

## Kikuchi disease

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### Abstract

Kikuchi-Fujimoto Disease or Necrotizing Lymphadenitis is a rare, benign, self-limiting disease. It usually affects young females in the third decade of life. The most common presentation is cervical lymphadenopathy, though the etiology of the disease is still controversial. Clinical findings, histological diagnosis and immunohistochemistry help in diagnosis. Once diagnosed, steroids have been found to alleviate symptoms in patients with systemic manifestations. Antibiotics should not be prescribed until infective element is identified.

We report the case of a female patient who presented with tender cervical lymphadenopathy. She was diagnosed on excision biopsy of one of her lymph nodes. Anti-inflammatory drugs were started but the disease relapsed briefly after. Her symptoms have improved remarkably after initiation of steroid therapy, since her relapse.

**Keywords:** Kikuchi-Fujimoto Disease, Cervical lymphadenopathy, Lymph nodes.

### Introduction

Kikuchi-Fujimoto disease (KFD) is an enigmatic, benign and self-limited syndrome characterized by regional lymphadenopathy with tenderness, predominantly in the cervical region, usually accompanied by mild fever and night sweats.<sup>1</sup> Kikuchi's disease, or necrotizing histolytic lymphadenitis, is a rare disease that presents predominantly in young women in their 20s and 30s from the Far East.<sup>3</sup>

### Case Report

We report the case of a 23 year old female who presented in the Out Patient Department of Shalamar Hospital and Medical College Lahore, with a history of low grade fever accompanied by night sweats and malaise for one month. She also reported of tender neck swelling since one month. On examination, she was found to have enlarged

lymph nodes in the submandibular region and anterior cervical and posterior cervical lymph node chains, of 2x3 cm size. The rest of her physical examination was unremarkable. Her Abdominal Ultrasound was reported as normal, although blood complete examination revealed neutropenia. An excision biopsy of one of her posterior cervical lymph nodes was performed and sent for histopathology. Biopsy report revealed Kikuchi's Disease, following which non-steroidal anti-inflammatory drugs were started. Her symptoms improved and cervical lymph nodes regressed in size. But after 3 weeks she again reported in the OPD with the same enlargement of lymph nodes. At that time, Prednisolone therapy was initiated, to which she responded very well and her symptomatology improved. After 8 weeks, she became symptom-free. She is currently on regular follow up.

### Discussion

KFD is an extremely uncommon, self-limited, and perhaps under-diagnosed disorder of unknown cause, yet with an excellent prognosis.<sup>2</sup> The epidemiology of Kikuchi's disease is widespread, spanning the globe from Japan, where it was first described in 1972, and Asiatic people.<sup>3</sup> Kikuchi's disease is an idiopathic illness characterized by a self-limiting lymphadenitis that normally resolves over subsequent weeks or months without specific treatment.<sup>4</sup> There is much speculation about the etiology of Kikuchi's disease, with both infectious and autoimmune causes having been suggested.<sup>5</sup> It often presents as painful cervical lymphadenopathy in young females. Unilateral and posterior cervical lymph nodes are the commonest to be involved. Less common manifestations are in the form of axillary and mesenteric lymphadenopathy, splenomegaly, parotid gland enlargement, arthralgias, myalgia, aseptic meningitis, bone marrow haemophagocytosis and interstitial lung disease. The cutaneous lesions include erythematous macules, papules, plaques and nodules.<sup>6</sup> Routine laboratory investigations usually do not aid in the diagnosis except for erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) which might be elevated in some patients. Many patients have a low white blood count atypical peripheral blood lymphocytes. Fine-needle aspiration cytology (FNAC) only has a limited role. Diagnosis is based on histopathological findings of a lymph node biopsy.

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Morphologically, it is predominantly histiocytes and plasmacytoid monocytes but also immunoblasts and small and large lymphocytes. The immunophenotype of Kikuchi's disease is primarily composed of mature CD8-positive and CD4-positive T lymphocytes.<sup>7</sup> Although uncommon, the diagnosis of KFD should be considered in a differential diagnosis that includes tuberculosis, connective tissue diseases such as SLE, or lymph proliferative disorders and immunological diseases.<sup>8</sup>

Kikuchi-Fujimoto disease is typically self-limiting, resolving within 1 to 4 months, though a possible recurrence rate of 3 to 4% has been reported. Analgesics, antipyretics and non-steroidal anti-inflammatory drugs may be used to alleviate lymph node tenderness and fever. The use of corticosteroids has been recommended in severe extra nodal or generalized KFD but is of uncertain efficacy.<sup>9</sup> In case of severe and persistent symptoms in addition to high doses of glucocorticoids, intravenous immunoglobulins should be prescribed.<sup>10</sup> In our case prednisolone was given for relief of symptoms after her relapse.

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