

Case report

Kikuchi Lymphadenitis- a rare case report

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Abstract:

Kikuchi-Fujimoto disease or Histiocytic necrotizing lymphadenitis is a benign, self limiting condition with higher prevalence in Asians especially Japanese. Though the etiology of this disease remains unknown, viral or autoimmune origin of the disease has been suggested. It is clinically characterized by lymphadenopathy, fever, skin rashes/ erythema, and other systemic manifestations like diarrhea, vomiting, sore throat, arthralgia, myalgia and hepatosplenomegaly. Laboratory findings are usually non-specific and Kikuchi disease is generally diagnosed based on characteristic histopathological findings of the involved lymphnode. Its recognition is very important as one of the close differentials is a systemic lupus erythematosus or a malignant lymphoma. Clinicians and pathologists knowledge of the disease helps prevent misdiagnosis and provide appropriate treatment to the patients. Here we present a case of a young teenaged girl presenting with a tender right cervical lymphadenopathy and fever. A diagnosis was made on biopsy after repeated aspirations and clinical management strategies could not establish any diagnosis.

Introduction:

Kikuchi-Fujimoto disease (KFD) was first described in Japan by Kikuchi and Fujimoto in 1972, (1,2) and has been called variously called as Kikuchi disease, necrotizing lymphadenitis, histiocytic necrotizing lymphadenitis, histiocytic necrotizing lymphadenitis without granulocytic infiltration, Kikuchi necrotizing lymphadenitis, subacute necrotizing lymphadenitis, and phagocytic necrotizing lymphadenitis. It is a very rare cause of lymphadenopathy. In spite of many case reports and numerous reviews, the etiology of the disease remains unknown. A viral or an autoimmune mechanism has been suggested but not fully established (3). The disease commonly affects young women (4) and patients usually present with cervical lymphadenopathy, However, rarely it can affect other

locations like the axilla, mediastinum, and retroperitoneum. (4) Systemic manifestations like fever or flu-like symptoms, malaise, weight loss, loss of appetite, nausea, vomiting, diarrhea, chest pain, splenomegaly and hepatomegaly have been reported. (5,6) Laboratory tests are mostly nonspecific with an elevated Erythrocyte Sedimentation rate (ESR), Leukocytosis with Neutropenia, atypical lymphocytosis etc., (7) The diagnosis of KFD is established by identifying characteristic histopathologic features in the effected lymph node. Presence of areas of necrosis with intermingled distinctive histiocytes with karyorrhetic debris and absence of neutrophils gives a clue to the diagnosis of KFD (8). The histological differential diagnosis of KFD includes Hodgkin's and non-Hodgkin's

lymphoma, lupus lymphadenitis, tuberculosis, Kawasaki's disease, and even metastatic deposits (4). It is a self limiting disease and most patient's symptoms resolve in 1-4 months of time (9).

Case presentation:

An 18 year old young female attended the general out patient department with fever of one week duration. On examination the patient was febrile and there was a tender right cervical lymph node in the posterior cervical region measuring 2 X 1cm. Routine investigations like complete blood picture and ESR revealed only mild lymphocytosis and an elevated ESR. There were no atypical cells noted in the peripheral smear. An FNAC done revealed only a polymorphous population of lymphocytes and histiocytes suggesting a Chronic non specific lymphadenitis (fig 1). Chest radiograph was normal, Tuberculin skin test was negative, Serology for HIV was non reactive. The patient was admitted and the lymphadenopathy did not resolve on a course of antibiotics with antipyretics. Subsequently a biopsy of the lymphnode was performed. In the department of Pathology we received a small grey white soft tissue bit measuring 1X1 cm. The cut surface revealed few necrotic areas along with solid grey white areas. The whole of the tissue was processed and stained with hematoxylin and eosin. On examination of multiple sections, there were areas of necrosis with Karyorrhectic debris and numerous histiocytes along with polymorphous population of lymphocytes (fig 2-4). There were no neutrophils seen in comparison to the amount of necrosis, thus ruling out the possibility of acute suppurative Lymphadenitis. There were no Malignant cells seen in the section. Thus based on the clinical manifestations and the histopathological examination a diagnosis of Kikuchi disease was made. The patient

was treated symptomatically with antipyretics and steroids. The patient responded to treatment and the lymphadenopathy completely resolved in three months duration.

Discussion:

Kikuchi disease is a very rare cause of lymphadenitis with a predilection to female population in their third or fourth decade of life (10). It was first described in Japan by two pathologists separately in 1972 namely Kikuchi and Fujimoto (1). The disease is prevalent in Asians with most common occurrence in Japanese population, but case reports have been obtained from world wide (11). The disease usually has an acute onset with systemic manifestation along with tender lymphadenopathy. The etiology is unknown and many viral factors like Epstein Barr virus (EBV), Herpes simplex virus (HSV), HIV, dengue have been implicated (12). Establishing a diagnosis is important so as to rule out other common causes of lymphadenopathy like Tuberculosis, SLE, or other reactive causes and rarely Lymphomas (10). Cytology may sometimes give a clue to diagnosis if the cytospins yield abundant necrosis with intracellular and extra cellular apoptotic bodies and absence or presence of few granulocytes and typical histiocytes with a crescentic nucleus (3). Histopathology is the key to diagnosis as most of the other laboratory investigations are non specific. The histology shows a typical paracortical involvement of the lymphnode with necrosis and distinctive crescentic histiocytes, transformed lymphocytes and other polymorphous population of lymphocytes surrounding the necrotic debris. Paucity or absence of Neutrophils substantiates the diagnosis and rules out any acute suppurative lymphadenitis (4). Literature shows that the disease is known to be classified into three histopathological forms namely

Proliferative, Necrotic, Xanthomatous. Recognition of each of these forms is important so as to differentiate from other causes of lymphadenopathy. For example, in most cases of proliferative form, a misdiagnosis of a Lymphoma can be established (11). The necrotic form can sometimes be mistaken for SLE lymphadenitis. The course of the disease is self limiting with complete recovery in 1-4 months. In our case a young female was diagnosed with Kikuchi disease based on histopathology.

The patient responded to treatment with steroids and antipyretics. The systemic manifestations resolved in 5 days after the treatment. On regular follow up the lymphadenopathy resolved in three months.

Conclusion:

Kikuchi disease is a rare benign condition with systemic manifestations. Correct diagnosis with histopathology is important to differentiate the disease from other infectious and malignant causes of lymphadenopathy. Both the clinicians and the pathologists should be aware of the condition so as to avoid mistreatment of the patient.

Figure 1: Cytosmear showing features of chronic non specific lymphadenitis.

Figure 2: Hematoxylin and eosin stained section, scanner view showing areas of necrosis intervening the lymphoid cells.

Figure 3: Hematoxylin and eosin stained section at low power showing necrotic areas and polymorphous population of lymphoid cells.

Figure 4: High power showing Histiocytes, Lymphoid cells, apoptotic bodies, Absence of Neutrophils.

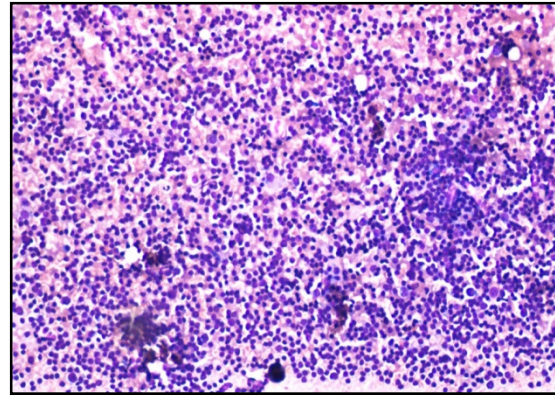


Figure 1

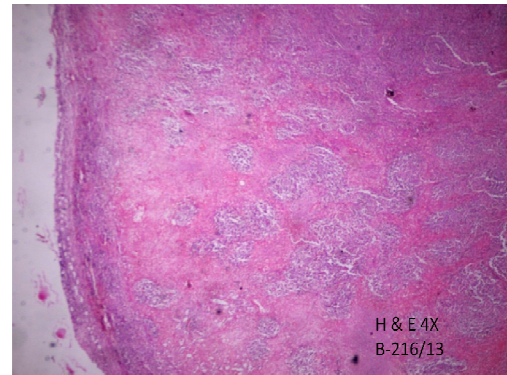


Figure 2

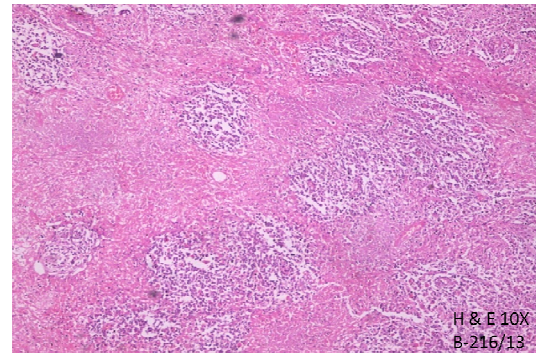


Figure 3

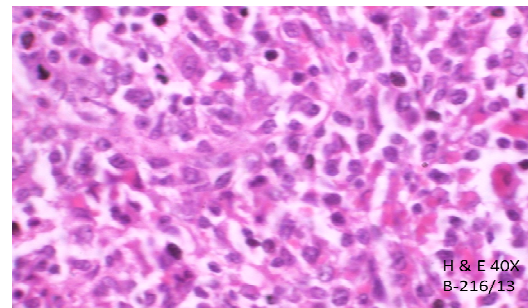


Figure 4

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