Case Report

Bilateral painful parotid lumps and a lump in the groin: An uncommon presentation of common Kikuchi's disease

Sumeet Prakash Mirgh[1], Jinendra Satiya[2], Jehangir Soli Sorabjee[1]

[1]Department of Medicine, Bombay Hospital Institute of Medical Sciences, [2]M.B.B.S. Grant Government Medical College and Sir. J.J. Group of Hospitals, Mumbai, Maharashtra, India

Abstract

Kikuchi-Fujimoto disease (KFD) is an under-recognized disease most commonly presenting with cervical lymphadenopathy, fever, and cytopenias in young females. Bilateral parotid enlargement is usually caused by infections (e.g., mumps) and autoimmune conditions (e.g., Sjogren syndrome). Parotid enlargement, inguinal lymphadenopathy, and pyrexia of unknown origin are uncommon presenting features of KFD and should be suspected in the appropriate setting.

Keywords: Inguinal, Kikuchi-Fujimoto disease, lymphadenopathy, parotid

Introduction

We report a case of a 31-year-old female presenting with bilateral painful parotitis, a lump in the right groin, and prolonged pyrexia who was diagnosed with Kikuchi-Fujimoto disease (KFD) on lymph node biopsy. Very few cases of KFD presenting with bilateral painful parotid swelling[1,2] and inguinal lymphadenopathy[3,4] have been reported in the literature. Our case illustrates the importance of diagnosing Kikuchi's disease in a patient presenting with bilateral parotitis and pyrexia of unknown origin (PUO), which is an unusual presentation of the disease.

Case Report

A 31-year-old female with a history of travel to Malaysia 6 weeks prior presented with a 3-week history of high-grade fever with chills. She reported daily 38.3°C fever without diurnal variation. She had a history of watery, nonbloody diarrhea 3 days prior and occasional cough. She denied any history of breathlessness, chest pain, diabetes mellitus, or hypertension.

On examination, her general condition and vital signs were normal. The patient had prominent bilateral parotid swelling measuring roughly 5 by 4 cm which was soft, tender and nonfluctuating [Figure 1], and mild right inguinal lymphadenopathy. Rest of the systemic examination was normal.

The patient was investigated as shown in Table 1. Viral markers and multiple blood cultures were negative. In view of clinical features of persistent fever with leukopenia, serological tests for *Cytomegalovirus* IgM antibody and Epstein–Barr virus IgM were done, which were also negative.

The patient was started on injection amoxicillin-clavulanate (dose 1.2 g twice a day) and symptomatic treatment. A computerized tomogram (CT) scan (chest and abdomen) showed multiple mildly enlarged nonenhancing, nonnecrotic discrete lymph nodes in the sub-mental and bilateral deep cervical regions and upper

Address for correspondence: Dr. Sumeet Prakash Mirgh, No. 401, Jai Gurudev C.H.S, Plot No. 6, Sector No. 1, Sanpada, Navi Mumbai - 400 705, Maharashtra, India.
E-mail: drsumeetmirgh@gmail.com

Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/2249-4863.192378

How to cite this article: Mirgh SP, Satiya J, Sorabjee JS. Bilateral painful parotid lumps and a lump in the groin: An uncommon presentation of common Kikuchi's disease. J Family Med Prim Care 2016;5:465-7.

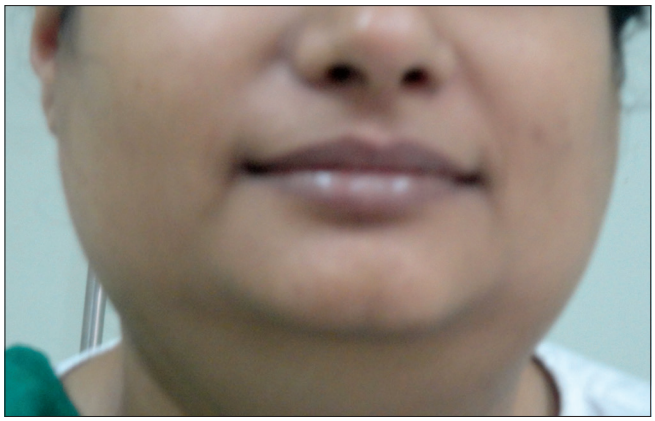

Figure 1: Bilateral parotid swellings

abdominal lymphadenopathy, suggestive of a granulomatous etiology. A biopsy of the cervical lymph node showed brownish fragment of 2 cm on gross examination and histiocytic necrotizing lymphadenitis with no evidence of acid-fast *Bacillus* or malignancy on microscopy. Tuberculosis (TB) mycobacterial growth indicator tube and gene expert *Mycobacterium tuberculosis* RIF done from the lymph node were negative.

Table 1: Routine investigations for pyrexia of unknown origin on admission

Investigations (normal values with conventional units)	Patient's values
Hemoglobin (14-17.5 g/dL)	9.40
Mean corpuscular volume (80-100 fL)	80.9
Mean corpuscular hemoglobin (27-31 pg)	27.5
Total leukocyte count (4.4-11.3×10^3/mm^3)	1400
Erythrocyte sedimentation rate (0-15 mm/h)	54
Lactate dehydrogenase (140-280 mU/ml)	913
Serum sodium (135-145 mEq/L)	130
Serum potassium (3.5-5.5 mEq/L)	4.0
SGOT† (12-38 U/L)	187
SGPT‡ (7-41 U/L)	59
Random blood sugar (<200 mg %)	85
Ferritin (0.3-1.3 mg/dl)	971.5
Calcium (9-10.5 mg/dl)	8.0
Plasma fibrinogen (150-400 mg/dl)	244.8
HIV ELISA	Negative
Dengue IgM	Negative
Blood culture	Negative
Peripheral blood smear - Mild hypochromia, anisocytosis, occasional microcytes, and elliptocytes	-

†SGOT: Serum glutamate oxaloacetate transaminase; ‡SGPT: Serum glutamate pyruvate transaminase

Discussion

KFD or histiocytic necrotizing lymphadenitis was first described by Kikuchi and Fujimoto in Japan in 1972. It typically affects young adults (mean age, 20–30 years). Women are affected more often than men, with a ratio of 3:1.[1]

The most common clinical manifestation is cervical lymphadenopathy (80%). Lymph nodes are usually described as mildly tender, firm, and 2–3 cm in diameter. A flu-like prodrome with fever is present in 50% of the cases.[1] Less common symptoms include headache, malaise, weight loss, arthralgias, and night sweats. Parotidomegaly, as seen in our patient, may be present, but is uncommon. Cutaneous involvement is observed in 30–40% of cases. There have been fatal reports of myocarditis, pulmonary hemorrhage, and disseminated disease.

Our case was unusual as our patient presented primarily with bilateral massive parotid enlargement and right inguinal lymphadenopathy on a background of PUO. Very few cases have reported the presentation of KFD with parotid involvement[5] and inguinal lymph node involvement.[6,7] KFD can masquerade as a single, rapidly-growing, parotid tumor in a middle-aged male. It has presented as PUO in a 9-year-old boy[8] as well as a 72-year-old male.[9] In a series of 21 patients of KFD evaluated by positron emission tomography-CT, most commonly involved lymph nodes were cervical followed by axillary whereas inguinal nodes were least commonly involved.[10]

The differential diagnosis of bilateral parotid enlargement includes infectious parotitis, chronic autoimmune parotitis (Mikulicz disease, Sjogren syndrome, and lymphoepithelial lesion of Godwin), and idiopathic causes (such as sarcoidosis and chronic nonspecific parotitis). Patients with HIV-parotitis and sarcoidosis may be asymptomatic. Sjögren syndrome presents as recurrent or chronic swelling of one or both parotid glands with no apparent cause, frequently associated with other autoimmune diseases.[2]

A close mimic of KFD is TB. In any patient from a TB endemic area such as India, presenting with classical clinical features of fever, weight loss, and lymphadenopathy, one should wait for a 6-week culture before making a diagnosis.[3] KFD is a rare self-limiting cervical lymphadenitis, often a diagnosis of exclusion. It is important for pathologists and clinicians to be aware of this possibility.

Supportive measures, including nonsteroidal anti-inflammatory drugs (NSAIDs) and antipyretics, can be used for symptomatic relief. Corticosteroids are reserved for severe cases or intractable symptoms. Immunosuppressive agents (hydroxychloroquine, cyclosporine, and azathioprine) have been used successfully. The course of the illness is usually 1–3 months, but longer follow-up may be appropriate as a 4% relapse rate has been reported. Monitoring for the development of systemic lupus erythematosus (SLE) is necessary. Case studies have described the use of minocycline and ciprofloxacin supporting an infectious cause. These, however, were the results of single case reports with no follow-up studies. Recognizing the significant limitations of the data, antibiotics or antivirals are not recommended. A steroid regimen is useful as it is effective in short durations. There is a single case report describing intravenous immunoglobulin treatment (0.4 g/kg per day for 3 days) in a patient with severe disease.[4]

Our patient was given oral steroids (oral prednisolone 10 mg thrice a day for 7 days followed by a taper over next 2 weeks) in view of persistent malaise and low-grade fever despite being

on NSAIDs. Her fever and malaise improved within 72 h. Over a course of 4 weeks, her parotid enlargement decreased and leucopenia normalized. At 6 months' follow-up, she is stable without any signs of relapse or progression to SLE.

Conclusion

KFD is an under-diagnosed, yet important cause of fever and lymphadenopathy in the young population. Posterior cervical lymph nodes are the most commonly involved. Biopsy is recommended to establish a definitive diagnosis and also to rule out other more sinister causes such as malignancy, lymphoma, and TB. It is highly responsive to treatment and rarely recurs. Steroids are useful in cases not responding to NSAIDs.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Chaitanya BN, Sindura C. Kikuchi's disease. J Oral Maxillofac Pathol 2010;14:6-9.
2. Chen JJ, Li YS, Chen TA. Kikuchi disease manifesting as a parotid tumor - Case report. Tzu Chi Med J 2006;18:442-4.
3. Wilson MR, Milne G, Vryonis E. Kikuchi-fujimoto disease: A rare cause of fever in the returning traveller. Case Rep Med 2014;2014:868190.
4. Mahmood A, Mir R, Salama SR, Miarrostami RM, Lapidus C, Pujol F. Kikuchi's disease: An unusual presentation and a therapeutic challenge. Yale J Biol Med 2006;79:27-33.
5. Kuo T, Jung SM, Wu WJ. Kikuchi's disease of intraparotid lymph nodes presenting as a parotid gland tumour with extranodal involvement of the salivary gland. Histopathology 1996;28:185-7.
6. Shah S, Vijendren A, Obichere M. Kikuchi – Fujimoto disease: A rare presentation of a groin lump. BMJ Case Rep 2013;2013. pii: Bcr2012006841.
7. Bernasconi C, Rusca T, Balestra B. An "exotic" lymphadenopathy: Kikuchi-Fujimotor histiocytic necrotizing lymphadenitis. Case report and discussion. Praxis (Bern 1994) 2000;89:671-4.
8. de Gaudio M, Moshal K, Malone M, Novelli V. Kikuchi-Fujimoto disease causing fever of unknown origin in a nine-year-old boy. Scand J Infect Dis 2010;42:946-9.
9. Fitzsimmons PR, Akpan A, Michael B. Kikuchi-Fujimoto disease as a rare cause of fever of unknown origin in a septuagenarian. Age Ageing 2008;37:233-4.
10. Kong E, Chun K, Hong Y, Hah J, Cho I. 18F-FDG PET/CT findings in patients with Kikuchi disease. Nuklearmedizin 2013;52:101-6.