A Case of Recurring Kikuchi Disease and Autoimmune Hepatitis

Umaima Dhamrah[1], Branden Ireifej[1], Sibghatallah Ummar[2], Nuzhat Batool[1], David Song[3], Nirali Sheth[1], Abhigan Shrestha[4], and Vikash Jaiswal[5]

[1]Icahn School of Medicine at Mount Sinai
[2]Bahria University Medical & Dental College Karachi, Pakistan
[3]Icahn School of Medicine at Mount Sinai Elmhurst Hospital
[4]M Abdur Rahim Medical Medical College Hospital
[5]AMA School of Medicine

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Abstract

We present a case of a 47-year-old female with a history of diagnosed KD and autoimmune hepatitis 13 years ago who presented with recurrent fevers and a desquamative rash on the lower extremities. Patient has elevated ASMA titer, and a subsequent liver biopsy confirmed the diagnosis of autoimmune hepatitis

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Authors :

1. Umaima Dhamrah[1](dhamrahumaima@gmail.com),(Department of Internal Medicine, NYC Health & Hospitals/Elmhurst Hospital Center, Elmhurst NY, USA
2. Branden Ireifej[1](*ireifejb@nychhc.org*), (Department of Internal Medicine, NYC Health & Hospitals/Elmhurst Hospital Center, Elmhurst NY, USA)
3. Sibghatallah Ummar[2](*ummarsibghatallah@gmail.com*), Department of Medicine, Bahria University Medical & Dental College Karachi, Pakistan
4. Nuzhat Batool (*batooln@nychhc.org*)[1](Department of Internal Medicine, NYC Health & Hospitals/Elmhurst Hospital Center, Elmhurst NY, USA
5. David Song DO, (*xdavidsong@gmail.com*), (Department of Internal Medicine, NYC Health & Hospitals/Elmhurst Hospital Center, Elmhurst NY, USA)
6. Nirali Sheth (*niralisheth221@gmail.com*), (Department of Internal Medicine, NYC Health & Hospitals/Elmhurst Hospital Center, Elmhurst NY, USA)
7. Abhigan Babu Shrestha MBBS (*abigan17@gmail.com*), (Department of Medicine, M Abdur Rahim Medical College, Dinajpur, Bangladesh)
8. Vikash Jaiswal (Larkin Community Hospital, South Miami, FL, USA)

Corresponding Author:

Abhigan Babu Shrestha MBBS

Department of Medicine, M Abdur Rahim Medical College, Dinajpur, Bangladesh

Email Id: *abigan17@gmail.com*

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Abstract :

We present a case of a 47-year-old female with a history of diagnosed KD and autoimmune hepatitis 13 years ago who presented with recurrent fevers and a desquamative rash on the lower extremities. CT neck showed enlarged lymph nodes, and with her daily fevers and skin rashes the presentation was concerning for recurrence of her KD. Patient was also found to have an elevated ASMA titer, and subsequent liver biopsy confirmed the diagnosis of autoimmune hepatitis. She was started on methylprednisolone with improvement. Our case emphasizes the association of KD with autoimmune conditions other than SLE. Given the recurrence of the disease after a decade of quiescence, long-term follow-up of patients with KD should be implemented.

Key Clinical Message:

Kikuchi disease (KD) can relapse despite a remission period, with other recurrent autoimmune diseases closely linked. Our case emphasizes the association of KD with autoimmune conditions other than SLE. Given the recurrence of the disease after a decade of quiescence, long-term follow-up of patients with KD should be implemented.

Keywords: Kikuchi disease, Hepatitis, Pathology

Background:

Kikuchi's disease (KD) is a rare, benign disease that is characterized by fever and localized lymphadenopathy, and its association with autoimmune diseases has been scarcely reported. We describe a report of recurring KD, originally in remission for several years, along with a concomitant flare up of biopsy proven autoimmune hepatitis (AIH).

Case Presentation:

A 47-year-old Hispanic female with a history of KD diagnosed 13 years ago presented with recurrent fevers, desquamative rash on the legs, and diffuse arthralgias. She reported daily fevers at noon and in the evening starting one month ago. One week prior to her presentation, she developed abdominal pain associated with nausea, anorexia, and dark urine. Vital signs on arrival were unremarkable without evidence of hypotension, tachycardia, or hypoxia. Physical examination was significant for a desquamative rash on her thighs bilaterally with no clinical lymphadenopathy. Infectious work up including chest x-ray, urinalysis, complete blood count, and blood cultures were unremarkable. Further laboratory results revealed an elevated AST 241 units/L, ALT 277 units/L, and ALP 118 units/L (previously normal 3 months ago). Autoimmune work up including ANA, anti-dsDNA, anti-SSA/SSB were negative but did have a weakly positive ASMA with titers of 1:40. Ultrasound of the abdomen showed cholelithiasis without cholecystitis and hepatic steatosis. CT neck showed a 1.8 cm right submandibular, 1.6 cm left submandibular, subcentimeter submental and supraclavicular lymph nodes. Her lymphadenopathy with daily fevers, skin rash and joint pain was concerning for recurrence of her KD. Given her elevated liver function tests, there was also concern for concomitant acute AIH. Patient then underwent liver biopsy which confirmed AIH. Patient was subsequently started on methylprednisolone with improvement in her liver function tests along with her fevers and arthralgias. She was started on Azathioprine at discharge and continued to follow up in rheumatology and liver clinic as outpatient for management of her disease.

The patient had a known history of AIH associated with KD, which was diagnosed 13 years ago via liver biopsy and axillary lymph node biopsy, respectively **(Figure 1)** . Work up at the time of diagnosis was negative for ANA, anti-dsDNA, anti-Sm, anti-RNP and positive for elevated liver enzymes. She was started on corticosteroids and azathioprine, which was self-discontinued after 8 years. Her symptoms were quiescent for years until now.

Discussion:

The extranodal symptoms of KD are uncommon and diverse including skin rash, night sweats, weight loss, headache, cough, and abdominal pain [1]. The disease resolves spontaneously in weeks to months, but in some cases, it may have systemic involvement or evolve to SLE [2-4]. A definite diagnosis of KD is

based on characteristic pathologic findings on biopsy that differentiate this disease from others such as lymphoma, SLE, and infectious lymphadenopathy. Characteristic histopathologic findings include necrotic and thrombotic blood vessels. The karyorrhectic foci are formed by different cellular types, predominantly histiocytes and plasmacytoid monocytes, but also immunoblasts and small and large lymphocytes [5].

Recurrence of KD is rare. There have been more recent reports of recurrence rates as high as 13% in an Asian population in Korea. [6] In a study by Jung et al, 54 patients (11.3%) experienced 1–4 recurrent episodes of KD each. The initial recurrence occurred within a mean duration of 6 months (range: 1 month to 6 years). Patients with recurrent KD were more likely to have extranodal symptoms and lymphopenia. Although the etiology of KD recurrence is unknown, certain viral infections have been hypothesized to be among the triggers for KD relapse [8].

Patients with KD showing progression to autoimmune diseases were more likely to have fever, common extranodal symptoms, a higher recurrence rate, and a higher ANA positivity rate at KD diagnosis [1]. Hepatosplenomegaly was present in 18% of patients of KD [7] with few cases having clinically diagnosed AIH. Our case had biopsy proven AIH without clinical and laboratory findings of SLE. In addition, our case had recurrence of symptoms of KD after 9 years which improved after starting corticosteroids.

Conclusion:

KD is a rare condition, and it should be considered in differential diagnoses of tender lymphadenopathy, especially lymphadenopathy localized to the cervical region and recurrent fever with AIH. Although the disease takes a self-limiting clinical course in most cases, we reported a case of KD with a prolonged relapse of ten years. Our case emphasizes the association of KD with autoimmune conditions other than SLE. Full recovery with a good response to corticosteroid regimen was achieved after the recurrence; therefore, considering the recurrence of KD, long-term follow-up of patients with KD should be implemented.

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Ethical statement: Institutional Review Board (IRB) approval is not required for case reports at our institution.

Availability of data and materials

The data used in the case report are available on reasonable request.

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Not applicable.

Authors' contributions

UD, BI , NU, and DS were involved in patient care (diagnosis, treatment, and follow-up). UD, BI, NU, NS, VJ, AS and SU contributed to the collection of case information, writing of the manuscript, and manuscript revision. VJ and DS were involved in revising the manuscript critically for important intellectual content. All authors approved the final version.

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