adenovirus, mycoplasma and a fever are negative. Autoimmune studies, including rheumatoid factor, antinuclear bodies and lupus erythematosus are reported to be negative (Fulcher, 1994; Inamura et al., 1982).

The disease is a self-limiting autoimmune condition with unknown aetiology, and most patients recover without treatment in 2-3 months. The aetiology and pathogenesis of Kikuchi's disease remain unclear. A viral-induced autoimmune body response was considered by Imamura (Kuo, 1990; Ogana et al., 1991). Other authors suggested that this condition might represent an atypical form of toxoplasmosis (Pileri et al., 1982; Rudniki, 1988). The diagnosis is based on fine needle aspiration or excisional biopsy (Fulcher, 1993). A fatal case of multicentric Kikuchi's disease was reported by Chan in a patient with acute congestive heart failure (Steinkamp et al., 1992).

Histopathologic studies show the paracortical area to be the most affected, with "punched-out" appearance, because of marked lymphocytic depletion. Sometimes, the lesion may be located interfollicularly, perinodally or subcapsularly. The typical histologic features are: large number of immunoblasts, histiocytes and macrophages with nuclear debris, absence of granulocytes, paucity

of plasma cells and a variable degree of necrosis with extensive karyorrhexis. In cases with limited necrosis, atypical mononuclear cells may suggest lymphoma (Fulcher, 1993; Inamura et al., 1982; Kikuchi, 1972; Kikuchi et al., 1977). The ultrastructural studies revealed tubular reticular structures in the cytoplasm of stimulated lymphocytes and histiocytes, which are frequently seen in viral infection, SLE, SLE-related autoimmune disease and neoplasia (Sterman et al., 1989).

The differential diagnosis should include; malignant lymphoma, Hodgkin's lymphoma, Yersinia infection, systemic lupus erythematosus, cat scratch disease, toxoplasmosis, infectious mononucleosis, AIDS, lymph node infection, Kawasaki's disease (Fulcher, 1993). It is imperative to distinguish Kikuchi's disease from lymphoma in order to avoid extensive investigation, unnecessary surgery and chemotherapy or radiotherapy.

In spite of a large series of Kikuchi's disease presented in the pathologic literature, only a few case reports are found in the radiologic literature. Ogawa et al present the ultrasonographic features in a patient with Kikuchi's disease (Fujimoto et al., 1970). Fulcher (1993) presents a classical cervical Kikuchi's disease with US and CT patterns. Rudniki et al. (1988) present an unusual case of Kikuchi's disease with splenomegaly, paraaortic and parailiac lymphadenopathy.

We report a typical case of Kikuchi's disease with cervical lymphadenopathy and classical clinical presentation. The lymph nodes were well delineated by US and CT. The two imaging modalities did not reveal necrosis. In US some nodes showed an increased short axis bringing the long/short axis ratio to less than 1.5. This criteria may suggest malignancy (Turner et al., 1983). The large vessels and adjacent soft tissue were not involved.

In conclusion, the diagnosis of Kikuchi's disease should be considered in cases of isolated cervical lymphadenopathy in young females. The

condition should also be added to the list of possible causes of fever of unknown origin. Since Kikuchi's disease may mimic clinically, radiologically and histopathologically,

lymphoma and other malignant conditions of lymphadenopathy, errors can be avoided by the awareness of this benign, self-limiting clinicopathologic condition.

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Congratulation !

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