Kikuchi-Fujimoto Disease: An Unusual Presentation with Isolated Abdominal Lymphadenitis

Osama Sharaf, MBCHB, MSc, FRCR* Ahmad Ali, MBCHB, MSc**

Kikuchi-Fujimoto Disease (KFD) is a rare and benign cause of lymphadenopathy that is often associated with fever, night sweats and weight loss. The differential diagnosis could be lymphoma, tuberculosis, sarcoidosis and systemic lupus erythematosus. Lymph node biopsy confirms the diagnosis.

A thirty-three-year-old female presented with abdominal pain for 3 months and fever, weight loss of 5 kg, no nausea or vomiting and no bowel habit changes. Biopsy of the abdominal lymph nodes confirmed the diagnosis of KFD. The patient received a course of antibiotics, hydrocortisone and immunoglobulin IV 10 g/100 ml. Post-treatment, the patient's abdominal pain and general condition had improved and CT revealed significant regression of the abdominal lymph node.

Bahrain Med Bull 2017; 39(4): 244 - 246

Kikuchi-Fujimoto Disease (KFD) is a benign, self-limited disease; it was first described in 1972 by Kikuchi and Fujimoto in Japan[1,2]. It is associated with systemic lupus erythematosus, non-infectious inflammatory conditions, viral infection and is more common in females (77%), especially those of Asian origin[3].

The disease frequently mimics tuberculous lymphadenitis and malignant lymphoma and may be mistaken for and treated as lymphoma[4-6].

The disease is characterized by enlarged lymph nodes, mainly the cervical; involvement of abdominal lymph nodes is unusual.

Kikuchi-Fujimoto Disease (KFD) is a rare and benign cause of lymphadenopathy that is often associated with fever, night sweats and weight loss. The clinical and laboratory manifestations of KFD are similar to those of lymphoma, tuberculosis, sarcoidosis and systemic lupus erythematosus. Definitive diagnosis is accomplished by lymph node biopsy[6,7]. Awareness of KFD among clinicians and pathologists is essential to avoid misdiagnosis and inappropriate treatment of this self-limited disorder.

KFD is an uncommon and benign disorder and should be considered by the clinicians whenever evaluating patients with lymphadenopathy and fever; this would avoid unnecessary invasive testing with subsequent complications.

The aim of this report is to increase the awareness of the abdominal lymphadenitis diagnosed as Kikuchi-Fujimoto lymphadenitis, a benign self-limiting disorder that could be mistaken for other serious diseases.

THE CASE

A thirty-three-year-old female presented with abdominal pain for three months associated with fever and weight loss of 5 kg but no nausea, vomiting or bowel habit changes. Clinical examination revealed low-grade fever of 38°C and vital signs were normal.

The laboratory investigations were as follows: Anti-CCP ABS was normal, Antineutrophil cytoplasmic antibody, Anti-SSA, hepatitis B and Beta-2 Glycoprotein 1 IgG were negative. Tuberculin skin test was negative. Serological tests for Epstein Barr virus, cytomegalovirus, HIV and toxoplasma were negative.

CT scan of the abdomen and pelvis revealed multiple mesenteric and retroperitoneal enlarged lymph nodes around the celiac axis and paraaortic groups extending around the iliac arteries with many necrotic lymph nodes, see figures 1 and 2.

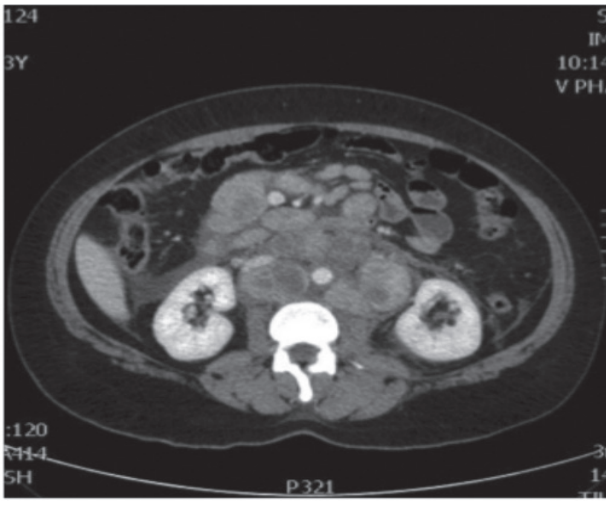

Figure 1: Axial CT Scan Showing Multiple Mesenteric and Retroperitoneal Enlarged Lymph Nodes with Many Necrotic Lymph Nodes

* Associate Consultant
** Assistant Consultant
Adult Medical Oncology
Radiology Department
King Fahad Specialist Hospital Dammam
Kingdom of Saudi Arabia
E-mail: osamasharaf66@hotmail.com

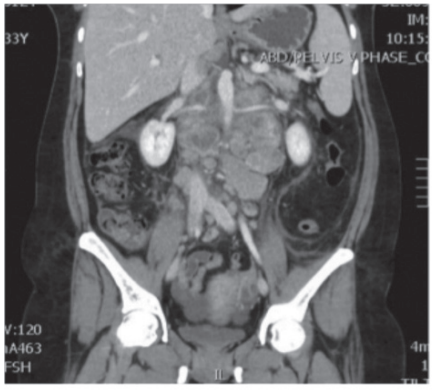

Figure 2: Coronal CT Scan Showing the Retroperitoneal Necrotic Large Lymph Nodes

The patient received a course of antibiotics (Cefazolin 1g intramuscular injection), hydrocortisone and immunoglobulin IV 10g/100 ml. Post-treatment, the patient's abdominal pain, and general condition had improved; follow-up CT of the abdomen and pelvis revealed significant improvement of the abdominal lymph nodal enlargement, see figures 3 and 4.

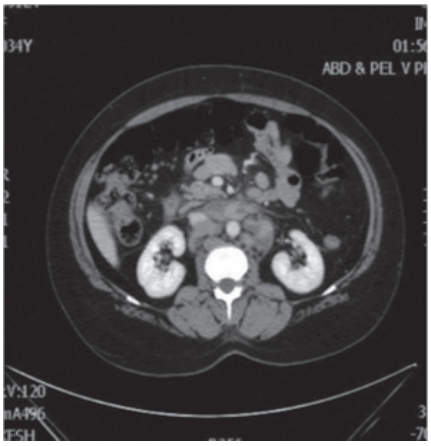

Figure 3: Axial CT Scan Showing Significant Regression of the Multiple Mesenteric and Retroperitoneal Enlarged Lymph Nodes

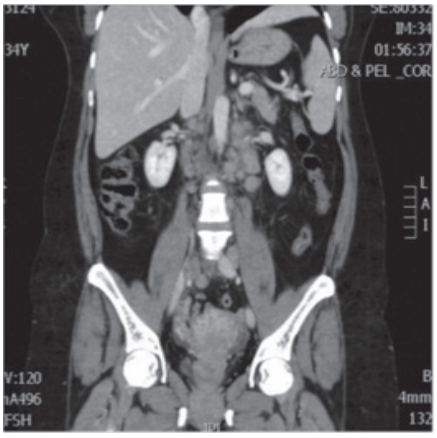

Figure 4: Coronal CT Scan Showing Interval Improvement

The differential diagnosis includes lymphoma, tuberculous lymphadenitis, HIV and systemic lupus erythematosus. The diagnosis of KFD was made as the patient's general condition has improved and the follow-up revealed a progressive reduction of the diseased lymph nodes. Biopsy of the abdominal lymph nodes was performed which revealed the diagnosis.

DISCUSSION

Kikuchi-Fujimoto disease (KFD) is a self-limited disease of the lymph nodes; cervical nodes are most commonly involved, while abdominal lymph nodes involvement is rare[1,2].

The disease mostly affects young individuals around 30 years old, and although less common, children are also affected[8,9]. The etiology of the disease is not definite yet; however, viral infection is believed to be a reason supported by the self-limited resolving condition and slow, insidious onset. Autoimmune etiology is also likely, as reported recently in literature[10].

Females are more prone than males; however, recent reports suggest equal prevalence of both genders[3]. The main clinical presentation is lymphadenopathy, persistent low-grade fever, weight loss, malaise, sweating and anorexia[11]. An open biopsy is the only reliable procedure to establish the diagnosis, however, according to some authors; a fine-needle biopsy may also be helpful[6].

Follow-up of these cases is important as recurrences have been reported. In addition, the expected regression with treatment is essential in differentiating the condition from other possibilities, especially lymphoma and tuberculous lymphadenitis[12].

CONCLUSION

Abdominal lymphadenopathy is a rare presentation in Kikuchi-Fujimoto disease; however, the diagnosis of KFD, particularly in young females with normal general health presenting with abdominal lymphadenopathy and fever should be considered as the differential diagnosis.

Treatment differs significantly from the other conditions such as SLE, lymphoma and TB. Lymph node biopsy would reveal accurate diagnosis.

Author Contribution: All authors share equal effort contribution towards (1) substantial contribution to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of manuscript version to be published. Yes.

Potential Conflicts of Interest: None.

Competing Interest: None.

Sponsorship: None.

Acceptance Date: 19 September 2017.

Ethical Approval: Approved by the Adult Medical Oncology Unit, King Fahad Specialist Hospital, Saudi Arabia.

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