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A Case Report on Kikuchi - Fujimoto disease

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ABSTRACT:
Kikuchi - Fujimoto disease is a rare disorder mostly affecting Asian females presenting systemic symptoms and cervical lymphadenopathy. It is also known as Kikuchi's disease or benign histiocytic nectrotizing lymphadenitis. The main complications of this disease include peripheral neuropathy, hepatitis, panuveitis that affect the eye , meningitis, aseptic encephalitis and cerebellar ataxia affecting the CNS.NSAID are prescribed for lymph node tenderness and corticosteroids are prescribed for extra nodal symptoms. For steroid resistant KD, iv immunoglobulins are given. A 29year old male patient was admitted with the complaints ofnocturnal fever with chills, recurrent migraine and painful oral ulcers, abdominal pain after meal with nausea along with diffused alopecia and bloody stools. He also presented disturbed sleep patterns, episodic mild rashes on left hand and leg that lasted a day. He also suffered early morning stiffness that lasted for about 15 mins especially in the left shoulder. On examination, He had left cervical lymph node enlargement. The USG of neck reported cervical adenopathy and FNAC of cervical lymph node showed reactive changes with histiocytic collection, without granuloma/ caseous necrosis/ malignant cells. On lab examination, ESR 50,TC 10500, Ca 9.5, UA 6.2, Hb 9.2, and MTB undetected. The biopsy report revealed necrotising lymphadenitis that confirmed Kikuchi disease. He was initially started with corticosteroids that were tapered as symptoms subsided. But the pain advanced as the dose was tapered and vice versa. The treatment regimen included Methylprednisolone 4mg twice daily along with a combination of Pregabalin and Mecobalamin.

KEYWORDS: Kikuchi -Fujimotodisease ,Lymphadenitis

I. INTRODUCTION:

Kikuchi - Fujimoto disease is a rare disorder mostly affecting Asian females presenting systemic symptoms and cervical lymphadenopathy. It is also known as Kikuchi's disease or benign histiocytic nectrotizing lymphadenitis. Although it is observed worldwide, Asians are mostly affected[1]. KD is a disorder that subsides on it's own. A long-term periodic follow up and monitoring should be conducted for patients with KD as they are at an increased risk of developing cutaneous and/or systemic lupus erythematosus. This is especially seen in patients with cutaneous KD. Males are least affected than females with a male to female ratio of 1:4. A cardinal feature of this condition is it's low morbidity and mortality rate, as though the painful suffering is agonizing. Moreover, this rate advances rapidly when the condition progresses to SLE[2].

Kikuchi's disease is an autoimmune disorder that usually appear post infection. Although the etiology of KD is unknown, it is presumed to be due to post infectious processes caused by Parvovirus 19, hepatitis B, HTLV-1 and toxoplasma[3].

The clinical manifestations of KD is typically of two types; Nodal and Extra nodal. Nodal findings comprise tender unilateral cervical lymphadenopathy with node size varying between 0.5 and 4cm. Lymphadenopathy located at a single site is a prominent feature of KD which is observed in 83% of patients. However, they may also exhibit as multiple nodal chains. They rather appear mobile and firm than fluctuant and draining[4].

The extra nodal features detected include low grade fever, upper respiratory symptoms and rarely hepatosplenomegaly. Whereas, involvement of skin, bone marrow and liver are rare. The distinguished extra nodal indications resemble the symptoms of SLE that include maculopapular lesions[5]. Morbilliform rash, nodules, urticarial and malar rash. These may resolve within few weeks to months.

Seldom, extranodal symptoms due to neurological involvement are observed at bone marrow, myocardium, uvea, thyroid and parotid

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glands[6]. Furthermore, asymmetric polyarthritis, enthesitis and dactylitis of the toes are the extranodal arthritic features.

There are no distinguishing evidences for diagnosing Kikuchi's disease as CT and MRI scans appear inconsistent. On analysis, these necrotic nodes may seem identical to metastatic carcinomas, lymphomas or tuberculosis. Polymorphous lymphoid cells with karyorrhectic debris, histiocytes and a smallsize nuclei are the cytological features that appear on conducting FNAC. These features are characteristics of diagnosing KD. Moreover, a necrotizing lymphadenitis in the presence of karyorrhexis and without granulocytes in lymph node biopsy is peculiar of KD[7].

Certain conditions such a systemic lupus erythematosus and lymphoma are the major differentially diagnosed disorders with KD along with infectious lymphadenitis[8,9,10].

The complications of KD is profuse that occur rarely and frequently. Such rare complications include peripheral neuropathy[11], hepatitis[12], panuveitis that affect the eye and treated with methotrexate[13]. Other complications include meningitis, aseptic encephalitis and cerebellar ataxia affecting the CNS[14]. Cardiac complications include Tamponade. Although its affect on mesenteric lymph nodes is rare, there are reports of axillary lymphadenopathy and isolated mediastinal lymphadenopathy[15]. Acute renal failure[16], interstitial lung disease and pleural effusion[17] are the other minor complications.

As the treatment regimen is initated, the symptoms subside within 1 to 4 months

The treatment is likely similar to other auto immune conditions.It is rarely fatal. As the lymph node involvement is major, NSAIDs are prescribed to treat lymph node tenderness and fever, Forextranodal symptoms, corticosteroids such as prednisone is recommended[18]. Whereas, hydroxychloroquine and iv immunoglobulin are recommended for steroid-resistant and recurrent Kikuchi disease[19]. Sometimes, antipyretics and analgesics are also recommended for fever and pain management[20].

II. CASE DESCRIPTION:

A 29 years old male patient presented with the chief complaints of nocturnal fever with chills,recurrent migraine and painful oral ulcers, abdominal pain after meal with nausea along with diffused alopecia and bloody stools. These symptoms persisted for over 2 months prior to admission. He also presented disturbed sleep patterns, episodic mild rashes on left hand and leg that lasted a day. He was unable to move his head as he experienced occipital headache that radiated to trapezius disturbing the sleep. He has been experiencing discharge from right ear since a day prior to admission. He also suffered early morning stiffness that lasted for about 15 mins especially in the left shoulder. On examination, He had left cervical lymph node enlargement.The USG of neck reported cervical adenopathy and FNAC of cervical lymph node showed reactive changes with histiocytic collection, without granuloma/ caseous necrosis/ malignant cells. The ANA and DsDNA were negative. On lab examination, ESR50,TC10500,Ca9.5,UA6.2, Hb9.2,and MTB undetected. Sigmoidoscopy indicated internal haemorrhoids and CT of PNS revealed mild mucosal thickening in ethmoid sinuses suggesting of sinusitis. X ray of thoracolumbar spine revealed scoliosis. The biopsy report revealed necrotising lymphadenitis that confirmed Kikuchi disease. He was initially started with corticosteroids that were tapered as symptoms subsided. But the pain advanced as the dose was tapered and vice versa. The treatment regimen included Methylprednisolone 4 mg twice daily along with a combination of Pregabalin and Mecobalamin.

III. DISCUSSION:

Kikuchi-Fujimoto disease is a self -limited condition that is benign and rare. This condition was first reported in 1972 in Japan by Kikuchi and Fujimoto.et al. Asian women are mostly affected. Lymph node involvement is mostly observed with cervical lymph node enlargement, although cervical lymphadenopathy is a potential differential diagnosis.

In this case, the patient experienced cervical lymph node enlargement along with fever, headache, mild rashes and shoulder pain. A study was done involving 1724 subjects in India by Mohan et.al. This study reported 35.6% subjects with non-specific lymphadenitis, 31.3% with tuberculous lymphadenitis, 25.9% with malignancy and <1% diagnosed with Kikuchi-Fujimoto disease[21].

IV. CONCLUSION:

Kikuchi's disease is a rare auto immune disorder that resolves gradually. A long term periodic follow up and monitoring should be

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conducted inorder to decrease the occurrence of cutaneous or systemic lupus erythematosus .NSAID are prescribed for lymph node tenderness and corticosteroids are prescribed for extra nodal symptoms . For steroid resistant KD, iv immunoglobulins are given. Inorder to decrease the potentially harmful events of this disease , clinicians should be aware of this condition even though the occurrence of disease is rare .

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