Kikuchi-Fujimoto Disease in Michigan: A Rare Case Report and Review of the Literature

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ABSTRACT

BACKGROUND: Kikuchi-Fujimoto Disease (KFD), also known as Histiocytic Necrotizing Lymphadenitis, is a rare cause of prolonged cervical lymphadenopathy in both the pediatric and adult populations. It was first reported in Japan in 1972, and since, this disease has been described worldwide, although most cases have been reported in Asia. The etiology of KFD is not fully understood, although there are 2 theories that are described in detail in this review. Kikuchi-Fujimoto Disease typically follows a benign course, with resolution of the lymphadenopathy within 6 months. It is important to recognize KFD as a cause of persistent lymphadenopathy, as it shares many characteristics with and must be differentiated from other causes of lymphadenopathy, including lymphoma, inflammatory disorders, autoimmune conditions, and infectious causes of lymphadenopathy.

CASE PRESENTATION: Here is presented a case of an 11-year-old male who was born in Vietnam, but subsequently adopted and raised in the United States, who presented to a private practice community-based Otolaryngology group. His chief complaint was a persistent neck mass of approximately 3 months duration. He underwent excisional biopsy for suspected lymphoma, but final pathology rendered a diagnosis of KFD.

CONCLUSION: The purpose of this article is not only to review the literature but also to contribute awareness of this entity in the differential diagnosis of persistent lymphadenopathy, especially for the general Otolaryngologist in a community-based setting. In addition, this review would be beneficial for other practitioners as well, specifically Pediatricians, Infectious Disease Physicians, Rheumatologists, Pathologists, and Medical Oncologists.

KEYWORDS: Kikuchi-Fujimoto Disease, histiocytic necrotizing lymphadenitis, lymphadenopathy, systemic lupus erythematosus, diagnosis, treatment, prognosis

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Introduction/Background

Kikuchi-Fujimoto Disease (KFD), also known as Histiocytic Necrotizing Lymphadenitis, is an uncommon disease characterized mainly by fever and lymphadenopathy. It was first described in Japan in 1972.[1,2] Typically, this clinicopathologic disease is a benign and self-limited condition. It is known to be uncommon in the United States, with most cases occurring in Asia, although there have been cases described worldwide in the literature. There is a higher incidence of KFD in women, aged 20 to 35 years.[3] However, there are numerous recent reports now describing the diagnosis in younger, pediatric patients.

In 2003, Lin et al[4] published an article with their experience in Taiwan, demonstrating 61 patients with KFD over a 15-year period, from 1986 to 2000. In 2007, a review by Kucukardali et al identified 330 cases of KFD (including both adults and children) worldwide during a 15-year period. Of these 330 cases, the majority of them were diagnosed in Asian countries, with only 22 cases (7%) diagnosed in the United States.[5] In 2013, the Children's Medical Center of Dallas published their experience with KFD. Over a 15-year period, from 1995 to 2009, only 9 patients were diagnosed with KFD.[6] More recently, in 2014, the Connecticut Children's Medical Center published their experience with KFD. Between 1996 and 2012, there were only 4 confirmed cases of KFD diagnosed. According to this article published in 2014, there were only 10 previously published case reports of KFD acquired in the United States identified.[7] These numbers demonstrate the paucity of cases of KFD diagnosed in the United States.

Kikuchi-Fujimoto Disease usually manifests with fever and lymphadenopathy, with the jugulo-carotid and cervical lymph nodes being the most commonly affected.[8–11] It is important to recognize KFD as a clinical identity, as it may mimic other conditions such as lymphoma, infectious etiologies, and autoimmune diseases, specifically systemic lupus erythematosus (SLE). According to 1 study, 30% of cases of KFD were incorrectly diagnosed as lymphoma.[12] This review reports an 11-year-old male that presented to a private practice Otolaryngology clinic (North Oakland ENT) in Clarkston, Michigan, with cervical lymphadenopathy. The precise etiology of KFD is unknown. In this article, a review of the proposed etiologies will be discussed, along with clinical presentation, diagnostic modalities, treatment options, and prognosis.

Case Report

The patient was an 11-year-old male with a past medical history positive for psoriasis that presented in December 2016 with a complaint of an enlarged lymph node on the right side of the neck of approximately 3 months' duration. He was born in Vietnam, but adopted at a young age and raised in Michigan. Therefore, family history could not accurately be obtained. There was no history of any precipitating viral upper respiratory infection or any other inciting events. Over 3 months, there were no changes or fluctuation in size of the enlarged lymph node. There was no history of dysphagia, odynophagia, dysphonia, or otalgia. There was no history of weight loss or night sweats. In addition, there was no history of fever.

The patient had previously been evaluated by a primary care physician who trialed a course of antibiotics without any improvement or resolution of the neck mass. In addition, an ultrasound of the neck was obtained which demonstrated multiple large right-sided neck lymph nodes, with the largest measuring 2.7 cm. Multiple lymph nodes in the left neck, which measured approximately 1 cm, were noted as well. Epstein-Barr Virus (EBV) serology was also performed. Epstein-Barr Virus Capsid IgG was positive, EBV Capsid IgM was negative, EBV Early Antigen AB was positive, and EB Early Antigen AB was positive. These results indicated the patient was likely exposed to an EBV infection in the past, with no active or recent infection. A Monospot test was also completed and negative.

On clinical examination, the patient's neck demonstrated palpable right-sided cervical lymphadenopathy, with a prominent 2-cm lymph node in level 5. It did not appear infectious, as there was no evidence of erythema, edema, or warmth. Other than this palpable lesion, the remainder of his otolaryngologic examination was unremarkable and the patient remained completely asymptomatic. No other infectious serology or workup was pursued.

Given the chronicity of this enlarged lymph node, a CT soft-tissue neck with and without contrast was obtained. The Computed Tomography (CT) scan demonstrated diffuse lymphadenopathy, particularly within the right posterior triangle of the neck. The largest lymph node measured approximately 1.6 to 2 cm in greatest dimension. At this point, there was high suspicion for lymphoma. Therefore, an excisional lymph node biopsy was performed and the gross specimen was sent fresh for histopathological diagnosis. Given the low suspicion for an infectious etiology, no specimen was sent for culture.

According to the pathology report provided by the local community hospital, the specimen demonstrated "extensive necrotizing granulomatous inflammation with abscess formation, suggestive of cat-scratch disease—pending final consultation." However, further analysis was requested, and the specimen was also sent to the University of Michigan Laboratories. On further analysis, the final pathology report concluded the specimen as a benign reactive lymph node, consistent with KFD. In detail, their report revealed the specimen to have retained nodal architecture with extensive patchy areas of necrosis. Surrounding the areas of necrosis, the cellular elements were composed by numerous histiocytes, small lymphocytes, and a few plasma cells with no neutrophils or eosinophils present. In addition, the histiocytes present in the specimen featured "crescentic" nuclei.

Additional immunohistochemical stains were performed. The majority of T-cells expressed CD-8, rather than CD-4. Numerous histiocytes stained positive for CD-68. Furthermore, the histiocytes expressed CD-163, as well as myeloperoxidase. Finally, CD-123 immunostain highlighted plasmacytoid dendritic cells predominantly located around necrotic areas. Flow-cytometry studies were also performed, showing a decreased CD4/CD8 ratio and no definitive evidence of monoclonal B-cell population or aberrant T-cells. These findings showed a benign process, consistent with KFD lymphadenitis.

The patient's cervical lymphadenopathy resolved in 2 months without complication. On recent follow-up via phone call in July 2018, the patient continues to be asymptomatic with no development of any signs and symptoms consistent with SLE.

Discussion

Kikuchi-Fujimoto Disease is an uncommon cause of lymphadenopathy in the United States. Localized lymphadenopathy is the most common symptom associated with KFD.[3] In addition to cervical lymphadenopathy, axillary and supraclavicular lymphadenopathy has also been reported.[13,14] Usually, the lymphadenopathy associated with KFD is <3 cm, but there have been reports of lymph nodes reaching 5 to 6 cm.[5] Regarding fever, reports have demonstrated that the duration may range from 1 to 7 weeks with the temperature ranging from 38.6°C to 40.5°C.[6] Tenderness with palpation of the lymph node may or may not be present. There may be a number of other associated signs and symptoms, which include chills, weight loss, headache, fatigue/malaise, vomiting, night sweats, arthralgia, splenomegaly, and rash. It has been reported that up to 30% of patients with KFD will have some sort of skin manifestation of the disease, which may include acneiform eruptions, facial erythema, papules, plaques, purpura, or nodules. When skin manifestations are present, it is indicative of a more severe clinical presentation.[3]

The differential diagnosis of a neck mass/cervical lymphadenopathy is broad and must include infectious, congenital, inflammatory, neoplastic, and vascular causes. *Staphylococcus aureus* and Group A Beta-Hemolytic *Streptococcus* (*S. pyogenes*) are the most common bacterial causes of suppurative lymphadenitis. Cat-scratch disease, caused by *Bartonella henselae*, must also be considered. Viral lymphadenopathy is also very common and can be caused by numerous viruses, including but not limited to EBV, cytomegalovirus (CMV), human immunodeficiency virus (HIV), rubella, rhinovirus, and adenovirus. Other

infectious causes of lymphadenitis should also be considered and include both typical and atypical mycobacterium, toxoplasmosis, and various fungal infections.

Serologic testing can be used to help diagnose infectious mononucleosis (caused by EBV), cat-scratch disease, and toxoplasmosis. A purified protein derivative (PPD) skin test may help aid the diagnosis of Mycobacterium tuberculosis (TB) (*Mycobacterium tuberculosis*). However, if atypical tuberculosis is suspected, diagnosis is usually made with biopsy/curettage.[7]

Various congenital disorders, including branchial cleft anomalies, thyroglossal duct cysts, and dermoids, may also present with a neck mass and must be entertained in the differential diagnosis. Inflammatory conditions or autoimmune disorders that may manifest similarly include Kawasaki disease, sarcoidosis, Rosai-Dorfman disease, and SLE. Finally, neoplastic etiologies must also be considered, such as cervical metastatic disease of an upper aerodigestive tract primary or lymphoma.

There are no specific labs that are specific for the diagnosis of KFD. Lab studies have been reported to show a wide variety of results, including an increased lactate dehydrogenase (LDH), leukopenia or leukocytosis, anemia, increased erythrocyte sedimentation rate, increased C-reactive protein, and elevated transaminases.[3,6] The literature has reported that leukopenia is present in anywhere from 25% to 58%, and leukocytosis is present in approximately 2% to 5% of patients with KFD.[3] Workup should include imaging with Ultrasound (US) and/or CT. Definitive diagnosis is obtained with excisional biopsy and histopathological examination.

Histologically, KFD is characterized by partially preserved nodal architecture with intermittent areas of fibrinoid necrosis and apoptosis, surrounded by histiocytes (with crescentic nuclei), activated T-lymphocytes, and plasmacytoid monocytes. The crescentic histiocytes are normally found in the necrotic foci with karyorrhectic debris. Characteristically, there is a paucity of neutrophils and eosinophils.[3,7,10,12,15] It is important to understand KFD shares similar histopathologic features with other important diagnoses, including lymphoma, SLE, TB, and infectious mononucleosis. However, there are differences, which prove helpful in distinguishing these entities.

In lymphoma, necrosis may not be as severe, and neutrophils and granulomata are usually absent. In addition, SLE is usually associated with the presence of hematoxylin bodies, which are particles of denatured nuclear material. Immunohistochemical staining is also helpful in distinguishing KFD and increasing the specificity of diagnosis. In KFD, there are a large number of CD8-positive lymphocytes, as well as large numbers of CD68-positive histiocytes. This is in contrast to large B-cell lymphoma, whereas the neoplastic cells stain CD20-positive.[3,7] Also, the histiocytes found in KFD typically express myeloperoxidase.[15]

The exact pathophysiology of KFD is unknown, but there are 2 theories that have been proposed. It has been hypothesized that KFD may result as a reaction to a viral infection, or it may be the manifestation of an autoimmune disease.[3,7,10] Support for the viral etiology is provided by the dramatic presence of histiocytes and CD8-positive lymphocytes in KFD affected lymph nodes. There have been numerous studies that have tried to demonstrate an association between KFD and various viruses. In a study by Cho et al,[16] 50 lymph node specimens diagnosed with KFD were analyzed using polymerase chain reaction (PCR) for the presence of Human Herpes Virus (HHV)-6, -7, and -8. This study failed to show an association between KFD and HHV-6, -7, or -8. However, another study performed by Zhang et al[17] did identify an association between parvovirus B19 and KFD. Hudnall et al examined 30 lymph nodes affected by KFD and showed that HHV-1, varicella zoster virus, and HHV-8 DNA were not detectable, and HHV-2, CMV, HHV-6, and HHV-7 were detected on occasion. Hudnall et al's[18] study concluded that it was unlikely that these viruses served as the etiology of KFD.

The other proposed theory is that KFD may be associated with an autoimmune disorder, mainly SLE. There have been many case reports and studies that demonstrate this possible link, but the association is not fully understood.[4] Lymphadenopathy, fever, fatigue, malaise, weight loss, and skin manifestations are clinical manifestations that may be present in both KFD and SLE. In 2007, a study was performed which evaluated 244 cases of KFD worldwide, from 1991 to 2005. From their results, 32 of these 244 patients (13%) also had a diagnosis of SLE.[5] More recently, in 2016, Baenas et al performed a MEDLINE/PubMed search through 2015 and identified 3 patterns of association between KFD and SLE. In their review, 30% of patients were diagnosed with KFD prior to the onset of SLE. In 47% of patients, there was simultaneous occurrence/diagnosis of both KFD and SLE. Finally, in 23% of patients, KFD was diagnosed after a diagnosis of SLE was already known and established.[19] A case series published by Goldblatt described 3 Asian females with KFD. At the time of diagnosis of KFD, Antinuclear Antibodies (ANA) testing was negative for all 3 patients and none experienced any signs and symptoms consistent with SLE. However, within a 3- to 14-month time period following the diagnosis of KFD, all 3 patients developed a positive ANA result and symptoms of SLE.[20] There is published data confirming the association between KFD and SLE, but the exact relationship is still unclear. Given the above evidence, it may be wise to follow patients diagnosed with KFD and consider an autoimmune workup or rheumatology referral.[10,19]

As mentioned earlier, KFD is classically believed to be a self-limited, benign condition that typically resolves within 6 months. An estimated 3% to 4% of patients may experience relapse.[3] Treatment is aimed at symptomatic control, and Non-Steroidal Anti-Inflammatories (NSAIDS) have been recommended for lymph node tenderness or febrile illness. If severe, glucocorticoids have been recommended, but there is no consensus on dosing or duration.[21,22] A review published in 2000 discusses the use of glucocorticoid use for cases refractory to standard

treatment with Aspirin (ASA) or NSAIDS. This article presents 3 cases of KFD where symptoms of tender lymphadenopathy and fever were not adequately controlled with NSAIDS. Glucocorticoid use was given in doses ranging from 30 to 60 mg/day for 3 to 5 days with tapering doses over 7 to 10 days. All 3 cases reported demonstrated significant improvement after glucocorticoid use.[22] Hydroxychloroquine has also been described as a possible treatment.[23]

However, there are also rare case reports in the literature that demonstrate an association with KFD and more aggressive and life-threatening outcomes.[24-29] Recently, in 2017, a case report described a 21-year-old, otherwise healthy female, who presented to the Emergency Department (ED) with a 2-day history of dyspnea, fever, and malaise. A CT of the chest, abdomen, and pelvis was performed, which demonstrated significant cervical and axillary lymphadenopathy, lung consolidation, and pericardial effusion. Cervical lymph node biopsy was performed, and histology was consistent with KFD. Despite aggressive Intravenous (IV) antibiotics and high-dose steroids, this patient's clinical course declined, eventually passing away due to the development of disseminated intravascular coagulation (DIC).[24] In addition to the case report described above, there have been 3 other case reports published which cite DIC as the cause of death in patients also diagnosed with KFD.[25,26] The association between DIC and KFD is not yet understood, and little is known how or if at all KFD plays a role in the development of DIC.[24] Apart from DIC as the cause of mortality, there have been other causes of death reported in patients with KFD, which include hemophagocytic syndrome and severe infection,[27] pulmonary hemorrhage,[28] and acute heart failure.[29] It is imperative to understand that these case reports do not imply a causal relationship between KFD and the development of DIC or other life-threatening conditions mentioned above, but rather an association. The pathophysiologic mechanisms are not understood, and further research is needed before any conclusions can be discerned.

Conclusion

Kikuchi-Fujimoto Disease is a rare disease that typically follows a benign and self-limiting course. It manifests classically with lymphadenopathy and fevers, but may be associated with a number of other symptoms. It should be considered in the differential diagnosis of patients presenting with persistent lymphadenopathy. The precise pathophysiology of this disease is unknown, but it is hypothesized the etiology may be post-viral or associated with an autoimmune disease, specifically SLE. Therefore, some thought should be given to follow patients and consider an autoimmune workup. Treatment is symptomatic, but if severe, corticosteroids may be considered. Although symptoms usually resolve within 6 months, there are reports of KFD being associated with poor outcomes.

Here is reported a patient who presented to a private Otolaryngology practice and was diagnosed with KFD. The purpose of this article is not only to review the literature but also to highlight and bring attention to the possibility of KFD in the differential of a neck mass or cervical lymphadenopathy in the adult or pediatric patient. This is especially important for the general Otolaryngologist practicing in a community setting, rather than a tertiary center where most of the previous cases have been reported.

It is also the aim of this review to popularize this clinicopathologic disease among other medical subspecialists, including Pediatricians, Infectious Disease Physicians, Rheumatologists, Pathologists, and Medical Oncologists. As discussed previously, the diagnosis of KFD requires a surgical specimen and an accurate histopathological analysis. It absolutely must be distinguished from lymphoma, as recommendations regarding treatment of these entities are vastly different. All of the above-mentioned specialists may play a significant role in establishing this diagnosis and providing treatment, and thus this review seeks to help educate and provide a resource to meet this goal.

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Author Contributions

JMS is the corresponding author and was the resident physician involved in this case. He is the author that comprehensively performed the literature review, compiled all of the information, and wrote the article. Also, he was the author primarily involved in the submission of this manuscript. CBS was the attending physician responsible for this patient. This patient was initially seen at North Oakland ENT, a community-based private practice ENT specialty group managed by CBS. Both authors, JMS and CBS, read and reviewed the manuscript prior to submission.

Availability of data and materials

All the data generated or analyzed during this study are included in this published article.

Ethical approval

The McLaren Health Care Institutional Review Board (MHC IRB) waived ethics approval.

Informed consent

Written consent was obtained to include details, images, or videos relating to an individual person. The patient's mother obtained the written consent.

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