Case Report

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Mortal Complication of Kikuchi–Fujimoto Disease; Lower Gastrointestinal Bleeding, Case Presentation and Literature Review

Sultan Tuna Akgöl Gür[1], Aslı Leyla Tahiroğlu[1], Abdullah Osman Koçak[1]

[1]Department of Emergency Medicine, Faculty of Medicine, Ataturk University, Erzurum, Turkey.

Abstract

In this report, we inform you that a patient with CFD, known as a benign disease, died due to massive lower GI bleeding. Although KFD is a benign disease, we have seen in this case that it can result in death. A 28-year-old Turkish male patient was brought to the hospital with complaints of high fever, fatigue, sweating and difficulty breathing. A cervical examination revealed that the patient had lymphadenopathy. The patient did not have any cutaneous lesions. Genital examination revealed melena. A normal cranial and abdomen, thorax CT was obtained. The patient was intubated because of respiratory failure and unstable hemodynamics. The patient developed pancytopenia as a result of blood tests. The patient died due to massive lower gastrointestinal bleeding.

Keywords: Kikuchi–fujimoto disease, lower gastrointestinal bleeding, histiocytic necrotizing lymphadenitis

Introduction

Kikuchi–Fujimoto Disease (KFD) or histiocytic necrotizing lymphadenitis is known to be a benign, generally self-limiting condition, which usually affects female patients under the age of 30 years. Most of the cases improve within six months. It is a benign disease mainly characterized by high fever, lymph node swelling, and leukopenia (1,2). It presents most often with cervical lymphadenopathy, which may be painful and can be accompanied by fever and upper respiratory tract symptoms. Unilateral involvement of the posterior cervical group is the most common manifestation (3). Less common manifestations include fever, axillary and mesenteric lymphadenopathy, splenomegaly, parotid gland enlargement, cutaneous rash, arthralgias, myalgias, aseptic meningitis, bone marrow haemophagocytosis, and interstitial lung disease. The cutaneous lesions include erythematous macules, papules, plaques, and nodules (4). Since the first description of the disease by the Japanese pathologists Kikuchi and Fujimoto (1,2) its etiology has remained unknown, although environmental factors, in particular viruses, have been suspected (5). Treatment is symptomatic with nonsteroidal anti-inflammatory drugs, and most symptoms improve within a few months (6). Laboratory findings are usually normal except for inflammatory syndrome or mild cytopenia, sometimes associated with hemophagocytosis (7). With this case, we present the coexistence of KFD and lower GIS bleeding for the first time in the literature. Our case aims to inform that there may be complications with a mortal course in KFD and to inform our physicians about these complications.

Case Report

A 28-year-old male patient was brought to the hospital with complaints of high fever, fatigue, sweating and difficulty breathing. In August 2022, it was found that the patient has been diagnosed with KFD as a result of a bone marrow biopsy. Biopsy is the gold standard diagnostic method for this disease. It was clarified that the patient had no additional disease. His vital signs during admission were as follows: blood pressure 121/71 mmHg, pulse 132/minute, SpO2 81%, and a fever of 38,3. His ECG showed a sinus tachycardia rhythm. His cervical examination revealed lymphadenopathy. The patient did not have cutaneous lesions. His breath sounds were normal bilaterally. Genital examination revealed melena. Laboratory results at our hospital were as follows: white blood cell count, 5,84/µL (33,1% neutrophils); absolute neutrophil count, 1,94/µL; hemoglobin level, 8.1 g/dL; platelet count, 32,000/µL; aspartate aminotransferase, 1210 IU/L; alanine aminotransferase, 1187 IU/L; and γ-glutamyl transferase, 197 IU/L. The patient's radiological imaging (brain, thorax and abdominal CT) was reported as normal. Epstein-Barr virus (EBV), Parvovirus B19, Anti-nuclear antibody (ANA), Brucella, Toxoplasma,

Corresponding Author: Sultan Tuna Akgöl Gür
e-mail: sultantuna@hotmail.com
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anticardiolipin IgM tests, as well as negative cytoplasmic and perinuclear anti-neutrophil cytoplasmic antibodies (C-ANCA and PANCA), anti-double-stranded DNA, and HLA-B57 values that have been tested at an external medical center were found to be negative, these tests are used to confirm the diagnosis. The patient was started on immediate replacement therapy (blood replacements, 2 Units of ES, 2 units of TDP). During the treatment, his hemodynamics deteriorated and intubation was performed in the emergency room for airway protection. After his hemodynamics improved, the patient was admitted to the internal medicine ICU from the emergency room. The blood panel that was taken in the ICU and showed hgb value of 5.4 mg/dl. He developed cardiac arrest 10 hours after hospitalization and the patient died despite all interventions.

Discussion

Although KFD usually presents with fever, cervical lymphadenopathy, or involvement of other lymph nodes, it may also present with symptoms related to other systems. These include maculopapular rash due to skin involvement, anemia due to bone marrow involvement, night sweats due to systemic exposure, weight loss, nausea, vomiting, sore throat due to respiratory system involvement and abdominal pain hepatomegaly and splenomegaly due to gastrointestinal system involvement (3).

Kucukardali et al. reported the characteristics of the patients in a study that analyzed 244 cases published by them. Accordingly, in these cases, the most common symptoms were weakness and joint pain, while the most common physical examination findings were lymphadenomegaly and erythematous rashes. Our case was also compatible with this information. In this case report, anemia and lymphopenia were frequently detected as laboratory tests, and this was consistent with our case. While additional diseases were detected in 51% of the patients in this article, there were none in our case. Küçükkardalı et al. reported that 64% of the patients recovered on their own without any treatment. In our case, he was not currently taking any medication (3).

Dumas et al. reported asthenia, weight loss and fever as the most common symptoms in their series of 91 cases. As for laboratory tests, high inflammatory parameters and neutropenia came to the fore. Gastrointestinal bleeding was not reported in either of the two multi-case analyses (6).

The association of KFD disease with bleeding has been reported only rarely in the literature. Bilateral preretinal hemorrhage has also been reported in a recent scientific article (8).

As the rate of diagnosis of KFD due to medical progress increases, the disease is better recognized and more complications are reported. Akhavanrezayat et al. have linked the mechanism of bilateral preretinal hemorrhage to autoimmunity. According to this, the cause of KFD disease is not clearly known and many causes are being investigated, but the most plausible is autoimmune causes. Increased vascular permeability due to direct vascular damage by autoimmune reactions or in response to the release of immune-mediated cytokines can lead to vascular injury and bleeding. Increased fragility of superficial capillaries due to vasculitis is also considered as one of the possible mechanisms (8). In our article, we think that gastrointestinal bleeding is due to increased vascular permeability and auto-immune system involvement. Since gastrointestinal system bleeding would not be expected in a normal young patient, we think that this patient had bleeding due to the effect of Kikuchi syndrome.

Our article reports the association between KFD and gastrointestinal bleeding, which was reported for the first time in the medical literature. Reported complications help us to manage diseases properly. It is much more important to report complications, especially in rare diseases.

Conclusion

In conclusion, the association between KFD and gastrointestinal bleeding, which was reported for the first time in the medical literature, is very important. Physicians need to know the complications that develop especially in rare diseases. It should be kept in mind that CFD should be considered especially in patients with fever, cervical lymphadenopathy and fatigue triad, and bleeding may occur as a complication of KFD.

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