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## Case Report

# Kikuchi Disease with enlargement of intramammary lymph node<sup>☆</sup>

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## ABSTRACT

We describe a rare case of intramammary lymphadenopathy due to Kikuchi-Fujimoto disease. A 15-year old female presented to the Breast Clinic with complaints of a tender, palpable right breast lump. An ultrasound of the area of concern demonstrated an enlarged 2.9 cm intramammary lymph node with preservation of the fatty hilum. An ultrasound guided core biopsy of the lymph node confirmed the diagnosis of Kikuchi-Fujimoto disease.

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## Introduction

Kikuchi-Fujimoto disease (KFD), also known as histiocytic necrotizing lymphadenitis, is a rare cause of lymphadenopathy that most commonly involves the cervical chain. It is a self-limiting disorder with the most common clinical features being fever and cervical lymphadenopathy. In rare instances involvement of the retroperitoneal lymph nodes has been noted. The exact pathogenesis remains unknown although multiple viruses have been associated with the disease. We describe a case of KFD presenting as a palpable intramammary lymph node.

## Case

A 15-year-old female presented to the Breast Clinic with complaints of a tender, palpable right breast lump. The patient first noticed the lump four months prior to presentation. She denied having any constitutional symptoms including night sweats, fever, and cough. The patient denied having any recent vaccinations, animal bites, or trauma.

A diagnostic ultrasound of the area of palpable concern was performed and revealed an enlarged lymph node measuring 2.9 cm on the right, 9:00 position 8 cm from the nipple. There was cortical thickening measuring up to 7.5 mm, the fatty hilum and oval shape of the lymph node was maintained (Fig. 1A). A large node within the right axillary tail was also noted (Fig. 1B). Other lymph nodes in the right and left axilla were not enlarged.

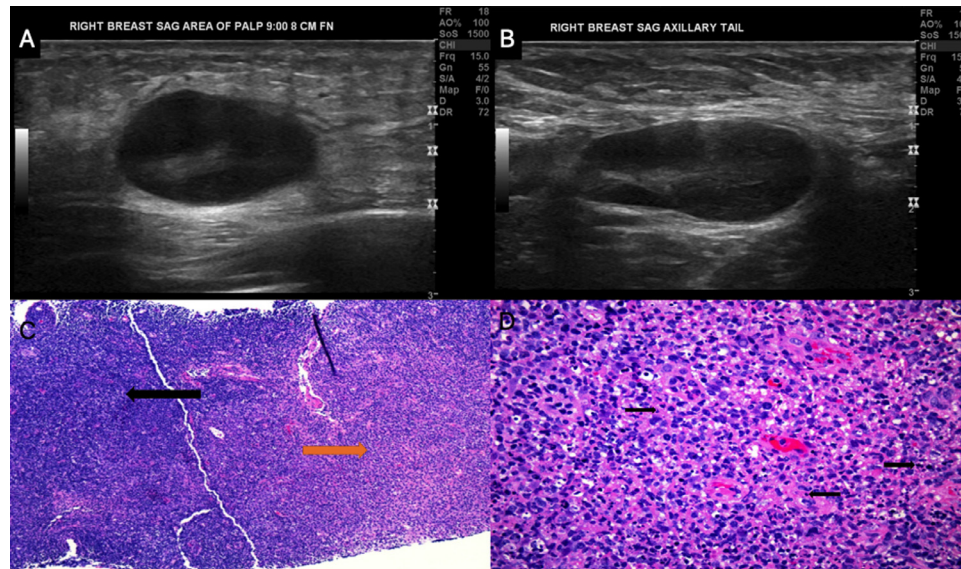
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**Fig. 1** – An 15 year-old with palpable mass right breast at 9:00. (A) Ultrasound image of palpable mass within the right breast demonstrates an enlarged lymph node at the 9:00 position, 8 cm from the nipple. The node measuring 2.9cm has a preserved oval shape, fatty hilum and thickened cortex. (B) Ultrasound image of right axilla demonstrates an enlarged node with thickened cortex. Low power hematoxylin and eosin (H & E) stain (C) demonstrates reactive lymph nodes (black arrow) showing lympho-histiocytic proliferation with karyorrhectic and apoptotic debris (orange arrow). High power image (D) shows detail of karyorrhexis and apoptotic debris (black arrows) consistent with Kikuchi-Fujimoto Disease.

An ultrasound-guided core biopsy of the intramammary node was performed. Pathology showed reactive lymph nodes with lympho-histiocytic proliferation with karyorrhectic and apoptotic debris features consistent with KFD. (Fig. 1C and D). Flow cytometry did not identify any clonal lymphocytic lineage.

## Discussion

KFD, also known as histiocytic necrotizing lymphadenitis, is a rare cause of lymphadenopathy that most commonly involves the cervical chain. Although initially described in Asian women, further studies have demonstrated that KFD affects all races and ethnicities [1].

Lymphadenopathy in KFD is typically accompanied by a low-grade fever in approximately 35% of patients [1]. Additional symptoms can include rash (10%), myalgias (7%), and fatigue (7%) [2, 3]. Laboratory findings are usually within normal limits; however, cases of leukopenia, elevated erythrocyte sedimentation rate, and abnormal liver function tests have been reported [1,2,4].

Lymphadenopathy seen with KFD typically does not exceed 2.5 cm on short axis measurement. Although KFD classically involves the cervical chain; retroperitoneal, mediastinal, and axillary nodal involvement have also been reported [5,6]. To our knowledge only two reports describe involvement of intramammary nodes, as in our patient [7,8].

The pathogenesis of KFD remains unknown. However, multiple viral illnesses, including Epstein Barr Virus and Cytomegalovirus, have been associated with the disease [5,9].

Notably, there are reports of patients developing systemic lupus erythematosus following resolution of KFD [5]. Due to this association, close follow-up is warranted following a diagnosis of KFD.

There is no specific treatment for Kikuchi disease as it is self-limiting with resolution typically seen within 1-4 months of disease onset. Supportive care with anti-pyretics is the mainstay of treatment.

In adult women, the presence of unilateral axillary adenopathy raises the question of occult breast carcinoma; however, breast cancer in women younger than 20 years is rare, with an incidence of 1 in 1 million patients [10]. Differential diagnosis for unilateral axillary adenopathy in a teenager includes reactive enlargement (post-vaccination), hidradenitis suppurativa, hematologic malignancy (Hodgkin's Disease), ipsilateral arm or hand infections including cat scratch fever (caused by the bacterium *Bartonella henselae*), *Mycobacteria tuberculosis* and collagen vascular diseases, including juvenile rheumatoid arthritis.

## Conclusion

We present a rare case of KFD presenting as a palpable right breast intramammary node in a young female with a solitary enlarged right axillary node. Diagnosis was confirmed on ultrasound-guided core biopsy with characteristic pathology. Close monitoring is warranted since development of SLE has been reported in these patients.

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## Patient Consent

Formal consents are not required for the use of entirely anonymized images from which the individual cannot be identified- for example, x-rays, ultrasound images, pathology slides or laparoscopic images, provided that these do not contain any identifying marks and are not accompanied by text that might identify the individual concerned. Therefore consent was not obtained for our case report.

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