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Kikuchi-Fujimoto disease: A comprehensive review

Mahajan VK *et al*. Kikuchi-Fujimoto disease

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Abstract

Kikuchi-Fujimoto disease, a rare form of necrotizing lymphadenitis, is an uncommon, benign, self-limiting disorder of obscure etiology. It affects mostly young adults of both genders. Clinically, it presents with fever and lymphadenopathy of a firm to rubbery consistency frequently involving cervical lymph nodes while weight loss, splenomegaly, leucopenia, and elevated erythrocyte sedimentation rate feature in severely affected patients. Cutaneous involvement occurs in about 30%-40% of cases as facial erythema and nonspecific erythematous papules, plaques, acneiform or morbilliform lesions of great histologic heterogeneity. Both Kikuchi-Fujimoto disease and systemic lupus erythematosus share an obscure and complex relationship as systemic lupus erythematosus may occasionally precede, develop subsequently, or sometimes be associated concurrently with Kikuchi-Fujimoto disease. It is often mistaken for non-Hodgkin lymphoma while lupus lymphadenitis, cat-scratch disease, Sweet's syndrome, Still's disease, drug eruptions, infectious mononucleosis, and viral or tubercular lymphadenitis are other common differentials. Fine needle aspiration cytology mostly has features of nonspecific reactive lymphadenitis and immunohistochemistry studies usually show variable features of uncertain diagnostic value. Since its diagnosis is exclusively from histopathology, it needs to be evaluated more carefully; an early lymph node biopsy will obviate the need for unnecessary

investigations and therapeutic trials. Its treatment with systemic corticosteroids, hydroxychloroquine, or antimicrobial agents mostly remains empirical. The article reviews clinicoepidemiological, diagnosis, and management aspects of KFD from the perspective of practicing clinicians.

Key Words: Cervical lymphadenopathy; Histiocytic necrotizing lymphadenitis; Kikuchi's disease; Lymphadenopathy; Lymphoma; Systemic lupus erythematosus

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Core Tip: Kikuchi-Fujimoto disease or Kikuchi disease described originally in young Japanese women is a rare benign cause of fever and lymphadenopathy usually involving cervical lymph nodes. The disease has been reported worldwide in both genders across ethnic and age groups. Histopathologically, histiocytic necrotizing lymphadenitis is characteristic and needs differentiation from more serious conditions such as malignant lymphoma in acute or subacute form. Its long-term prognosis is favorable albeit long-term follow-up is recommended because of a rare but increased risk of developing systemic lupus erythematosus/other autoimmune disorders. Patients with a severe or recurrent disease need treatment with nonsteroidal anti-inflammatory drugs, systemic corticosteroids, and/or other immunomodulators.

INTRODUCTION

Kikuchi-Fujimoto disease (KFD; *syn*: Kikuchi's disease, histiocytic necrotizing lymphadenitis), a rare self-limiting disorder of uncertain etiology, presents with prolonged lymphadenopathy with or without systemic features. The exact role of microbial infection and other non-infectious triggers in its etiopathogenesis remains obscure. Most patients present with firm to rubbery cervical lymphadenopathy and intermittent fever while weight loss, splenomegaly, leucopenia, and elevated

erythrocyte sedimentation rate occur in severely affected patients. Cutaneous involvement occurs in about 30% to 40% of cases[1,2]. Its diagnosis is mainly retrospective from characteristic histopathology of a diseased lymph node whereas treatment mostly remains empirical. Interestingly, both KFD and systemic lupus erythematosus (SLE) share a complex and as-yet unelucidated relationship as SLE may occasionally precede, develop subsequently, or sometimes be associated concurrently with KFD[3]. KFD is a clinical entity often misdiagnosed as malignant lymphoma in acute or subacute form or missed altogether owing to its rarity, similarity to other common conditions, and lack of awareness among clinicians. Herein we revisit the clinicoepidemiological, diagnosis, and management aspects of KFD from the perspective of practicing clinicians.

EPIDEMIOLOGY

Kikuchi[4] and Fujimoto *et al*[5] in the year 1972 independently described two separate cases from Japan one presenting with 'lymphadenitis showing focal reticulum cell hyperplasia, nuclear debris, and phagocytosis' and another having 'subacute necrotizing cervical lymphadenitis', hence the nomenclature 'Kikuchi-Fujimoto disease'. Since then several cases of KFD have been reported worldwide both in children and adults in all racial and ethnic groups across countries including populations of western and Asian origin, particularly with a higher prevalence among Japanese[6-10]. While most reports have appeared from Asia, its prevalence varies widely across ethnic groups. Seventy-five percent were White individuals in a study of 88 patients with KFD in a study from the United States[6]. Only 0.5% of 920 Lymph nodes had histopathology characteristics of KFD in a study from Saudi Arabia[11]. KFD-associated adenitis was present in only 5.7% in a report from Taiwan compared to 35% of 147 Korean patients with cervical lymphadenitis[2,12]. However, the exact prevalence of KFD remains understudied perhaps for want of clinical suspicion and validated diagnostic criteria. Although initial reports suggested a female preponderance, it affects mostly young adults of both genders with variable frequency. It has been reported in patients aged

between 6 and 80 years with a mean age of 30 years at presentation but the majority of affected individuals were younger than 40 years of age across studies[6,13-16]. The women were affected more often than men in three separate reports with a male to female ratio of 1:4, 1:1.6, and 1:1.3, respectively[6,17,18]. However, in a series of 20 Korean patients aged less than 18 years both genders were affected equally[19]. Among children, boys were affected slightly more frequently than girls compared to patients in the older age group in other reports[20,21]. A large review of 733 patients diagnosed worldwide since 1972 the male:female ratio was 1.4:1.8 for 19% of pediatric patients but a higher propensity for male gender was in children younger than 12 years of age[22,23].

ETIOLOGY

The clinicopathologic features of KFD favor a T-cell and histiocytes-mediated immune response to an infectious or a non-infectious trigger. Increased levels of IFN-α and its stimulators including 2',5'-oligoadenylate synthetase, and tubuloreticular structures in the cytoplasm of stimulated lymphocytes, histiocytes, and vascular endothelium favor a viral etiology whereas a favorable therapeutic response to minocycline, ciprofloxacin, and ofloxacin in few studies suggest a pathogenic role of other non-viral microorganisms sensitive to these antimicrobial agents[8,22-25]. Nonetheless, several microbial agents including Epstein-Barr virus (EBV), human herpesvirus (HHV) 6 and 8, human immunodeficiency virus (HIV), parvovirus B19, paramyxoviruses, parainfluenza virus, *Yersinia enterocolitica*, and *Toxoplasma*, *Streptococcus pneumoniae* have been implicated as inciting factors[26-31]. Although EBV had been the most observed trigger, immunohistochemistry detected EBV-encoded protein in only one of the 10 patients showing EBV by in-situ hybridization[23,26,27,30]. A minivirus resembling circovirus, the causative agent for swine necrotizing lymphadenitis, too has been isolated in three Korean patients[32]. Although the exact relationship remains obscure, a postviral hyperimmune reaction too is a potential trigger for KFD[33,34]. B-cell lymphoma, rupture of silicone breast implant, infections due to cat-scratch disease, and *Mycobacterium szulgai* are other proposed triggers for KFD[8,35,36].

KFD has been infrequently reported following vaccination for influenza, Japanese encephalitis, and HPV[37,38]. Recently, KFD has been linked to coronavirus disease 2019 infection and SARS CoV2 vaccination[39-48]. However, whether the association is causal or epiphenomenon remains uncertain as most patients recovered rapidly and completely. One of the two patients developed KFD disease 10 d after vaccination while hemophagocytic reticulosis complicated KFD after the vaccination in another[45,46].

It has been suggested that KFD most likely develops due to an autoimmune mechanism disease as cases had reportedly developed clinicopathological features similar to Sjögren's syndrome and systemic lupus erythematosus/mixed connective tissue disease[49,50]. Since KFD shares age and gender predisposition as well as histologic features with SLE, it has been also proposed that KFD perhaps represents a self-limiting SLE-like autoimmune disorder caused by virus-infected transformed lymphocytes[51]. The hypothesis of autoimmune pathogenesis is further supported by an association of KFD with an increased risk of its evolution into an autoimmune syndrome, particularly in females or the concurrent presence of other autoimmune disorders such as Hashimoto thyroiditis, primary Sjögren's syndrome, antiphospholipid syndrome, and leukocytoclastic vasculitis as observed in 53% of thirteen women in a follow-up study of twenty patients with KFD[52-55].

The reports implicating allogeneic hematopoietic stem cell transplantation and bariatric surgery for triggering KFD remain anecdotal[56,57].

PATHOGENESIS

No genetic transmission or inheritance of a definite pattern has been identified despite being reported uncommonly to affect twins and even human leukocyte antigen (HLA)-identical non-twin siblings[58-61]. Some of these familial cases were from the common living environment and developed symptoms almost simultaneously and demonstrated serological evidence for the same infection. However, one of the two HLA-identical sisters but not twins from Saudi Arabia developed KFD after a gap of 10 years with no identifiable infectious trigger[58,59]. Stasiuk *et al*[58] reported a similar case of two

Aboriginal sisters from northern Ontario reflecting a genetic predisposition among affected individuals. KFD and HLA class 2 alleles have been linked suggesting a possible positive relationship between DPA1*01 and DPB1*0202 alleles which are much more frequent in Japan than in Europe and the United States[62]. This perhaps accounts for a higher prevalence of KFD in the Asian population.

Up-regulation of cell-cycle-associated genes and apoptosis-associated genes including caspase, and down-regulation of apoptosis-inhibitory genes such as *BCL2* too have been observed in KFD-associated lymph nodes and not in nonspecific lymphadenitis[63]. The apoptotic cell death induced by the Fas-Fas ligand system mediated by cytotoxic CD8+ cells is the principal mechanism of cellular destruction in KFD[64-66]. The apoptosis is enhanced by histiocytes as evident from the morphology of apoptotic cells on transmission electron microscopy characterized by nuclear chromatin condensation, fragmentation along the nuclear membrane with intact organelles, and histiocytes phagocytosing karyorrhectic debris (apoptotic bodies). To summarise, primarily the activation of T lymphocytes and histiocytes occurs and the activated T cells enter the apoptotic cycle manifesting with areas of necrosis in lymph nodes, and the cellular debris is then cleaned up by the histiocytes. Also the cytokine and chemokine pathways of interferon (IFN)-γ, interleukin (IL)-18, MIG, and IFN-γ-induced protein 10 have been suggested to play a significant role in the apoptosis associated with KFD[67]. The serum levels of IFN-γ and IL-6 were elevated but not of IFN-α, tumor necrosis factor (TNF), or IL-2 in these patients during the acute phase of illness which returned to normal during convalescence[68,69]. This further reinforces the possible role of IFN-γ and IL-6 in the pathogenesis of KFD. In general, the shared and common HLA and cytokines involved in the molecular pathogenesis of this disease suggests that like other HLA-linked disorders, KFD is a two-step disease requiring a predisposing HLA and a secondary trigger such as an infection in individuals with a genetic predisposition for the exuberant T-cell mediated immune/inflammatory response[70]. However, the exact molecular pathogenesis of KFD remains complex and needs further elucidation.

CLINICAL FEATURES

KFD usually manifests as an acute or subacute illness with fever and painful posterior cervical lymphadenopathy generally in a previously well individual and evolving over 2-3 wk[71]. Lymph nodes of firm to rubbery consistency are usually small ranging from 0.5 to 4 cm in size[18,72,73]. The most common signs and symptoms were lymphadenopathy (100%), fever (35%), erythematous rash (10%), arthritis (7%), fatigue, (7%), and hepatosplenomegaly in 3% of patients in a series of 244 individuals with KFD[74]. The association of splenomegaly too has been reported as an uncommon feature of KFD[75]. Typically, the fever is of low grade and intermittent which persists for about one week, rarely lasting for up to one month (median duration 9 d), and is a primary symptom in 30%-50% of patients[6]. Patients with higher fever (≥ 39.0 °C), larger lymph nodes, and leucopenia can have a more prolonged clinical course[76]. However, the diagnosis of KFD is unlikely to be considered without lymph node biopsy in patients with pyrexia of unknown origin. Other sporadic symptoms and signs include rigors, myalgia, arthralgia, and chest and abdominal pain[6]. Nausea, vomiting, diarrhea, night sweats, and weight loss have been reported more often in patients with extranodal disease[77].

Lymph node involvement

The lymph node involvement is the commonest presenting feature. The lymph nodes are usually enlarged only moderately varying in size from 1 to 2 cm in diameter but sometimes they are much bigger up to 7 cm in diameter[78]. The enlarged lymph nodes are typically discrete, mobile, firm, smooth, and often associated with dull or acute pain. In the majority, the lymph node involvement is limited to unilateral cervical lymphadenitis particularly localized to posterior cervical lymph nodes but there may rarely be bilateral cervical adenopathy[6,78]. Generalized lymphadenopathy involving axillary, epitrochlear, mediastinal, mesenteric, inguinal, intraparotid, iliac, retrocrural, celiac, peripancreatic, and retroperitoneal nodes occurs in about 1% to 22% of patients[71]. Extracervical disease especially with bilateral cervical adenopathy and

leukopenia in a review of 60 patients (mean age of 21 years) with confirmed KFD manifested with abdominal in 52%, pelvic in 47%, inguinal in 41%, and axillary lymph nodes in 30% of patients, respectively[79]. Mediastinum lymphadenopathy can occur more often while the involvement of mesenteric lymphadenitis is rare and often mimic appendicitis[80-83]. However, nodal enlargement may be minimal or remain limited to mediastinal or retroperitoneal nodes only[84,85].

Cutaneous manifestations

The involvement of skin in 30%-40% of KFD patients is the most commonly affected extranodal organ[1,2]. Facial erythema, SLE-like malar rash, and other nonspecific lesions including macules, patches, papules, nodules, or plaques, occasionally pruritic, and histologically resembling KFD occur typically and are observed more frequently in children than adults[2,21,86,87]. More sick patients may develop transient skin rashes resembling rubella or drug-induced eruptions[88,89]. Other cutaneous manifestations include pruritus, lichen planus, polymorphous light eruptions, scattered indurated lesions, leukocytoclastic vasculitis, ulcers, scales, and alopecia[2,26,86,90]. Mucosal involvement in the form of oropharyngeal redness, oral ulceration, and conjunctival injection/papillary conjunctivitis may occur[8,91].

Systemic manifestations

Multiple systemic complications may occur. Weight loss is common in adult patients and leukocytosis is perhaps more frequent than leukopenia[20,87]. An association of KFD with various other inflammatory disorders such as hemophagocytic syndrome, arthritis, myocarditis, and hepatic dysfunction has been indicated[87,91-97]. Macrophage activation syndrome, a secondary form of hemophagocytic lymphohistiocytosis (HLH), has been recognized in some older patients hospitalized under intensive care with KFD and HLH who required systemic corticosteroids or had ended fatally[98].

Involvement of the central nervous system presenting as aseptic meningitis, meningoencephalitis, acute cerebellar symptoms with tremors and ataxia, and optic

neuritis has been reported frequently[93,98-104]. Aseptic meningitis mostly at the time of the lymphadenopathy is the commonest neurological complication seen in some of these cases and may be associated with very high levels of intracranial pressure, low cerebrospinal fluid (CSF)-serum glucose, and recurrent subdural effusions requiring intervention[105,106]. While recurrent meningitis occurs more often in males, encephalopathy in children may occur after 10 d to 3 mo which is characterized by very high CSF protein levels and extensive magnetic resonance imaging changes requiring early treatment.

DIAGNOSIS

Distinguishing KFD from malignant lymphoma and SLE remains a major diagnostic challenge owing to similar histopathologic appearances. Patients with KFD especially in its proliferative phase have been often misdiagnosed as having non-Hodgkin or Hodgkin lymphoma leading to extensive investigations and, in some cases, aggressive treatment with cytotoxic agents owing more so to unfamiliarity with this uncommon entity[6,107]. Parotid gland tumor, lupus lymphadenitis, Kawasaki's disease, cat-scratch disease, Sweet's syndrome, sarcoidosis, lymphogranuloma venereum, drug eruptions, infectious mononucleosis, and viral or tubercular lymphadenitis are other more common differentials[6,8,107-109]. Mesenteric lymphadenopathy presenting as acute appendicitis too has been described[69].

The diagnosis of KFD is mostly retrospective and based exclusively on lymph node histopathology and excisional biopsy has been recommended frequently in the past. However, fine needle aspiration (FNA) histology with features varying from nonspecific reactive lymphadenitis to those of KFD can be a useful diagnostic tool with an overall accuracy of about 56% in expert hands; nonetheless, it provides clues to exclude other common causes of lymphadenopathy[110]. Ultrasound-guided FNA using smears and cell block preparations designed to preserve lymph node architecture is considered a better alternative[111-113]. However, ultrasound-guided core needle biopsy with a diagnostic accuracy of 95.6% as compared with FNA is now the suggested

diagnostic modality of choice[114]. Extracellular debris and intracellular apoptotic debris embedded in the cytoplasm of crescentic and phagocytic macrophages are characteristic of KFD[115,116]. Nevertheless, lymph node biopsy should be preferred for an early diagnosis, to avoid unnecessary investigative work up, and to exclude more serious conditions such as lymphoma requiring early and intensive therapeutic intervention.

Histopathology

The cut surface of the involved lymph nodes may show yellowish necrotic foci on gross examination. Histological examination (Figure 1) usually shows single or multiple foci of focal or complete loss of follicular architecture often with necrosis of the cortical and paracortical areas and extensive infiltrate consisting of small lymphocytes, immunoblasts, histiocytes, and plasmacytoid T-cells[8]. Perinodal inflammation is common and the capsule may be infiltrated. The necrotizing process is often localized to areas of eosinophilic fibrinoid material with fragments of nuclear debris distributed irregularly. However, obvious coagulative necrosis is not essentially diagnostic of KFD.

The histologic appearance changes as the disease progresses and can be classified into three histologic subtypes; an early phase of “proliferative type” in more than 30%, a late phase of “necrotizing type” in 50% being the commonest, and the end phase of “xanthomatous type” the least common form seen in < 20% cases[78]. The presence of follicular hyperplasia and paracortical expansion by lymphocytes, T and B cell blasts, plasmacytoid monocytes, and histiocytes with numerous background apoptoses is the hallmark of the early "proliferative phase" of KFD. The numerous blast cells may evoke the possibility of lymphoma or EBV/HHV infection. While preservation of the nodal architecture, polyclonal infiltrate, absence of monoclonal T-cell receptor rearrangement, and negative viral serology will exclude these possibilities, its oligoclonal pattern and spontaneous regression favor a more benign immune reaction[117]. The "necrotizing phase" with an absence of a neutrophilic infiltrate is predominately associated with progressive dominance of histiocytes often with crescentic nuclei and phagocytosed debris as the major cell type. Histiocytes with predominantly CD8+ T lymphocytes and

CD68+ plasmacytoid monocytes are visualized on immunohistochemical staining[118]. The absence of neutrophils in the necrotizing phase differentiates KFD from SLE and drug-induced lymphadenopathy. The "xanthomatous type" seems to be a distinct histologic variant or perhaps indicates a healing phase characterized by the predominance of foamy histiocytes, and necrosis may or may not be present[119,120].

Systemic lupus erythematosus, tubercular or viral lymphadenitis, and Hodgkin and non-Hodgkin lymphoma are common histological differentials. The absence of Reed-Sternberg cells, the presence of numerous histiocytes, and relatively low mitotic rates favor the diagnosis of KFD rather than lymphoma[121]. The histology of the lymph node in KFD can be differentiated easily from most known infectious causes of fever and lymphadenopathy but its differentiation from lymphadenopathy in SLE may be particularly difficult. However, positive results for immunohistochemistry taken together with clinicopathologic features may be of diagnostic help[2,6,13,122].

In contrast, the histologic features of skin lesions in KFD are mostly nonspecific and highly variable featuring epidermal changes mainly of interface dermatitis, necrotic keratinocytes, non-neutrophilic karyorrhectic debris, basal vacuolar change, papillary dermal edema, and a lymphocytic infiltrate[2,123]. Vacuolar interface changes in 75%, necrotic keratinocytes in 68%, karyorrhexis in 100%, superficial and deep lymphohistiocytic infiltration in 100% and 56%, respectively, and panniculitis in 60% were major histologic features noted in a retrospective review of skin biopsies from sixteen patients with KFD[124]. Atwater *et al*[2] proposed that diagnosis of KFD can be reasonably made in an appropriate clinical setting if the biopsy specimen meets the following criteria: (1) a dermal (lympho)-histiocytic infiltrate; (2) epidermal changes with an emphasis on necrotic keratinocytes; (3) non-neutrophilic karyorrhectic debris; (4) basal vacuolar change; and (5) papillary dermal edema. However, confirmation is needed by immunohistochemical staining with CD68 for the presence of histiocytes.

Immunohistochemistry

Hassan *et al*[125] observed positive immunostaining by antibodies Mac 387, KP1 (CD 68), and Ki M1P, and the majority of the cells were a mixture of CD8+ and CD68+ in the affected foci (Figure 2).

Additionally, a variable number of T-cells immunostained by antibody MT1 (CD 43) or UCHL1 and (CD 45RO) and CD8+ T cells stained positive with antibody CD8/144 in all lesions. They opined that immunohistochemistry can be useful in differentiating KFD from other chronic cervical lymphadenopathies. KFD can be distinguished from necrotizing lymphadenitis due to other causes with digital quantification of CD123 immunohistochemical staining[126]. There are fewer surrounding mononuclear cells and neutrophils are usually present in HHV-associated lymphadenitis. Contrasting with KFD, necrosis in Hodgkin lymphoma usually includes neutrophils and CD15, CD30, and CD45 positive Reed-Sternberg cells, the large atypical cell variants. The plasmacytoid dendritic cells infiltrate the lymph nodes more frequently in KFD irrespective of its duration than in either reactive lymphadenitis or T or B cell lymphoma and can be useful cytologic indicators in the diagnosis of KFD[127]. The presence of abundant CD8+ cytotoxic T cells around necrotic areas can help in differentiating necrotic lymph nodes from head and neck cancer or metastatic disease from SLE and reactive lymphoid hyperplasia[128,129]. CD68+ and CD163+ histiocytes and CD3+ T lymphocytes are predominant infiltrating cells in skin lesions whereas, neutrophil-predominant inflammation with superficial dermal edema features in skin lesions with Sweet syndrome-like morphology[124,130].

Laboratory investigations

No specific laboratory investigations are recommended for the diagnosis of KFD but these are advocated to exclude other causes of (cervical) lymphadenopathy. Although atypical lymphocytes in up to 25% and leukopenia in up to 43% have been observed, the blood counts are usually normal in a majority of the patients with KFD[6,12,17,74]. Thrombocytopenia, pancytopenia, and anemia of chronic disease in those with severe disease are other reported hematological abnormalities[26,74,131]. The erythrocyte

sedimentation rate and C reactive protein can be normal but were elevated (> 60 mm/h) in 70% of patients in one series[84]. Mildly abnormal liver function tests and elevated serum lactate dehydrogenase are other nonspecific findings[97].

Increased macrophages without atypical cells are frequently seen in bone marrow cytology whereas, increased numbers of mature hemophagocytic histiocytes in the bone marrow may cause diagnostic confusion with virus-associated hemophagocytic syndrome[17,78]. However, bone marrow studies are rarely recommended for the diagnosis of KFD.

Serodiagnostic studies

Serology for EBV, HHV, cytomegalovirus, HIV, toxoplasmosis, *Y. enterocolitica*, cat scratch disease, and other infectious agents is often performed in a suspected case of KFD to exclude other differentials of fever and lymphadenopathy. Antinuclear antibodies (ANA) and other investigations for SLE, and rheumatoid factor are, although not always, negative and help distinguish the two. However, patients initially diagnosed with KFD may have concurrent or developed SLE subsequently, it is generally recommended to perform an ANA test in patients suspected of KFD with features suggestive of SLE and to exclude this diagnosis[6,132]. There may be a transient rise in anti-deoxyribonucleic acid and anti-ribonuclear protein antibody levels[17]. Screening for the presence of adult-onset Still's disease in patients with suspected KFD is also recommended because of reports of their concurrent occurrence[133].

Imaging studies

No specific radiographic finding has been established to make a diagnosis of KFD. However, a chest X-ray should be obtained in a routine workup of a patient with fever and cervical lymphadenopathy for any evidence of pulmonary tuberculosis or malignancy. Ultrasonography of the neck for lymph nodes can be performed but results need careful interpretation as in a series of 29 lymph nodes evaluated by sonography 66% showed features suggestive of malignancy whereas tubercular lymphadenitis

remains another common differential[122,134]. Features such as posterior neck involvement, echogenic hilum, absence of internal calcification and necrosis (rarely present in partial necrosis), normal vascular pattern (hilar vascular structures are central or branch radially from the hilum in both longitudinal and transverse planes) on power Doppler ultrasound are considered typical of KFD[122]. In Doppler ultrasonography, the lymph nodes are smaller, less rounded, and mostly have an echogenic hilum and abnormal posterior cervical region but necrosis and internal calcifications are less likely in KFD as compared to tuberculous adenitis[122]. Computed tomography (CT) of the neck may be helpful before biopsy and the features may be similar to lupus lymphadenitis and malignant lymphoma. It exhibits unilateral, uniform homogeneous enlargement and post-contrast enhancement of cervical lymph nodes preferably affecting levels II-V in most patients with KFD[135]. Perinodal infiltration is typically seen in about 81% and homogenous nodal contrast enhancement occurs in 83% of affected lymph nodes[135]. The pattern of necrosis, when present, suggests tuberculosis while the absence of necrotic lymphadenopathy and nodal cortical attenuation and its ratio to adjacent muscle on CT imaging differentiates KFD from tuberculous lymphadenopathy and nodal reactive hyperplasia[136]. Magnetic resonance imaging findings of 52 enlarged cervical lymph nodes predominantly showed unilateral distribution at level II-V in a study[137]. Areas of hypointensity in peripheral distribution and clear margins representing necrosis in paracortical areas, and occasional focal non-enhancing areas suggestive of necrosis within the enlarged lymph nodes were observed in T2-weighted images in the same study. Positron emission tomography scanning has been used to assess disease severity and most patients are found to have multiple hypermetabolic lymph nodes but only a few were enlarged[138,139]. Radiating vessels from the central hilum to the periphery of the lymph node and normal or scant hilar vascularity within the whole lesion in 92% of patients with KFD on color Doppler imaging indicates its benign nature[140]. Lee *et al*[141] performed real-time elastography in two patients with KFD to measure the elasticity score of affected cervical lymph nodes. Based on a scale of 1 to 4 devised by Ying *et al*[142] the elasticity score of 1 in both

patients was consistent with the benign nature of KFD. However, all these investigative modalities need further evaluation for their diagnostic accuracy compared to histopathology, the gold standard in diagnosis.

SYSTEMIC COMPLICATIONS

Although considered benign and self-limiting, systemic complications in KFD may develop prompting urgent intervention. These include cardiac tamponade, pleural effusions, pulmonary infiltrates and nodules (interstitial lung disease), symmetrical polyarthritis, thyroiditis and parotid enlargement, autoimmune hepatitis, acute renal failure, bilateral papillary conjunctivitis and panuveitis, ocular vasculitis and subretinal macular infiltrates, polymyositis, hemophagocytosis, limb paresis because of brachial plexus neuritis, peripheral neuropathy, and antiphospholipid syndrome with multiorgan failure[143-159]. Still's disease, cryptogenic organizing pneumonia, hemophagocytic syndrome, and B cell lymphoma have been described occasionally in association with KFD[160-166].

KFD AND SYSTEMIC LUPUS ERYTHEMATOSUS

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KFD shares gender and age predisposition as well as histologic features with SLE. SLE may occasionally precede, sometimes be associated with KFD, or develop subsequently. In a review of 55 patients with KFD in the context of definite connective tissue disorder fifty were associated with SLE; 22 (40%) patients had simultaneous onset, 19 (35%) patients predated the onset, and 14 (25%) patients developed KFD after the onset of SLE[3]. KFD may also complicate preexisting SLE as described by Tarabichi *et al*[167] requiring intensive therapeutic intervention. Their patient of SLE, a 20-year-old girl, had been doing well with hydroxychloroquine (HCQS 200 mg/d)/analgesics. But, after two months she developed KFD affecting multiple cervical/extra cervical lymph nodes and hepatomegaly and needed high doses of systemic methylprednisolone (250 mg/d, given as pulse therapy for 5 d) and 40 mg/d thereafter in addition to HCQS (200 mg twice daily) for adequate control.

Many reports of KFD cases also support the idea that the two diseases share common immunopathological and clinical features such as the disappearance of the lesions without any specific treatment whereas, the possibility of recurrence suggests the involvement of immune mechanisms and that KFD could be a common aspect of SLE[168-170]. One ultrastructural study proposed that KFD perhaps reflects a self-limiting, SLE-like autoimmune condition triggered by virus-infected transformed lymphocytes[51]. The histological findings of focal necrosis with immunoblastic infiltration in lupus lymphadenitis are indistinguishable from KFD. The absence of neutrophils is characteristic of both KFD and lupus lymphadenitis[8]. The tubuloreticular structures in the lymphocytes and endothelial cells in patients with SLE have similarities to those seen in KFD whereas a similar tubuloreticular structure in human lymph nodes is not seen except in cases of SLE. However, the detection of this peculiar structure of unknown origin in an ultrastructural study of necrotizing lymphadenitis suggests a direct relationship between SLE and KFD[51]. In addition, the key distinguishing histologic feature of SLE lymphadenitis is the presence of hematoxylin bodies, an amorphous aggregate of basophilic material[169].

Skin lesions in KFD may also clinically and histologically resemble those seen in SLE or subacute lupus erythematosus[171]. Skin biopsy of KFD lesions reveals a pattern of interface dermatitis in KFD that evolved into SLE suggesting that it could be a histopathological marker of evolution into SLE and might predict clinical outcomes in KFD[128]. An association of KFD with hemophagocytic syndrome also suggests a common pathogenesis[170].

According to Tabata *et al*[129], CD30 immunostaining may help in differentiating KFD from SLE as CD30+ cells significantly are more numerous in KFD than in SLE and most of these were located around necrotic areas in their study of 30 patients with KFD and six patients with SLE. These CD30+ cells were CD8+ activated cytotoxic T cells around necrotic areas and characteristic of KFD and occurred predominantly in females having only mild symptoms and normal laboratory data[129]. The relationship between KFD and SLE remains complex and uncertain requiring long-term follow-up because of late

evolution of KFD to SLE or other autoimmune diseases after several months to years[49,123,173].

KFD IN PREGNANT WOMEN AND CHILDREN

Although KFD affects women younger than 40 years of age more often, there is a paucity of data on its impact on gestation and pregnancy outcomes. The treatment with antibiotics, steroids, or both had no adverse impacts on the mother, fetus, or the course of pregnancy in two patients with KFD manifesting during pregnancy[173,174]. Another patient with KFD had a miscarriage reportedly with evolving SLE[175].

The presenting clinical features in children are similar to those in adults, although fever, rash, and bilateral cervical lymphadenopathy are more frequent in children younger than 18 years than in adults[72,176-178]. Generalized lymphadenopathy was less common while fever and rash were more common in children as compared to adults[177]. The presentation may be atypical in children < 6 years old. Recurrent disease seen in approximately 3% of pediatric patients is uncommon and occurs more often in boys compared to adults where recurrences have been more frequent in women[21,74].

TREATMENT

No specific treatment has been established as signs and symptoms usually resolve spontaneously within 1 to 4 mo without serious sequelae[179]. Fever usually subsides after removal of the affected lymph node suggesting the possible therapeutic benefit of excisional biopsy by removal of the focus for the inflammatory process in addition to being diagnostic[180]. Pharmacotherapy is usually targeted to reduce morbidity and prevent complications. Nonsteroidal anti-inflammatory drugs (NSAIDs) usually suffice to alleviate fever and lymph node tenderness in mild cases. Patients with prolonged fever, severe or symptoms lasting for more than 2 wk, and recurrent disease have been treated with immunomodulators, systemic corticosteroids (prednisolone 1-2 mg/kg body weight) alone or combination, high-dose corticosteroids and intravenous immune globulin (IVIg) especially in patients presenting with extranodal or generalized severe

disease or hemophagocytic syndrome with good therapeutic outcome[181-183]. There have been reports of recurrent or steroid-resistant KFD treated successfully with hydroxychloroquine monotherapy or in combination with systemic corticosteroids[184,185]. Rezai *et al*[184] treated a patient of KFD with systemic symptoms with chloroquine and achieved a rapid response in four days. One patient unresponsive to steroid therapy reportedly responded well to the IL-1 inhibitor, anakinra[186]. The anecdotal efficacy of ciprofloxacin, minocycline, and ofloxacin in remitting KFD needs further evaluation[8,24,25].

PROGNOSIS

In general, the prognosis in KFD is good as it is self-limiting in a majority of the patients and the symptoms may spontaneously subside in 1-6 mo, although recurrences can occur in 3% to 4% cases[131,187-189]. In a series with a median of 32 mo of follow-up 92% of patients were alive and well[6]. Recurrences respond well to treatment similar to the first occurrence. Nevertheless, affected patients should be followed up for some years as unpredictably they may develop SLE, and sometimes recurrences of KFD can continue for many years. For instance, in a Korean case series recurrence was seen in 11.3% and 2.7% of patients who developed autoimmune diseases after an initial diagnosis of KFD[190]. Similarly, four episodes of lymphadenopathy over 18 years occurred in a patient and another had two episodes separated by 6 years[131,187]. In another Korean series of 102 patients (mean age 26.7 years) followed up between 2001 and 2006, late recurrence happened in 13% of patients while early relapse occurred in 8% of patients while 3% of patients developed SLE[188]. These recurrent cases had fever and fatigue with extranodal involvement and remained symptomatic for a longer period than non-recurrent cases. A positive fluorescent antinuclear antibody test was found to be associated with a significantly higher risk of recurrent disease. An EBV seropositive and ANA negative KFD presenting initially with severe disease relapsed frequently despite being treated with multiple courses of systemic corticosteroids and hydroxychloroquine[191]. With a reported fatality rate of 2.1% and severe and fatal cases

having been generally associated with hemophagocytic syndrome or connective tissue disease, a long-term follow-up remains essential[3,74,192].

CONCLUSION

Kikuchi-Fujimoto disease is an uncommon, idiopathic, benign lymphadenopathy primarily affecting both genders at a young age between 20 and 40 years worldwide across ethnicities. The acute or subacute onset of adenopathy is usually associated with fever, body aches, night sweats, and weight loss (B symptoms). Its exact etiology remains obscure. The results of studies looking for viral etiology have been inconsistent. Molecular pathways implicated in its pathobiology are also not well understood. Concomitant autoimmune disorders have been reported or they may also be diagnosed more frequently following the resolution of KFD. Despite differing opinions, SLE is perhaps the commonest disorder linked to KFD and has been suggested to be its *forme fruste*. However, further research is necessary to reach a definitive conclusion.

Although modern hematopathological methods have made misdiagnosis less likely, a diagnosis of malignant lymphoma for KFD is still reported. Since the diagnosis is exclusively based on histopathology, an early excision biopsy of the affected lymph node will be diagnostic in doubtful cases. However, using high-throughput nucleotide sequencing of lymph node tissue to identify exomes and transcriptomes, candidate single nucleotide polymorphisms for markers of KFD have been identified and the altered gene expression found may help in better understanding of the disease, diagnosis, and treatment[193]. The proposed diagnostic criteria using a scoring system based on temperature, maximum lymph node size, and serum B2 microglobulin described for children avoiding lymph node biopsy had 100% sensitivity and specificity in a study[194]. However, further evaluation with multicenter studies involving larger numbers of patients across all age groups remains highly desirable.

Treatment guidelines for KFD have not been established and recommendations are mostly based on case reports, small case series, and the opinion of the experts. Treatment with systemic corticosteroids, NSAIDs, hydroxychloroquine or other

immunomodulators, used alone or in combination, in a few patients with severe symptoms or recurrent disease remains the mainstay in therapeutics. Anecdotal therapeutic efficacy and place of chloroquine, anakinra, IVIg, and antimicrobial agents in the treatment ladder need to be evaluated further.

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