Kikuchi's Disease

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ABSTRACT

Kikuchi-Fujimoto disease is a self-limiting disease that usually presents with lymphadenopathy and fever. We present the case of a young girl who presented with complaints of cervical lymphadenopathy and fever. A lymph node biopsy was performed which was suggestive of Kikuchi disease. Our patient was treated symptomatically and did well. Awareness and knowledge of this disorder is essential for physicians and health care workers to ensure appropriate care and treatment of such cases.

Keywords: Kikuchi-Fujimoto disease; Cervical lymphadenopathy; Necrotizing lymphadenitis; Histiocytes

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INTRODUCTION

Kikuchi-Fujimoto disease is an idiopathic and uncommon condition characterized by tender lymphadenopathy. It usually affects cervical lymph nodes and is often accompanied by mild fever [1, 2]. The disease was initially discovered by Dr. Masahiro Kikuchi in Japan in 1972. In the same year, Fujimoto and colleagues defined and described Kikuchi's disease. Outside Japan, the first case of Kikuchi-Fujimoto disease was identified in 1982 [3]. The disease is more prevalent among females. Individuals younger than 30 years are more affected than older age groups [4]. The cause of Kikuchi-Fujimoto disease is not known; it has been proposed that the disease may have viral or post-viral etiology [5].

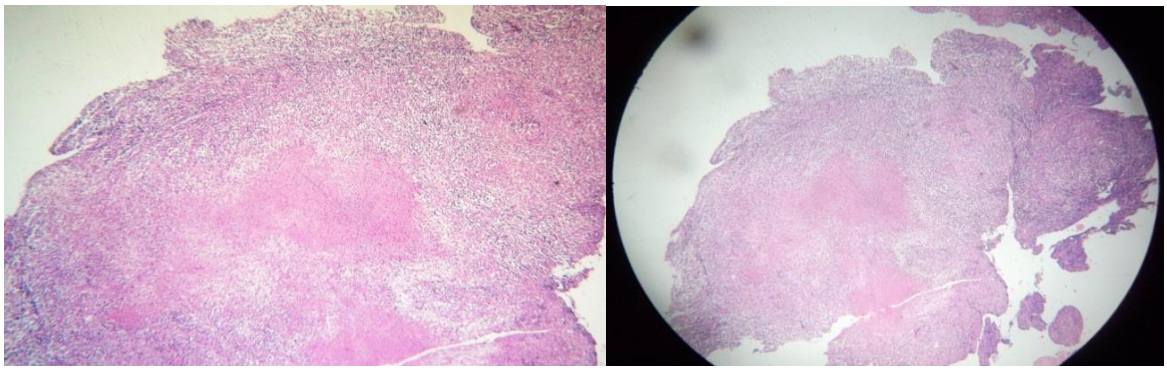
Kikuchi-Fujimoto disease is very rare condition. In Pakistan, very few cases of Kikuchi's disease have been reported and there is little knowledge of this disease among physicians and other medical professionals [6-8]. We present the case of Kikuchi-Fujimoto disease characterized by lymphadenopathy from a tertiary care hospital of Karachi, Pakistan.

CASE REPORT

An 18-year-old female presented to the outpatient clinic with complaints of bilateral swelling of her upper neck for the past year. Swelling had progressed over time and was associated with low-grade fever and weight loss. There were no other associated symptoms such as malaise, night sweats, fever, chills or cough. There was no known history of trauma or infection of ears, nose, throat or mouth.

On examination, she was thin and had conjunctival pallor. She had bilateral enlarged lymph nodes (mostly involving the posterior groups of cervical nodes), which were tender to touch, not matted and absent of any discharge. Mobility of the lymph nodes could not be assessed because of severe tenderness. Her full blood count was normal except for the erythrocyte sedimentation rate, which was slightly elevated at 30. Her chest radiograph was normal. A skin tuberculosis test (Mantoux test) was also negative. Excision biopsy of an enlarged and tender lymph node was performed. Histopathological examination showed fragmented lymphoid tissue with necrotic areas and mixed inflammatory infiltrate with early granuloma formation. There were epitheloid cells, histiocytes and occasional giant cells, as shown in **Figure 1**. This appearance strongly suggested necrotizing lymphadenitis, which was consistent with the clinical picture and consequently, the patient was diagnosed with Kikuchi-Fujimoto disease. Out patient was given symptomatic treatment to alleviate suffering and to promote comfort. She responded well to symptomatic treatment. She attended regular follow-up and has had no relapsing signs or symptoms for approximately 6 months.

Figure 1: Histopathology of a lymph node demonstrating distorted architecture with areas of necrosis



DISCUSSION

Kikuchi-Fujimoto disease is a benign clinical condition that is characterized by lymphadenopathy, lymph node tenderness, fever, malaise, night sweats, chills, cough, arthralgia, nausea, vomiting and weight loss. Chest and abdominal pain has also been reported in a few cases. In addition, the patient may also present with hepatosplenomegaly. It is a rare disease more prevalent among Japanese [8]. Kikuchi's disease is also prevalent in rest of the Asia, with very few cases being reported in the United States and Europe [9-11]. The disease mostly affects individuals under 30 years of age and females are affected more than males [4]. Kikuchi's disease is self-limiting and its symptoms resolve within a few weeks to months [5]. The onset of Kikuchi's disease is acute as it evolves within 2 to 3 weeks. Symptoms of the disease resemble tuberculosis and some head and neck infections, therefore, other diseases associated with cervical lymph node enlargement need to be evaluated [12]. Lymph node size may vary from 0.5 cm to 4 cm and mostly involves posterior cervical lymph nodes [13, 14]. Our patient also presented with lymphadenopathy, fever and weight loss, and screening for tuberculosis was negative.

The cause of Kikuchi's disease is unknown; viral agents including human immunodeficiency virus (HIV), Epstein Bar Virus (EBV), dengue virus, herpes simplex virus and parvovirus B19 have been reported as possible etiological factors [15]. Few reports suggest that there is an association with autoimmunity. On the basis of histopathological classification Kikuchi disease is broadly classified into three main types: proliferative, necrotizing and xanthomatous. Histiocytes including plasmacytoid monocytes are the predominant cells found on immunohistochemistry.CD8(+) T cells can be found in variable numbers but B cells are almost always absent. Activation of T lymphocytes and histiocytes are thought to play an integral role in pathogenesis of the disease. Routine investigations do not aid diagnosis of Kikuchi disease. ESR and C-reactive protein may be elevated in some patients and 25-30% of patients may have atypical lymphocytes on peripheral film [17]. None of the available radiological or laboratory findings can confirm the diagnosis of Kikuchi's disease. The diagnosis can only be confirmed via histological examination of the excised lymph nodes [18]. The current treatment of Kikuchi disease is symptomatic. Commonly non-steroidal anti-inflammatory drugs (NSAIDs) are prescribed to reduce fever and lymph node tenderness. In addition, corticosteroids can also be given in severe forms of the disease [20]. Some studies report successful use of intravenous immunoglobulins [21]. Kikuchi disease has a recurrence rate of 3-4% [5, 15]. Our patient was treated with NSAIDs for 5 days and corticosteroids (prednisolone) for 1 week. As symptoms starts to improve, corticosteroids were tapered and discontinued over a period of 2 weeks. She was followed up at the out-patient department for 6 months and no recurrence was observed.

CONCLUSION

Kikuchi-Fujimoto is a rare disease that should be considered a differential diagnosis when a young female patient presents with lymphadenopathy and fever. It may mimic other conditions such as lymphoma, tuberculosis or systemic lupus erythematosus, but histopathological examination plays a key role in diagnosis. Knowledge and awareness about disease among physicians, histopathologists and other medical professionals

is essential to ensure early recognition, correct diagnosis and appropriate treatment of the disease.

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