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Kikuchi-Fujimoto Disease: A Rare Cause of Intestinal Obstruction

Dr. Basavaraja C[1], Dr. Aniruddha Desai[2], Dr. Anjana Kaza[3], Dr. Siddharth Kalke[4]

[1]Assistant Professor, Dept. of General Surgery, JJM Medical College, Davangere

[2, 3, 4]Post Graduate Resident, Dept. of General Surgery, JJM Medical College, Davangere

Abstract: ***Kikuchi–Fujimoto Disease (KFD) was first described in Japan in 1972. It is a benign and self-limited disorder, characterized usually by regional cervical lymphadenopathy with tenderness, usually accompanied with mild fever and night sweats. Kikuchi-Fujimoto disease is an extremely rare disease. Diagnosis requires histopathologic examination and exclusion of other factors by ancillary studies. Treatment involves supportive measures, and the symptoms usually resolve spontaneously within 4 months. Although a rare presentation, Kikuchi disease can present similarly to Abdominal Tuberculosis & treatment approach varies. Necrotic lymph nodes should raise the suspicion of the disease. Here we present to you a case report of a similar case with a disease presenting similar to tuberculosis but yet completely different from it.***

Keywords: Kikuchi Disease; Intestinal Obstruction; Lymphadenitis

1. Introduction

Kikuchi–Fujimoto Disease (KFD) was first described in Japan in 1972. The disease frequently mimics tuberculous lymphadenitis, malign lymphoma, and many other benign and malignant conditions.(1) It is a benign and self-limited disorder, characterized usually by regional cervical lymphadenopathy with tenderness, usually accompanied with mild fever and night sweats. Less frequent symptoms include weight loss, nausea, vomiting, sore throat. Kikuchi-Fujimoto disease is an extremely rare disease known to have a worldwide distribution with higher prevalence among Japanese and other Asiatic individuals.(2) Diagnosis requires histopathologic examination and exclusion of other factors by ancillary studies. Non-Hodgkin lymphoma and systemic lupus erythematosus should be ruled out before diagnosis of Kikuchi-Fujimoto disease, given the overlapped clinical and histologic features as well as the different therapeutic approaches. Treatment involves supportive measures, and the symptoms usually resolve spontaneously within 4 months.(3)

Intestinal Obstruction is one of the common emergencies found in the emergency room of most of the district hospitals in India. According to data, adhesions following previous surgery and tuberculosis remain the most common causes of this emergency in our country. It is in the subconscious of a surgeon practising in India to conclude tuberculosis of the abdomen after noticing adhesions with multiple lymphnodes with inflammation and infected ascites in a patient presenting with intestinal obstruction.

Here we present to you a case report of a similar case with a disease presenting similar to tuberculosis but yet completely different from it. There are very few reports of the disease causing intestinal obstruction as it is usually associated with cervical lymphadenopathy.

2. Case Report

A 43 years old adult male presented to the hospital with distension of abdomen, pain in abdomen, vomiting and constipation since 5 days. On and off abdominal pains were present since the past 1 year. Patient had previous history of hospitalizations for the abdominal pain 2 months back and was managed conservatively for the same with analgesics and antibiotics. The details of the previous hospitalization were unavailable with the patient. There was no history of previous surgery. Patient did not give history of any comorbid conditions including Tuberculosis, Diabetes, Asthma, Epilepsy or Hypertension.

On examination, the patient was in a state of distress lying uncomfortably on the examination bed due to the abdominal pain. Pulse rate was 102 beats per minute and blood pressure was 90/60 mm of Hg. On per abdominal examination, the abdomen was noted to be distended, tense with guarding. There was diffuse tenderness and a uniform hyperresonant note heard all over the abdomen. Per rectal examination was normal and showed a collapsed rectum.

The patient was shifted to the radiology room where an Erect X-ray of the abdomen and an Ultrasound of the abdomen were performed. Erect X-ray showed multiple air fluid levels with characterless loops in the central region, suggestive of small bowel obstruction. Ultrasound of the abdomen showed dilated, fluid-filled bowel loops with thickened bowel walls & minimal peristalsis. Multiple mesenteric lymph nodes were seen to be enlarged.

Patient was shifted to the Operation theatre and an exploratory laparotomy was proceeded with. The abdomen was opened with a lower midline incision. Multiple firm to flimsy adhesions were noticed in the abdominal cavity. There was kinking of the ileum loops over the mesentery. The ileal loops were dilated with multiple adhesions attached between the bowel, abdominal wall and mesentery. Multiple dark bluish coloured discreet lymphnodes were noted in the mesentery (Fig. 1,2). The lymph nodes were

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excised for biopsy (Fig. 3).Adhesiolysis was performed and after a thorough wash the abdomen was closed.

Histopathology showed Lymph nodes demonstrated architecture partially effaced by paracortical expansion composed of circumscribed foci of apoptotic necrosis with abundant karyorrhectic debris and numerous histiocytes of different types at the edge of the necrotic foci suggestive of histiocytic necrotizing lymphadenitis i.e. Kikuchi-Fujimoto disease. (Fig. 4)

In the post-operative period, the patient was started on NSAIDs and antibiotic prophylaxis. The patient recovered comfortably over a week. Sutures were removed on the 10th post-operative day and the patient was discharged with oral NSAIDs, PPIs and Antibiotics.

On regular follow ups over two weeks each done thrice, the patient showed good recovery without having a recurrence of any of the symptoms. There was no difficulty in passage of stools or flatus either.

3. Discussion

Kikuchi–Fujimoto Disease (KFD) was first described in Japan in 1972. The disease frequently mimics tuberculous lymphadenitis, malign lymphoma, and many other benign and malignant conditions.[1]

It is an enigmatic, benign and self-limited syndrome characterized by regional lymphadenopathy with tenderness, predominantly in the cervical region, usually accompanied with mild fever and night sweats.[4]

Affected patients are most often young adults under the age of 30 years; the disease is seldom reported in children.[5]

There is much speculation about the cause of KFD; infection or autoimmune has been suggested. Some initial reports hinted at Yersinia enterocolitica and Toxoplasma gondii as possible causative agents of KFD, mainly on the basis of positive serologic test results. The role of Epstein-Barr virus (EBV), as well as other viruses, in the pathogenesis of KFD remains controversial.[6]

KFD is a self-limiting disorder that does not require any specific management. The patients with KD require a systemic survey and regular follow-up for several years. An effective communication between the surgeon and pathologist is needed because the clinical and pathologic characteristics of KD are essential in making an accurate diagnosis.[5]

4. Conclusion

Although a rare presentation, Kikuchi disease can present similarly to Abdominal Tuberculosis & treatment approach varies. Necrotic lymph nodes should raise the suspicion of the disease. Patients should be evaluated for other auto-immune disorders like SLE, etc.

It is important to differentiate between KFD and Tuberculosis as the treatment protocol varies in both. KFD has a better outcome with early therapy and regular follow up and hence one should have an eagles eye for suspecting & diagnosing the same.

5. Figures

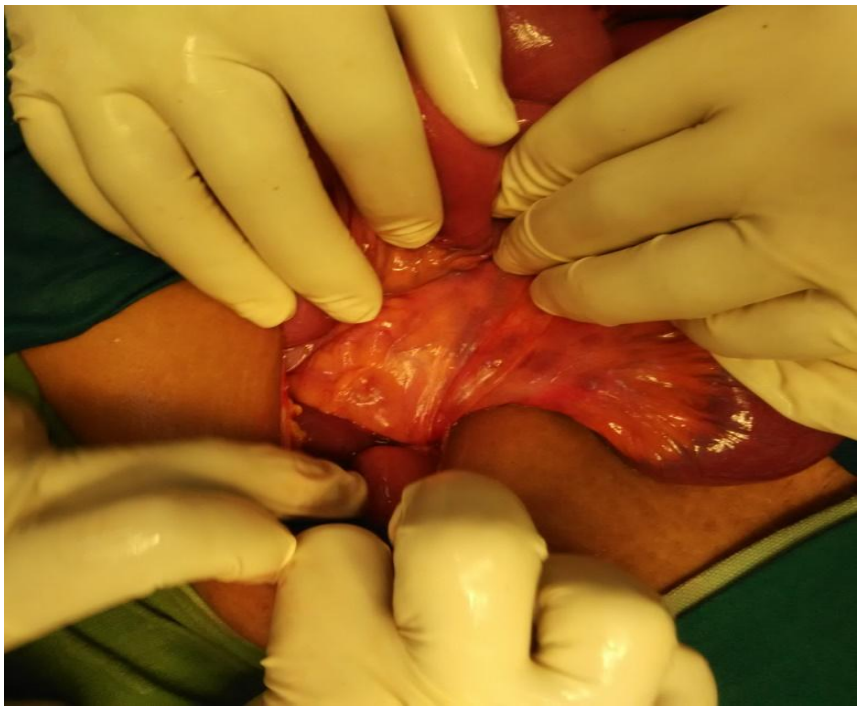

Figure 1: Multiple dark bluish coloured discreet lymph nodes noted in the mesentery

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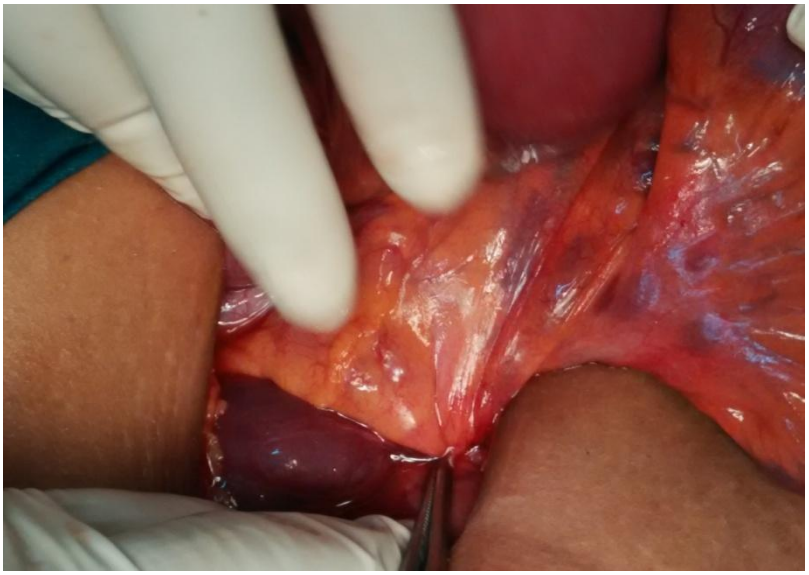

Figure 2: Multiple dark bluish coloured discreet lymph nodes noted in the mesentery.

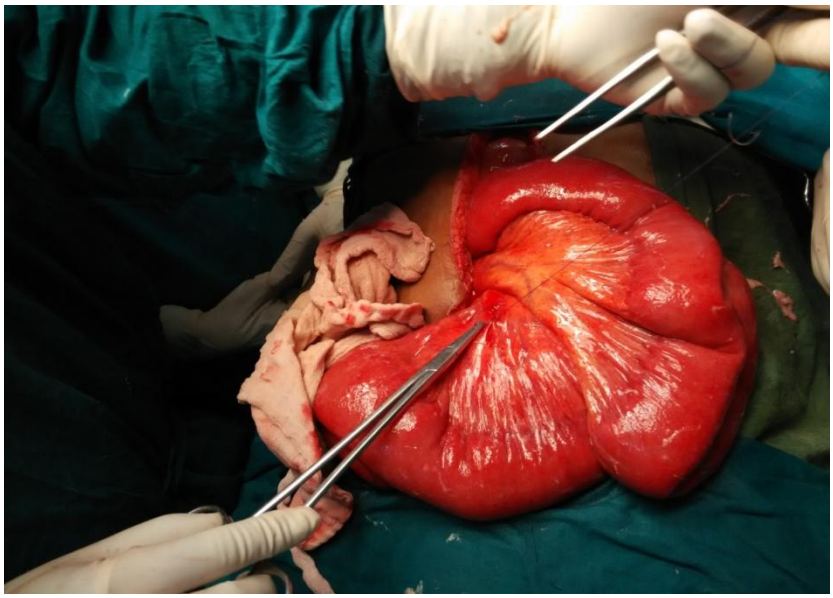

Figure 3: The Mesentric lymph nodes excised for biopsy

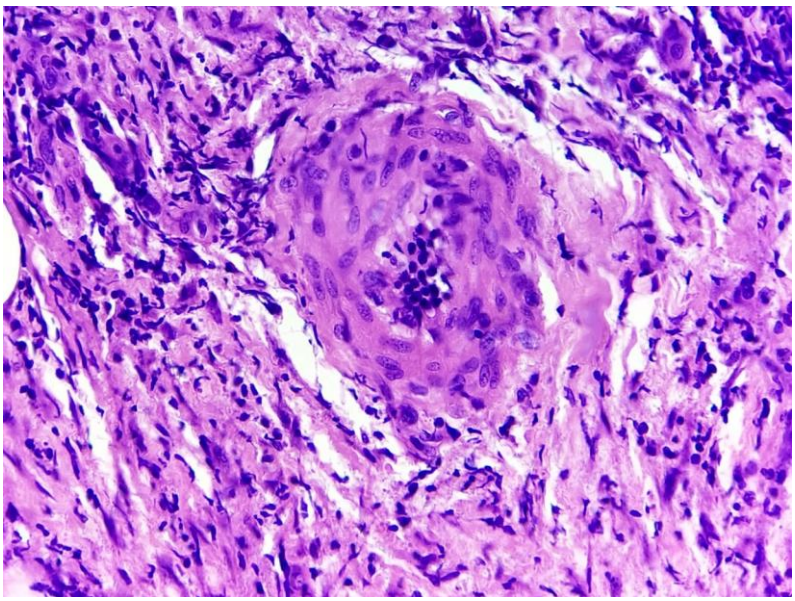

Figure 4: Histopathology suggestive of histiocytic necrotizing lymphadenitis, Kikuchi-Fujimoto disease

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