

Kikuchi's Disease of the Mesenteric Lymph Nodes Presenting as Acute Appendicitis

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Kikuchi's disease is a benign self-limiting necrotizing lymphadenitis that occurs most commonly in young women, and is usually found in the cervical lymph nodes. When there is an unusual location of involved lymph nodes, the diagnosis can be difficult. We recently treated a patient with Kikuchi's disease who had ileocecal mesenteric lymph node involvement; the patient presented with symptoms of acute appendicitis in an 11-year old boy. Although mesenteric lymph node involvement of Kikuchi's disease is very rare, Kikuchi's disease should be added to the differential diagnosis of acute appendicitis in patients with enlarged ileocecal mesenteric lymph nodes on radiological evaluation.

Key Words : Kikuchi's disease; Mesentery; Lymph node; Appendicitis

Kikuchi's disease was first described by Kikuchi¹ and Fujimoto *et al.*² in 1972. They reported the distinctive histological appearance of the lymph nodes in descriptive terms such as "lymphadenitis showing focal reticulum cell hyperplasia with nuclear debris and phagocytosis" and "cervical subacute necrotizing lymphadenitis". This disorder is usually found in the cervical lymph nodes, less frequently in the axillary nodes and rarely presents as a generalized lymphadenopathy.³⁻⁵ Kikuchi's disease localized to the mesenteric lymph nodes is extremely unusual. Only a few reports have been previously described in the medical literature.^{3,6-10} Here we report an additional case of Kikuchi's disease with ileocecal mesenteric lymph node involvement that clinically presented as acute appendicitis.

CASE REPORT

An 11-year-old boy was transferred to our hospital with the impression of acute appendicitis based on the findings of the

local clinic. The patient presented with nausea and had vomited seven times in two days. Fever (39.2°C) and right lower abdominal pain developed one day prior to presentation. The past history showed bilateral tonsillectomy due to reactive follicular hyperplasia two years ago. Physical examination revealed rebound tenderness in the right lower quadrant of the abdomen. Serum electrolytes, liver function tests and routine urine analysis were within normal limits. C-reactive protein and erythrocyte sedimentation rate were elevated (2.52 mg/dL, 16 mm/h). The leukocyte count was elevated at 13,000/ μ L with 85% of neutrophils. Serologic and microbiologic tests for tuberculosis, systemic lupus erythematosus (SLE), Yersinia infection, cat scratch disease, Toxoplasma lymphadenitis and infectious mononucleosis were not performed. The abdominal plain radiograph was unremarkable. The abdominal computerized tomography showed suspicious features for acute appendicitis with multiple enlarged mesenteric lymph nodes around the ileocecal area (Fig. 1). With the impression of acute appendicitis, the patient underwent laparotomy via right oblique skin incision. A normal appearing appendix was



Fig. 1. An axial scan of contrast-enhanced abdominal CT revealed multiple homogeneously enhanced mesenteric lymph nodes (arrows) at ileocecal area.

located at the antececal area. Several enlarged ileocecal lymph nodes were present. The ileocecal mesentery showed mild congestion. Appendectomy and excision of a mesenteric lymph node were performed. On gross examination, the appendix was unremarkable. The mesenteric lymph node was enlarged and measured 1 cm in diameter. On microscopic examination, the lymph node showed the histological features of Kikuchi's necrotizing lymphadenitis including patchy necrotic lesions localized to the paracortical areas, marked proliferation of benign histiocytes with crescent or C-shaped form, necrosis with extensive karyorrhexis, marked phagocytic activity and mild perinodular polymorphous infiltrates (Fig. 2). Evaluation with periodic acid-Schiff and silver stains, to rule out fungal infection, were negative. The appendix was well preserved without evidence of acute inflammation. Recovery was uneventful and the patient was discharged three days after surgery.

DISCUSSION

Kikuchi's disease (Necrotizing lymphadenitis) is a well documented, benign self-limiting condition that usually presents with cervical lymphadenopathy and often with a fever of unknown origin.^{1,6,8} The etiology of Kikuchi's disease remains unknown. Although there is no proven etiological agent to date, virus, *Toxoplasma* or *Yersinia* are currently favored possibilities.⁵⁻⁷

The disease occurs much more commonly in young women than men, with most patients being under the age of 40 years.^{3,5} The classical presentation is one of painful tender cervical lymphadenopathy associated with pyrexia and frequently arthralgia and rash. The diagnosis is usually made by excision biopsy to

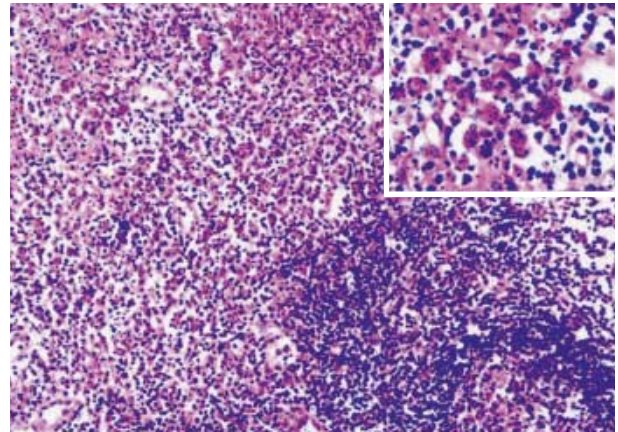


Fig. 2. The lymph node showed the characteristic histologic features of Kikuchi's disease including marked proliferation of histiocytes, necrosis with extensive karyorrhexis, and marked phagocytic activity (inset).

exclude either the clinical diagnosis of tuberculosis or lymphoma.⁷ The mean age of patients with mesenteric Kikuchi's disease in previously reported cases was 21.8 years (range, 12-29 years). Our case was an 11-year-old boy.

The histological features of Kikuchi's disease are characteristic. The involved lymph nodes have patchy necrotizing features localized mainly to the paracortical areas. The necrotizing areas show eosinophilic fibrinoid material associated with a striking degree of karyorrhexis. Fragments of nuclear debris are distributed irregularly throughout areas of necrosis associated with the presence of apparently atypical mononuclear cells. There is a mixture in variable proportions of benign histiocytes (so-called crescent or C-shaped forms), immunoblasts, plasmacytoid monocytes, and small lymphocytes. Granulocytes are absent and plasma cells are rare or absent. The capsule is intact and the perinodal adipose tissue may contain a polymorphous infiltrate.^{4,5,8,11} The histological findings in our case were consistent with the characteristics of Kikuchi's disease.

The main differential diagnosis includes several necrotizing lesions associated with SLE, *Yersinia* infection, cat scratch disease, *Toxoplasma* lymphadenitis and infectious mononucleosis. The histological features of SLE include a prominent infiltration of plasma cells, the presence of hematoxyphilic bodies aggregated toward the edges of the acellular necrotizing areas and the deposition of DNA on the vessel walls. In our case, infiltration of plasma cells and hematoxyphilic bodies were not present and the blood vessels were unremarkable. A *Yersinia* infection is characterized by the presence of microabscesses with polymorphonuclear leukocytes localized to germinal centers and aggregates of epithelioid histiocytes producing a granulomatous reaction. In

cat-scratch disease, the most characteristic finding is that of stellate microabscesses containing numerous granulocytes surrounded by palisading histiocytes. The histological findings of *Toxoplasma* lymphadenitis are characterized by florid follicular hyperplasia and clusters of epithelioid histiocytes in close proximity or infiltrating reactive germinal centers of follicles, and prominent monocytoid B-cell proliferation within sinuses. Infectious mononucleosis shows prominent follicular hyperplasia and a florid immunoblastic proliferation in the interfollicular areas.^{4,11} In our case, the features of granulomatous inflammation and neutrophilic infiltration to form an abscess or follicular hyperplasia were not present.

An unusual location of lymph node involvement can make the diagnosis of Kikuchi's disease difficult. Kikuchi's disease usually presents as solitary or multiple enlarged cervical lymph nodes and only rarely with generalized lymphadenopathy.^{3-5,8} Rare sites of extracervical lymphadenopathy include axillary, supraclavicular, mediastinal, inguinal, intraperitoneal, iliac, celiac, and peripancreatic.⁴ However, localization of Kikuchi's disease to the mesenteric lymph nodes is quite unusual. Only eight cases have been previously described in the medical literature.^{3,6-10} Of note is that the enlarged lymph nodes were located in the mesentery of the cecum and the distal ileum in all of the previously reported cases. All of these patients underwent laparotomy with the clinical impression of acute appendicitis. In our case, the patient's clinical symptoms and laboratory data were consistent with acute appendicitis; enlarged lymph nodes were located in the mesentery of the cecum and the distal ileum. However, there was no evidence of appendicitis on histological examination of the appendix.

Acute appendicitis is the most common diagnosis made in patients with symptoms of an acute abdomen. However, there are many conditions that mimic appendicitis including mesenteric adenitis, infectious enterocolitis, gynecological disease, fibrotic appendix, colonic diverticulitis, urinary tract infection, urolithiasis, small bowel obstruction, regional ileitis, cholecystitis, and Meckel's diverticulitis.^{12,13} Among these conditions, mesenteric adenitis is the second most common cause of right lower quadrant pain in patients suspected of having appendicitis.¹²

When Kikuchi's disease involves the mesenteric lymph nodes of the ileocecal region, the clinical symptoms and laboratory findings are similar to those of acute appendicitis as in previously reported cases^{3,6-10} and our case. Although the occurrence is ext-

remely rare, Kikuchi's disease with involvement of the ileocecal mesenteric lymph nodes should be considered in the differential diagnosis of acute abdominal pain mimicking acute appendicitis.

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