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KIKUCHI FUJIMOTO DISEASE – A CASE REPORT

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ABSTRACT Kikuchi Fujimoto Disease (KFD) is a rare disorder, it is a benign or non-cancerous/non-malignant of lymph nodes of young adults, to a great degree effecting young women. It is characterized by tissue abnormalities i.e., lymph node enlargement with inflammation and pain. The exact cause Kikuchi disease is unknown. There is no specific laboratory test that helps in contributing to find out the KFD diagnosis. KFD can be associated with systemic lupus erythematosus with similarities but can differentiate with help of differential diagnostic parameters. With supportive treatment the disease can be resolved within 4 months. In this study patient presented with fever since past 2 weeks and was treated with empirical antibiotics, antipyretics, antacids and other symptomatic supportive therapy. Patient is stable during discharge.

KEYWORDS : Lymph nodes, Kikuchi Fujimoto disease, Non-malignant, Benign, Lymphadenitis, Young women.

INTRODUCTION-

Kikuchi Fujimoto Disease [KFD], also termed as histiocytic necrotizing lymphadenitis or enlargement of lymph nodes in the body. It is a rare and self-limiting disorder that mostly effects young women. This disease is similar to cervical lymphadenopathyas the symptoms are similar [1, 2].

Epidemiology-

KFD is an extremely rare disease. It was found that Japanese Asian countries adult younger than 40 years of age are at high risk [5]. Later cases were emerging from West Germany and individual cases from Iran, Italy, South Korea, and Spain. Since then, it has been reported worldwide [5].

Clinical Features-

Kikuchi disease is characterized by cervical adenopathy [mainly posterior lymph nodes are affected], fever with or without chills, lymph nodes enlarged, tender and painful , an atypical lymphocytosis, myalgia's, weight loss, sweating, hepatosplenomegaly.Extra nodal involvement by KFD is seenuncommon, but cutaneous lesions associated with KFD are seen common[6]. Exact root of cause is not perfectly known; sometimes it may be associated with SLE, leukopenia's [3].

Cytomorphology of Kikuchi lymphadenitis-

- Necrotic debris, karyorrhexis
- Small phagocytic histiocytes with sharp angulated nuclei.
- Cytoplasmic tangible bodies
- Increased number of monocytes
- Absence of neutrophils.

Laboratory Diagnosis-

The is no specific laboratory diagnostic test available for KFD. Leukopenia can be witnessed in 30-70% cases. Other nonspecific findings like anaemia, atypical peripheral blood lymphocytes and increased erythrocyte sedimentation rate with low C-reactive protein values in maximum patients (30-50% of cases); serum hepatic transaminase activities and lactate dehydrogenase levels are also found to be increased [5] [9].

Diagnosis-

1. Diagnosis of KFD is generally done based on excisional biopsy of effected lymph nodes.
2. Fine-needle Aspiration Cytology is a cytologic diagnostic test which is less commonly used when compared to excisional biopsy of lymph nodes and accuracy was estimated at 56.3%.
3. Patients who have minor form of KFD symptoms are under diagnosed [5].

Differential Diagnosis-

Differential diagnosis is based on morphologic evaluation, immunohistochemical analysis has been used and it commonly rules out malignant lymphoma. The histiocytes of KFD characteristically express myeloperoxidase, in addition to lysozyme, CD68, and CD4 [7]. Plasmacytoid dendritic cells do not express any histiocytic markers or myeloperoxidase[8]. It also gives clear distinction of KFD during early stages of the disease (from large cells or high-grade lymphoma. CD8 expression is less common than CD4, and also a predominance expression of CD8 positivity is characteristic of KFD [5]. In the same way histologic findings of skin lesions of KFD patients can mimic cutaneous malignant T-cell lymphoma [5].

On histological findings, SLE and Kikuchi disease may be similar, but haematoxylin bodies are unique to SLE on histology compared to Kikuchi disease [8].

Clinical Course And Management-

KFD is a self-limited disease, which upto1 to 4 months. There's a low recurrence rate of 3% to 4% has been reported. Duration of this recurrence disease was found to be 8- to 9-years period. Very less fatal cases were found due to Kikuchi Fujimoto Disease [5].

There is no specific treatment available currently for KFD due to its unknown cause. Therefore, in general therapy is given for symptomatic relief, like relief of fever and lymph node tenderness by using analgesics and antipyretics. Corticosteroids are used in severe cases or relapsing disease [6]. Takada and co workers recently reported a case of KFD that resolved with oral Minocycline treatment, suggesting that the causative agent of KFD might be especially sensitive to this antibiotic [5]. Due to the disease's rarity, diagnosing individuals might be difficult because they may be mistreated for conditions with other causes. Thus, by being more knowledgeable about this uncommon illness, physicians and pathologists can contribute to improving patient outcomes [10].

Case Reoprt

A 27-Year-old-Female patient presented with complaints of high-grade fever since 2 weeks, on arrival temperature was 100.2 ^{0}F, H.R-100 bpm, SpO2- 98%, Blood pressure- 110/70 mm Hg.

Laboratory Data: Hb- 10.9 g/dl, plt-470 lakhs/cumm, WBC- 4.3 thousand/cumm, lymphocytes-24.2%, neutrophils-69.9%, eosinophils-0.9%, monocytes-5.1%, basophils-0.2%,Creatinine- 0.46 mg/dl, urea- 19mg/dl, Uric acid- 1.7 mg/dl, CRP- 36.4 mg/L, ESR- 49 mm/hr, SGOT- 52 U/L, SGPT- 59 U/L, ALP- 82 IU/L, serum cholesterol- 150mg/dl, total Bilirubin- 0.3 mg/dl, total Protein-5.9, albumin-2.4 g/dl, sodium-142 mmoles/L, potassium-4.2 mmoles/L, chloride-108 mmoles/L, magnesium-1.5mg/dl, pre-albumin-12

mg/dl, RBS-91 mg/dl, phosphorous-3.6mg/dl, amylase-84 CUE-N, culture blood and urine- negative[no growth reported], CECT Abdomen shown- mesenteric lymphadenopathy along right sided ileocolic, para-aortic group of lymph nodes. Laparoscopy + Lymph node biopsy under general anaesthesia-macroscopic description-mesenteric lymph node frozen measuring 10.5x0.4x0.3cms-showing reactive lymphoid hyperplasia with evidence of Kikuchi lymphadenitis. Immunohistochemistry results lymphoid cells predominantly CD3 positive with admixed CD20 positive cells. CD30 negative.

Microscopic description- sections of lymph nodes showing large areas of necrosis are seen with karyorrhectic debris and scarred histiocytes.

Patient differential diagnosis supports the diagnostic criteria of Kikuchi disease and was treated with empirical antibiotics, antipyretics, antacids, hydration and symptomatic treatment.

DISCUSSION

The patient came to Outpatient department with complaints of high-grade fever since past 2 weeks, was on medication since last 1 week T. cefixime 200mg Per oral twice a day, but fever not decreased, doctor advised admission for evaluation with evidence of high-grade fever, non-suppressive on antibiotic use. The patient's differential diagnosis satisfies criteria of Kikuchi's disease diagnostic criteria shows provisional diagnosis of Kikuchi Fujimoto disease [KFD].

CONCLUSION

The patient was spotted with Kikuchi Fujimoto disease with unknown cause. As it is self-limiting disorder, as the symptoms lasts for a week and is self-limited, use of antibiotics, steroids, antipyretics, antacids are instant symptomatic relief. Use of prophylactic treatment of antibiotics for longer duration of time can cause drug resistance.

Management of Kikuchi disease mainly focusses on inflammation or pain. Thus, patient was treated with hydrating fluids, antibiotic [ofloxacin], antacid, antipyretic, analgesic, steroid, along with multivitamin supplementation as it has no standard treatment guide lines.

Conflict Of Interest

None

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