Case Report

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Kikuchi Fujimoto Disease: A Rare Case Report

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Abstract

Introduction: Kikuchi-Fujimoto disease (histiocytic necrotizing lymphadenitis) is a rare, benign, self limiting syndrome characterized by tender regional lymphadenopathy, fever and night sweats. Highest prevalence is seen among Japaneese population. **Case Report:** We report a case of a young male presenting with fever with chills and tender cervical lympadenopathy not responding to the traditional treatment. A fine needle aspiration cytology of the lymph node was done, which was inconclusive. An excision biopsy showed features suggestive of Kikuchi Fujimoto's disease. Patient was started on low dose steroids with anti inflammatory drugs. Patient became symptom free within a week and has no recurrence in 6 months of follow up. **Discussion:** Kikuchi disease is mainly a diagnosis of exclusion. It is self-limited, and lasts 1 to 4 months with a low, but possible, recurrence rate of 3% to 4%. Only 3 fatal cases have been reported, occurring in the active phase of probably genuine disease. It has no specific treatment. Only symptomatic treatment measures to relieve local and systemic complaints should be used. Patients with a more severe clinical course or with relapsing signs and symptoms could benefit temporarily from corticosteroids. **Conclusion:** Kikuchi Fujimnoto disease is rare and a self limited disease, diagnosis of which is based on histopathological findings. It should be suspected in all the cases of unexplained fever and tender lymphadenopathy thus avoiding cumbersome investigations. It usually responds to symptomatic treatment and in unresponsive patients, low dose steroids for 10 days helps to prevent relapse and recurrence, as in our case.

Keywords: Kikuchi Fujimoto disease; Histiocytic necrotising lymphadenitis; Pyrexia; Lymphadenopathy; Corticosteroids.

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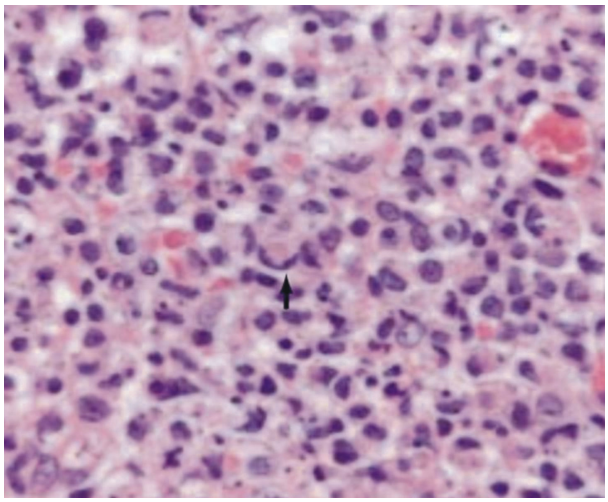
Introduction

Kikuchi-Fujimoto disease (KFD; so-called histiocytic necrotizing lymphadenitis) is an enigmatic, benign, and self limited syndrome characterized by regional lymphadenopathy with tenderness, usually accompanied by mild fever and night sweats.[1] It is a , benign, and self-limited syndrome. It is first reported almost simultaneously by Kikuchi and by Fujimoto and associates in 1972 as a lymphadenitis with focal proliferation of reticular cells accompanied by numerous histiocytes and extensive nuclear debris.[2] Disease has worldwide distribution with a higher prevalence among Japanese and other Asiatic people.[3,4] Most often seen in adults younger than 40 years. A female preponderance has been reported (female/male ratio, 4:1).[5,6] Recent reports from Eastern countries seem

Fig 1: Karyorrhectic Foci with Large Numbers of Histiocytes, Including Crescentic Histiocytes (Arrow), and Some Lymphoid Cells



to indicate that the female preponderance was overemphasized in the past and that the actual ratio is closer to 1:1. [5]

Case Report

A 35 yrs old male presented with complaints of fever with chills since 3 weeks. Fever is insidious onset, high grade, intermittent, with chills, reduced on medication, no diurnal variation. No other co relating history. On examination, axillary temperature was 103 ^{o}F. A single 2x2 cms, lymph node present in the right side of neck in the posterior triangle, firm in consistency, surface smooth. No signs of inflammation. Systemic and ENT examination is within normal limits. All the relavant laboratory investigations and infective parameters are within normal limits. Chest X-ray is normal. Patient was started on antibiotic and anti inflammatory and anti pyretic medications. No improvement was noted after 1 week. Hence, a FNAC of the lymph node was done. The aspirate was hemorrhagic and cytology showed hemorrhagic cells with clusters of polymorphic lymphoid cells comprising of centrocytes, centroblast, along with histiocytes and an ill defined granuloma. No caseating necrosis. An impression of Reactive lymphadenitis was given. An excision biopsy (Fig 1) of the cervical lymph node was done, which showed paracortical areas of expansion with karryorhectic debris. Focal areas show reactive follicles and is negative for granuloma or malignancy. A final diagnosis of Kikuchi Fujimotos disease was made. Patient was started on low dose prednisolone along with anti inflammatory drugs. Patient responded well and became symptom free in a week. Patient is on regular follow up for the last 6 months without any recurrence.

Discussion

The etiology is unknown. Viral or autoimmune cause is speculated. A viral infection is, nevertheless, possible by virtue of clinical manifestations, as described by Unger and colleagues.[7] Upper respiratory prodrome, atypical lymphocytosis, and lack of response to antibiotic therapy, and certain histopathologic features i.e. proliferation of immunoblasts, presence of necrotic zones localized to T-cell areas, expansion of the paracortex, and predominance of T cells. Histologic, ultrastructural, and immunohistochemical findings might support a hyperimmune reaction, perhaps to several organisms. It is possible that KFD might represent an exuberant T cell–mediated immune response in genetically susceptible people to a variety of nonspecific stimuli. DPA1*01 and DPB1*0202 alleles is significantly higher.[1] Electron microscopic studies have identified tubular reticular structures in the cytoplasm of stimulated lymphocytes and histiocytes.[2] The association between KFD and SLE has been reported with a frequency probably greater than that expected by chance. Imamura and coworkers hypothesized that KFD might reflect a self limited SLE-like autoimmune condition induced by virus infected transformed lymphocytes.[1] Some physicochemical factors have been pointed out anecdotally as triggers that might lead to KFD. A case of previous pacemaker implantation and simultaneous occurrence of KFD and