

## Kikuchi's disease presenting as multiple cervical lymphadenopathy

Waipoj Chanvimalueng\*

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*Kikuchi's disease (histiocytic necrotizing lymphadenitis) is an idiopathic disease. Its manifestations include enlargement of the cervical lymph nodes, occasionally with fever, and often associated with other non-specific clinical signs and blood chemistry. The diagnosis relies on the histological examination of lymph node biopsies. A spontaneous resolution of the disease occurs spontaneously within three months. The author describes the diagnosis and treatment of Kikuchi's disease in three patients. All patients were given oral form of prednisolone for one or two weeks and benefited significantly resolution within three to five weeks in two cases and concluded that Kikuchi's disease should be considered in the differential diagnosis of cervical lymphadenopathy.*

**Keywords :** *Kikuchi's disease, Lymphadenopathy.*

Reprint request: Chanvimalueng W. Department of Otolaryngology, Faculty of Medicine, Thammasat University, Bangkok 12120, Thailand.

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**ไวพจน์ จันทรวินเมลิอง. โรค Kikuchi มาด้วยอาการต่อมน้ำเหลืองบริเวณคอโตผิดปกติ.  
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*Kikuchi's disease (Histiocytic necrotizing lymphadenitis)* คือโรคซึ่งมาด้วยอาการก้อนที่คอโต ตรงตำแหน่งบริเวณต่อมน้ำเหลืองด้านข้างของคอ โดยไม่ทราบสาเหตุ และบางครั้งผู้ป่วยอาจจะมีไข้หรือความผิดปกติของสารเคมีในเลือดร่วมด้วยการวินิจฉัย โดยวิธีการตัดชิ้นเนื้อบริเวณต่อมน้ำเหลืองเพื่อส่งตรวจดูลักษณะทางพยาธิวิทยา และการดำเนินโรค ซึ่งโรค Kikuchi นี้ส่วนใหญ่จะสามารถหายได้เองภายใน 3 เดือน ผู้เขียนได้บรรยายถึงการวินิจฉัยและการรักษาโรค Kikuchi จำนวน 3 รายโดยทั้ง 3 รายได้รับยา กลุ่ม oral steroid ประมาณ 1-2 สัปดาห์ และมีผู้ป่วย 2 รายจาก 3 รายหายภายใน 3-5 สัปดาห์ และ โรค Kikuchi ควรพิจารณาไว้ใน การวินิจฉัยแยกโรค ในผู้ป่วยที่มีต่อมน้ำเหลืองมีคอโตร่วมด้วย

**คำสำคัญ :** โรค Kikuchi, ต่อมน้ำเหลืองโต.

Kikuchi's disease is a histiocytic necrotizing lymphadenitis and self-limited disease of unknown origin.<sup>(1-4)</sup> Its first description was by Kikuchi and Fujimoto in 1972, in Japan.<sup>(5)</sup> It is a benign disorder predominantly of young females who present with cervical lymphadenopathy, and is commonly misdiagnosed as malignant lymphoma.<sup>(6)</sup> Kikuchi's disease might be associated with systemic lupus erythematosus.<sup>(7)</sup> It predominantly in patient before the fourth decade of life.<sup>(8)</sup>

An ENT specialist is often confronted with an inquiry regarding the cause of lymph node enlargements in the neck. Common causes include inflammation, neoplasm, or autoimmune disease.

The author report three cases of Kikuchi's disease as a benign self-limiting lymphadenopathy of unknown etiology, which is usually found in young female between 20 and 30 years of age. Its main symptoms are indolent or light tender, enlarged lymph nodes in the neck area. The correct diagnosis requires the histologic examination of the lymph node. The three female patients suffering from Kikuchi's disease were

treated at the Department of Otolaryngology between August 2002 and July 2004.

## Case Report

### First case

A 24-year-old Thai female presented at the Out-patient Department of Thammasat University Hospital with one month history of bilateral multiple cervical lymphadenopathy. She had a neck mass in the left and three in the right with firm consistency and tenderness on both side with low-grade fever.

Physical examination revealed a firm and fixed left upper jugular lymph node measuring 3X5 cm in diameter (Figure 1), and a right upper jugular lymph node measuring 2 cm in diameter with two lower jugular lymph nodes measuring 1x1.5 cm<sup>2</sup> in diameter (Figure 2). The nasal cavity and nasopharynx were within normal limits. Indirect laryngoscopic examination showed a normal epiglottis, vocal cord and pyriform recess. Otoscopic examination showed normal tympanic membranes. There were no cranial nerve deficits.



**Figure 1.** Left upper jugular lymph node measuring 3 x 5 cm<sup>2</sup> in diameter.



**Figure 2.** A right upper jugular lymph node measuring 2 cm in diameter with two lower jugular lymph nodes measuring 1 x 1.5 cm<sup>2</sup> in diameter.

Fine needle aspiration biopsy (FNA) of the neck mass was performed and was negative for malignancy. Routine laboratory examinations, chest x-ray and liver function test findings were all normal.

Excision biopsy of the right neck node was performed on August 21, 2002. The pathologic report: sections of lymph node show fragments of lymphoid tissue. Lymphoid follicles are enlarged. There are nodules of histiocytic cell hyperplasia with necrosis. Stain for acid fast bacilli is negative, and pathologic diagnosis is histiocytic necrotizing lymphadenitis (Kikuchi's Disease).

The patient was treated by an oral form of prednisolone for two weeks and follow up in three weeks. The patient had a complete remission.

### Second case

A 25-year old Thai female presented with

only one week history of left multiple cervical lymphadenopathy. She had two masses on her left neck, firm and tender.

Physical examination revealed two firm and fixed left upper jugular lymph nodes measuring 1.5 and 2 cm in diameter. The ENT findings were within normal limits. The routine laboratory examinations, chest x-ray and blood chemistry findings were normal. Panendoscopic examinations were performed in this case and showed normal findings. An excision biopsy was performed on September 4, 2002. The pathologic report was consistent with histiocytic cell hyperplasia with necrosis, and pathologic diagnosis was histiocytic necrotizing lymphadenitis of left neck node (Kikuchi's disease).

The patient was treated by oral form of prednisolone for one week and follow up in two weeks. The patient had a complete remission.

**Table 1.** Differential diagnosis of neck mass.

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| <b>1. Neoplasms</b>  |   |
| <b>A. Benign</b>   | <b>B. Malignant</b>   |
| 1. Vascular – hemangioma,<br>lymphangioma, arteriovenous<br>malformation, aneurysm | 1. Neck primary   |
| 2. Chemodectoma  | A. Lymphoma   |
| 3. Neural – neurofibroma, Schwannoma   | B. Sarcoma  |
| 4. Lipoma  | C. Thyroid carcinoma  |
| 5. Fibroma   | D. Salivary gland carcinoma   |
| 6. Miscellaneous – fibromatosis  | E. Branchial cleft cyst carcinomas  |
|  | F. Thyroglossal duct cyst carcinomas  |
|  | <b>2. Metastatic</b>  |
|  | A. Head and neck primary-mucosal<br>surfaces, skin, salivary glands, thyroid  |
|  | B. Infraclavicular primary-lung, kidney,<br>prostate, gonads, stomach, breast |
|  | C. Leukemia   |
| <b>2. Infection</b>  | A. Abscess  |
|  | B. Cervical lymphadenitis   |
|  | 1. Bacterial  |
|  | 2. Granulomatous – tuberculosis,<br>actinomycosis, sarcoidosis                |
|  | 3. Viral – infectious mononucleosis   |
| <b>3. Congenital</b>   | A. Thyroglossal duct cyst   |
|  | B. Branchial cleft cyst   |
|  | C. Dermoid cyst   |
|  | D. Teratoma   |
| <b>4. Miscellaneous</b>  | A. Zenker's (hypopharyngeal) diverticulum                                     |
|  | B. External laryngocele   |
|  | C. Amyloidosis  |
|  | D. Neuroma (traumatic)  |
| <b>5. Normal structures</b>  | A. Hyoid  |
|  | B. Carotid bulb   |
|  | C. Transverse process of vertebrae  |
|  | D. Normal neck nodes (hyperplasia)  |

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### Third case

A 24-year-old Thai female presented with three months history of bilateral multiple cervical lymphadenopathy with firm consistency and tenderness on both side.

Physical examination revealed two upper jugular lymph nodes measured 1.5-2 cm in diameter on both sides of the neck. ENT examinations were within normal limits. The routine laboratory examinations, chest x-ray and blood chemistry finding were normal. Panendoscopic examination and nasopharynx biopsy were performed and the results were negative. Finally, a lymph node biopsy was performed and the pathologic diagnosis was histiocytic necrotizing lymphadenitis (Kikuchi's disease) in July 2004.

The patient was treated with oral form of prednisolone for two weeks and follow up in three months, the patient still had small bilateral neck nodes, but they were not tender and showed no progression of the disease.

### Discussion

Histiocytic necrotizing lymphadenitis, or Kikuchi's disease, is a newly recognized disease of unknown origin that causes cervical lymphadenitis.<sup>(9)</sup> Cases of Kikuchi's disease are being reported in Asia with increasing frequency but relatively rare in other regions of the world. The symptoms of Kikuchi's disease include tender cervical lymphadenopathy with or without fever. Laboratory tests are often normal. The diagnosis is established on the basis of histologic of lymph node excisional biopsy. It is likely to be misinterpreted as malignant lymphoma or systemic lupus erythematosus.<sup>(10)</sup> Corticosteroid<sup>(11)</sup> was

administered for one or two weeks, which resulted in complete remission in two patients and one patient showed improvement and regression of the enlarged lymph nodes. Clinical and epidemiologic information about cervical lymphadenitis can often lead to a presumptive diagnosis.

Therefore, it must be included in the differential diagnosis<sup>(12)</sup> in all patients who present with a neck mass (Table 1). The outline developed by Suen JY, Wetmore JY (1981) should be used.<sup>(13)</sup>

However, Kikuchi's disease presenting as multiple cervical lymphadenopathy (a neck mass) should be included in the differential diagnosis of neck mass.

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