CASE REPORT

Case Report: An unusual clinical presentation of Kikuchi's disease: a case report [version 1; peer review: 1 not approved]

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Abstract

Introduction

Kikuchi's disease is usually a benign condition presenting with fever and lymphadenopathy. Rarely, the severe form of this disease can have an unusual presentation.

Case Report

We report a case of 9 year old girl, a resident of Kerala (South India) with no significant past medical history, who presented with fever of 8 days with facial puffiness. She had tender cervical lymphadenopathy. Two days after admission, her condition worsened with venous congestion of the face and neck, throbbing headache, dyspnea and she developed seizures. Very soon, she developed pericarditis with cardiogenic shock and required adrenaline support. Lymph node biopsy confirmed Kikuchi's disease. The possibility of superior vena cava (SVC) obstruction due to mediastinal lymphadenopathy was considered and treated with steroids following which the child showed a dramatic improvement. The child also had four of the American Rheumatism Association (ARA) diagnostic criteria for systemic lupus erythematosus (SLE) (viz., serositis, seizures, hematological abnormalities and antinuclear antibodies (ANA) positivity.

Conclusion

Although Kikuchi's disease is a self-limiting condition, it is important to consider this possibility in any patient who fits the typical clinical scenario to avoid unnecessary workup. An unusually severe form of Kikuchi's disease can have a presentation like SVC obstruction syndrome due to enlarged mediastinal lymph nodes. Hence, timely recognition of this condition and prompt institution of steroid therapy will result in a dramatic clinical response.

Keywords

blood, lymph nodes, fevers

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Any reports and responses or comments on the article can be found at the end of the article.

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Introduction

Kikuchi's disease is a rare disease that was originally reported in patients of Asian heritage. It usually occurs in 8–16 year old aged children and presents with fever accompanied with lymphadenopathy especially in posterior cervical region. The diagnosis is made through lymph node biopsy¹. Here we report a case of superior vena cava obstruction as an unusual presentation of Kikuchi's disease in a 9 year old Indian girl.

Case history

A 9 year old girl presented with fever of 8 days duration with poor appetite, nausea and occasional headache. She was a previously healthy child with no significant past medical or family history. She had facial puffiness and swellings in the neck noted two days previously. There was no history of decreased urine output, vomiting, rash, arthralgia, breathlessness, skin or mucosal bleeds, significant weight loss or contact with a case of tuberculosis. On physical examination, she was febrile (102°F) and had marked periorbital edema, along with multiple, enlarged and tender lymph nodes in the posterior cervical region with the largest node measuring 3 cm. There was no rash, bleeding manifestations or enlargement of other groups of lymph nodes. Examination of the gastrointestinal, respiratory, cardiovascular and nervous systems was normal. Investigations revealed the following: hemoglobin 10.1 g/dL, white blood cell (WBC) count 2760 cells/mm (neutrophils-55%, lymphocytes-41%) with platelet count of 82,000 cells/cu.mm, erythrocyte sedimentation rate (ESR)–50 mm/hr and C-reactive protein (CRP)–44 mg/dL. Renal and liver function tests were normal. Blood Widal and Dengue NS1 antigen tests were negative. She was started on cefotaxim (1 g intravenously q8 hourly). The patient's condition worsened on the second day of admission with progressive dyspnea and increasing facial puffiness and she also complained of a throbbing headache. She had venous congestion of the head and neck region with conjunctival congestion. Since chest X-ray showed mediastinal widening, the possibility of enlarged mediastinal lymph nodes causing SVC obstruction was considered and she was scheduled for a CT chest examination but she developed multiple seizures at that time. Repeat investigations on the same day (day 2 of admission) showed a fall in hemoglobin (9.6 g/dL) and platelets (67,000 cells/mm³) from the baseline. Lumbar puncture findings were normal. Two hours later, she also developed pericarditis with pericardial effusion, went into cardiogenic shock and required adrenaline support (0.5 mcg/Kg/min continuous intravenous infusion). A cervical lymph node biopsy done showed necrosis with karyorrhexis, histiocytic infiltrates, crescentic plasmacytoid monocytes, and absence of neutrophils – a picture consistent with Kikuchi-Fujimoto disease. Her blood, CSF, urine cultures were sterile. Bone marrow aspiration studies were normal. Investigation also showed ANA-IF – 3+ positivity and anti-dsDNA was strongly positive. Epstein-Barr virus (EBV) DNA PCR was negative. In view of the child fulfilling four of the ARA criteria for systemic lupus erythematosus (SLE) (viz., serositis, seizures, hematological abnormalities and ANA positivity)⁸, a diagnosis of SLE with Kikuchi disease presenting as superior vena cava syndrome was made and was treated with intravenous methylprednisolone (600 mg over 30 minutes) for 3 days. The

patient showed a dramatic improvement with treatment, both clinically as well as with laboratory parameters. Her general condition improved, congestion of the face and neck decreased, the lymph nodes decreased in size and she became afebrile and remained so thereafter. After 3 days of intravenous methylprednisolone, she was changed over to oral prednisolone (1 mg/Kg/day) and oral hydroxychloroquine (5 mg/Kg/day). She was discharged after 1 week with advice to continue prednisolone and hydroxychloroquine (at the same dose). At the time of follow up (after 2 weeks) she remained asymptomatic, blood count and disease activity were normal and she did not develop any features of steroid toxicity and had no evidence of adrenal insufficiency.

Discussion

To the author's knowledge, this is the first report of SVC obstruction as a clinical presentation of Kikuchi's disease (due to enlarged mediastinal lymph nodes).

The etiology of Kikuchi's disease is not entirely known. It has been linked to sequelae of infection by human herpes virus 6 (HHV-6), cytomegalovirus (CMV), EBV and even human T lymphotropic virus 1 (HTLV-1)^{2–4}. Serological tests conducted with our patient ruled out the possibility of an associated infection. The most common clinical presentation of Kikuchi's disease is fever and cervical lymphadenopathy in a previously healthy young woman⁵. In a literature review describing 244 patients affected by Kikuchi's disease, the common presentations were fever, fatigue, and joint pain with cervical lymphadenopathy, leucopenia, elevated ESR, and anemia⁶. Our patient also had prolonged fever with constitutional symptoms and cervical lymphadenopathy with raised ESR.

Our patient also presented with features of SLE (she fulfilled four of the ARA criteria for SLE viz., serositis, seizures, hematological abnormalities and ANA positivity). According to Kucukardali *et al.*⁶, SLE-associated Kikuchi's disease is more common in cases from Asian countries than from Europe. Among the 28 cases studied by Kucukardali *et al.*, 18 cases presented with both SLE and Kikuchi's disease diagnosed simultaneously, 6 cases were diagnosed with SLE after being diagnosed with Kikuchi's disease and 4 were previously diagnosed with SLE⁶. The clinical features of SLE and Kikuchi's disease are very similar and definite discrimination between them is based on histopathological findings⁷. The absence of hematoxylin bodies and paucity of neutrophils indicate Kikuchi's disease rather than SLE and our patient showed definite features of Kikuchi's disease in the lymph node biopsy.

Conclusion

Kikuchi's disease should be considered in any child, especially from Asian heritage, who presents with the typical clinical features, as prompt diagnosis of this condition will avoid further unnecessary investigations. An unusually severe form of Kikuchi's disease can have a presentation similar to SVC obstruction syndrome due to enlarged mediastinal lymph nodes. Hence timely recognition of this condition and prompt institution of steroid therapy will result in a dramatic clinical response and a life saving measure.

Consent

Written informed consent for publication of clinical details was obtained from the child's parents.

Author contributions

Dr. Singaram A. wrote the manuscript. Dr. Ramesh Menon P. revised the first draft. Dr. Cherian N.C. and Dr. Geetha .P. managed the patient in the pediatric ward. Dr. Rajesh T.V. managed the patient in the intensive care unit. All authors were involved in the revision of the manuscript and have agreed to the final content.

Competing interests

No competing interests were disclosed.

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The histology of the lymph node in Kikuchi's disease can usually be easily differentiated from most known infectious conditions in the differential diagnosis of lymphadenopathy, but not from that seen in some patients with systemic lupus erythematosus (SLE). Even if we accept the histological diagnosis based on lymph node pathology, this does not necessarily explain the whole clinical picture if other more probable diagnoses have not been ruled out.

Beyond the fact that the child fulfilled four of the American Rheumatism Association (ARA) diagnostic criteria for systemic lupus erythematosus (SLE), they tested positive for anti-double-stranded DNA (dsDNA) antibodies, which are highly specific for SLE.

The diagnosis of superior vena cava syndrome was based on clinical manifestations (dyspnea, facial puffiness, throbbing headache and venous congestion of the head and neck region with conjunctival congestion). However, chest CT was not performed. Considering the fact that the child was definitely suffering by cardiac tamponade, which could also be manifested with dyspnea and congestion of face veins, I am not persuaded about the co-existence of SVC obstruction. In this specific clinical text this diagnosis should have been confirmed by chest CT.

In conclusion, after reading this case I was rather convinced that it was a case of SLE complicated by cardiac tamponade, effectively responding to corticosteroid treatment.

Competing Interests: No competing interests were disclosed.

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