

## KIKUCHIS-FUJIMOTO DISEASE

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### ABSTRACT

*Kikuchi–Fujimoto disease or Sub-acute necrotizing lymphadenitis is a benign self-limiting condition. A case in a 24-year-old young female who presented with swelling in the right submandibular region and pain is reported. The erythrocyte sedimentation rate was elevated to 50 mm/h. Other serological tests for rubella, hepatitis B, hepatitis C, HIV, and toxoplasmosis, as well as tests for tuberculosis were performed which were negative. The excisional biopsy of the lesion showed necrotizing lymphadenitis consistent with Kikuchi–Fujimoto disease. No further treatment was given, and a full recovery was achieved within one month. Tuberculous lymphadenitis and lymphoproliferative disease were considered in differential diagnosis.*

**Key words:** Khyber College of Dentistry, Kikuchi's–Fujimoto disease, Oral and Maxillofacial Surgery

### INTRODUCTION

In 1972, Kikuchi and Fujimoto were the two Japanese pathologist who for the first time described the clinical features of a self-limiting syndrome of benign necrotizing lymphadenitis. The characteristic histologic appearance of Kikuchi's disease resembles that of malignant lymphoma.<sup>1</sup> KFD patients typically present with painless but sometimes tender, enlarged unilateral cervical lymph nodes. Two or more than two chains of the cervical lymph nodes may be involved. Involvement of other sites have also been noted as, splenomegaly along with generalized lymphadenopathy. The clinical indicators usually suggest a hematological malignancy, particularly a lymphoproliferative disease.<sup>2</sup> A case report along with clinical manifestations, histology, and outcome of KFD is presented.

The condition is common in Japan, China and Taiwan but not in Pakistan.

### CASE REPORT

A 24 years old young female patient reported to oral and maxillofacial surgery department Khyber College of dentistry Peshawar with a swelling in the sub-

mandibular area for the last 3 weeks. The patient was extremely concerned about the swelling and according to her she had a similar swelling below the chin one year back which regressed spontaneously. The swelling was not responding to medications. A detailed intra-oral and extra-oral examination was performed. On intra oral examination she had fair oral hygiene and all dentitions in good state of health. Extra-oral examination revealed a swelling with diffused borders not attached to the underlying structures, the overlying skin was found normal (Fig 1) and the lesion was slightly tender on palpation. On routine investigations the ESR of the patient was found raised and the rest of routine blood tests were normal. A Fine Needle Aspiration Cytology (FNAC) was decided to know the nature of the lesion. The FNAC report showed numerous small lymphocytes with focal aggregates specially seen in small clotted area, along with mixed large lymphocytes and occasional plasma cells and no granulomas were seen. The overall picture favored a reactive inflammatory process and the histopathologist suggested an open scalpel biopsy because of the raised ESR and the lesion not responding to antibiotics for quite some time to rule out chronic granulomatous inflammation or lymphoproliferative disorder.

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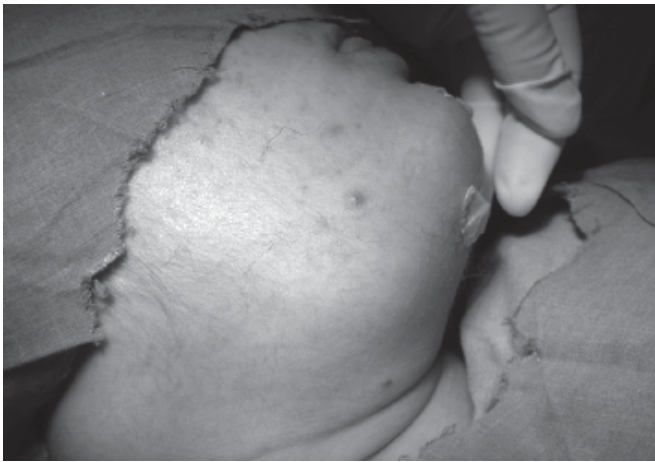


Fig. 1: Swelling in the right submandibular region

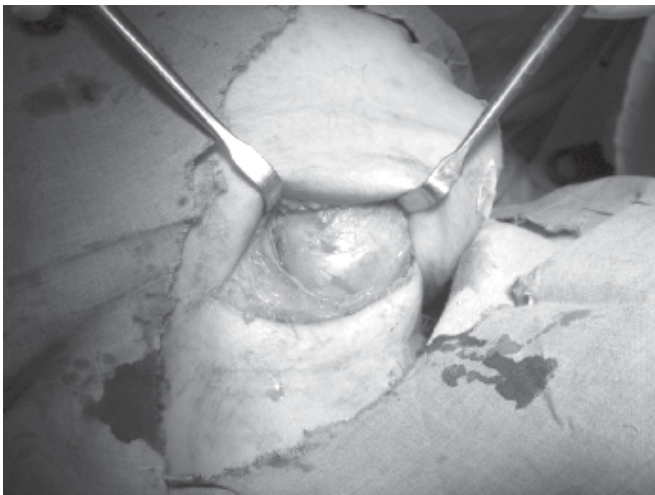


Fig 2: Exposure of the lesion in the subplatysmal plane

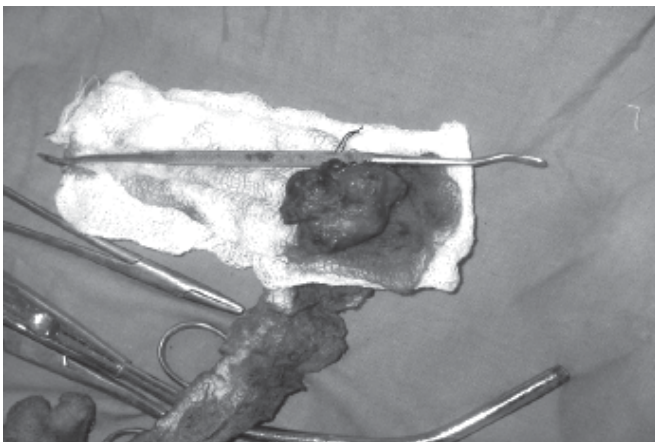


Fig 3: Excised lymph nodes

The patient was scheduled for an excisional biopsy, a 6 cm incision was given in the skin crease well below in the neck crease for better cosmetic results and to save the marginal mandibular branch of the facial nerve. The lesion was exposed and dissected from the under-

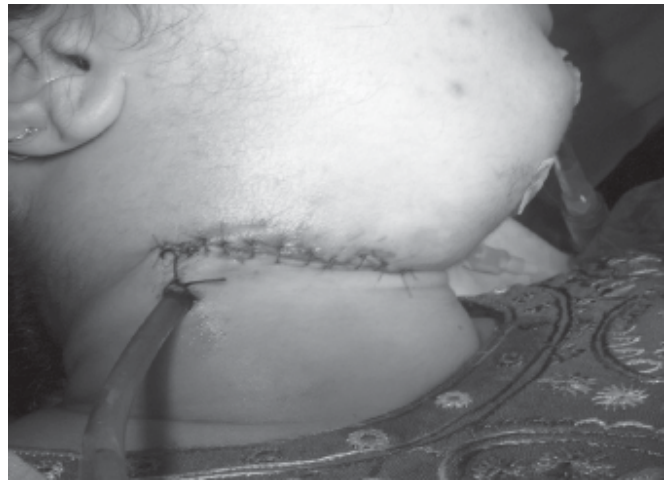


Fig 4: Stitched incision with suction drain in place

lying submandibular gland and freed from the superficial layer of deep cervical facial, (Fig 2) it was a bunch of lymph node (Fig 3) that has raised the suspicion of Tuberculous lymphadenitis. The lesion was removed, the incision closed in layers and a suction drain placed at the surgical site. (Fig 4) The biopsy report came as idiopathic necrotizing lymphadenitis (Kikochi's disease)

## DISCUSSION

KD or necrotizing histiocytic lymphadenitis is more common among Asians than the Western populations. However, an increasing number of cases are being reported from Europe and from the United States. Rare, but fair number of cases are common in Asia and Taiwan, for instance, 5.7% of patients undergoing lymph node biopsy have been diagnosed as KD.<sup>3</sup> KD is found to be more common in young women under the age of 30 years although it may affect the male gender as well. The characteristic feature of KD is painless but sometimes tender unilateral cervical lymphadenopathy. Occasionally, it may also present as hepato-splenomegaly<sup>4</sup> but in our patient the generalized lymphadenopathy and splenomegaly was absent. Some case reports show parotidomegaly and thyroiditis with kikuchi disease.<sup>5</sup>

One third to one half of patients have fever at the time of presentation. Other symptoms of KD are fatigue, malaise; headache, vomiting, night sweats, and weight loss.<sup>6</sup> Kikuchi's disease have also got cutaneous manifestations. Pigmented purpuric dermatitis, Lupus erythematosus, Erythema annulare centrifugum,

Erythema multiforme, Erythema nodosum, Kimura's disease, Lupus erythematosus, Sweet's and Behcet's disease are a few cutaneous manifestations of the disease<sup>7</sup> but none of them were associated with our patient.

The etiology of KFD has not been explained yet. Infectious agents (Epstein–Barr virus, herpes virus 6 and 8, toxoplasma, yersinia, brucella, human immunodeficiency virus, human T-cell lymphotropic virus type 1) and genetic factors (human leukocyte antigen class-2) are associated with the etiology of KD. The histological findings of KFD are not pathognomonic. Currently, there is no specific laboratory marker for KFD and must be excluded at the time of diagnosis. Diseases that can mimic histological features of KFD include *Yersinia*, *Toxoplasma*, and *Bartonella* infections, infectious mononucleosis, acquired immune deficiency syndrome, Hodgkin's lymphoma, and lymph node infarction.<sup>8</sup>

There are no predisposing factors for KFD, although Pileri and coworkers have described 40% of patients as having had tonsillectomies. Certain studies have shown that 32% had tonsillitis, otitis media, or tooth extraction before the emergence of KFD. It is suggested pathogen of, an ear, nose, or throat may be related to KFD, yet no direct evidence exists.<sup>9</sup>

Laboratory tests may show high C-reactive protein or erythrocyte sedimentation rate (ESR), leukopenia, and atypical lymphocytes. Definitive diagnosis depends on lymph node biopsy. Three histopathological types of KD have been reported: proliferative, necrotizing, and xanthomatous. In a study, done at Israel the most common histological type was necrotizing type 63%, followed by xanthomatous and proliferative type 21% and 16% respectively.<sup>7</sup> The proliferative phenotype is composed of various histiocytes, plasmacytoid monocytes, and lymphoid cells with kariorrhctic nuclear fragments and eosinophilic apoptotic debris in the absence of neutrophils. Coagulative necrosis in the lymph node of any degree will classify the variant as necrotizing, while an increased number of foamy histiocytes in the lesion is a classical feature of xanthomatous variant regardless of the presence of necrosis. The histopathological features may also resemble that of systemic lupus erythematosus (SLE), an important differential diagnosis that should be excluded on the basis of clinical characteristics and laboratory tests. The diag-

nosis of KD is confirmed by the histopathologic findings in affected lymph nodes.<sup>10</sup>

KD is a benign and self-limited disease. The lymphadenopathy disappears spontaneously in most patients within few weeks to months. Less than 5% develop recurrent lymphadenopathy. There is no specific treatment for KD. The best symptomatic treatment is non-steroidal anti-inflammatory drugs.<sup>11</sup> Recurrence rate in children is 10%, which is higher than that of the adult patients i.e. 3–4%.<sup>12</sup>

## CONCLUSIONS

Kikochi disease can be a cause of cervical lymphadenitis and may mimic certain serious conditions like lymphomas and tuberculosis; these conditions should be considered in differential diagnosis.

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