



**KIKUCHI DISEASE – A RARE CASE REPORT**

<sup>1</sup>Dr. S. Parthasarathy MD DA DNB PhD FICA, <sup>2</sup>Dr. S Ravi MBBS DLO

<sup>1</sup>Associate Professor, Department of Anaesthesiology and Critical Care, Mahatma Gandhi Medical College and Research Institute, Sri Balaji Vidyapeeth University, Pillayarkuppam, Puducherry – 607402. India.

<sup>2</sup>Consultant ENT Surgeon, Shree ENT Care, Kumbakonam, Tamilnadu – 612001.

**\*Author for Correspondence: Dr. S. Parthasarathy MD DA DNB PhD FICA**

Associate Professor, Department of Anaesthesiology and Critical Care, Mahatma Gandhi Medical College and Research Institute, Sri Balaji Vidyapeeth University, Pillayarkuppam, Puducherry – 607402. India.

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**ABSTRACT**

A 24 year old female presented to us with a history of painless swelling in the neck of 2 weeks duration. After a five day course of antibiotics and anti-inflammatory drugs which proved futile in clearing signs, an excision biopsy was done. This was pathognomonic of kikuchi disease. All drugs were discontinued. The swelling disappeared in four weeks thereafter. There was no other visceral involvement. A six month follow up was uneventful. This case is reported for its rarity.

**KEYWORDS:** kikuchi disease, cervical lymph node, lymphadenopathy.

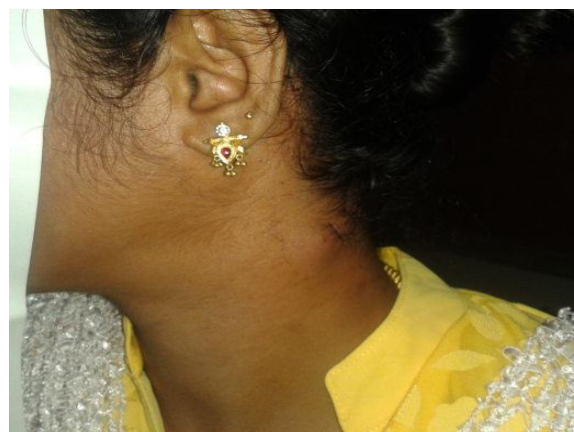
**INTRODUCTION**

Kikuchi disease, also known as Kikuchi-Fujimoto disease, is an uncommon, idiopathic, generally self-limited cause of lymphadenopathy predominantly posterior cervical lymph nodes. Multiple lymph node involvement, extra nodal features like hepatosplenomegaly, meningitis and arthritis may be present<sup>1</sup>. In the following article, we present a case of successful management of kikuchi's disease without extra nodal involvement.

**CASE CAPSULE**

A 24 year old female presented to us with five days fever followed by swelling of neck for two weeks. Headache with mild nausea, vomiting and fatigue were present. There was no history of weight loss, arthralgias, night sweats or thoracic/abdominal pain. There was no significant past history. On examination, the patient was mildly febrile, with two enlarged posterior cervical nodes. (Fig.1) There was no pain and the nodes were non tender. A significant search of the involvement of other sites including skin, liver, eye was not informative.

A five day course of a combination of cefixime and azithromycin with aceclofenac was administered. There was no response. An excision biopsy was done under local anesthesia which showed necrosis, with typical cell types, namely crescentic histiocytes and plasmacytoid monocytes. This was classical of kikuchi's disease. All drugs were stopped. After two weeks, the swelling decreased in size to disappear spontaneously. After a six month follow up, there were no symptoms and signs.



**DISCUSSION**

Kikuchi's disease, also known as histiocytic necrotizing lymphadenitis or Kikuchi-Fujimoto disease, is an uncommon, idiopathic, usually self-limited cause of lymphadenitis. Kikuchi first described this disease in 1972 in Japan. Fujimoto and colleagues independently described Kikuchi's disease in the same year<sup>2</sup>. Even though the disease is reported to be very rare the exact incidence is not yet known. To date, most cases have been reported from East Asia, even though there are definite case reports in UK and USA<sup>3</sup>. The cause of Kikuchi disease is not known, although infectious and autoimmune etiologies have been proposed. The most favoured theory proposes that it results when one or more unidentified agents trigger a self-limited autoimmune reaction. Lymphadenitis results from apoptotic cell death initiated by cytotoxic T lymphocytes. Some human leukocyte antigen (HLA) class II genes are more frequent in such patients

suggesting a genetic predisposition to the proposed autoimmune response<sup>4</sup>. Usually the disease is more common in young females, which is present in our case. It starts with a prodromal feverish illness with the lymphadenopathy usually cervical nodes being commonly involved. In our case, the posterior cervical nodes are involved which favours the rare diagnosis. There were neither other palpable lymph nodes nor any other systemic involvement. Arthritis, hepato splenomegaly and skin lesions<sup>5</sup> were notably absent in our case. The described incidence of such associated pathology ranges from 5–40 %. Routine investigations were normal and inconclusive in our case. Systemic lupus erythematosus is a differential diagnosis.<sup>6</sup> We did not resort to costly diagnostic tests like antinuclear antibodies (ANA), rheumatoid factor (RF), and lupus erythematosus (LE) preparations as they are not usually definitely conclusive. Histopathological examination (HPE) of the diseased tissue is the test of choice and we resorted to the same earlier to have a definitive diagnosis. We started on analgesics with antibiotics with infection in the back of mind but stopped the same with HPE report. We did not start the patient on steroids as they are indicated either in extra nodal involvement or in patients with multisite nodal involvement<sup>7</sup>. The limitation of the report is that we did not investigate for extranodal involvement and the findings were based on clinical examination only.

## CONCLUSION

We conclude that Kikuchi disease is a rare yet an important clinical diagnosis in cases with cervical lymphadenopathy of short duration. The diagnosis should be supported by histopathological examination. The disease is self-limiting in majority of the cases. Such patients need a long time follow up for the development of systemic lupus erythematosus later.

## REFERENCES

1. Louis N, Hanley M, Davidson NM, Kikuchi-Fujimoto disease: a report of two cases and an overview *The Journal of Laryngology Otolaryngology*, 1994; 108:1001-04.
2. Xavier Bosch and Antonio Guilabert. Kikuchi-Fujimoto disease - a review. *Orphanet Journal of Rare Diseases*., 2006; 1:18.
3. Vikrant Veer, Albert Lim, and Wolfgang Issing. Kikuchi-Fujimoto Disease: A Case Report and literature review *Case Reports in Otolaryngology* 2012(1): 1-5.
4. Tanaka T, Ohmori M, Yasunaga S, Ohshima K, Kikuchi M, Sasazuki T. DNA typing of HLA class II genes (HLA-DR, -DQ and -DP) in Japanese patients with histiocytic necrotizing lymphadenitis (Kikuchi's disease). *Tissue Antigens*., 1999; 54(3): 246-53.
5. Babu N Chaitanya and CS Sindura. Kikuchi's disease. *J Oral Maxillofac Pathol.*, 2010; 14(1): 6–9.
6. Anupam Patra, Sujit Kumar Bhattacharya, .SLE developing in a follow up patient of Kikuchi's disease: a rare disease. *Journal of Clinical and Diagnostic Research*., 2013; 4: 752 – 753.
7. Jang YJ, Park KH, Seok HJ. Management of Kikuchi's disease using glucocorticoid. *J Laryngol Otol.*, 2000 Sep. 114(9): 709-11.