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Kikuchi-Fujimoto disease: potential immune-mediated pathogenesis, a rare case and literature review

Payam S. Pahlavan

Terre Haute Regional Hospital, Terre Haute, Indiana, USA

Introduction

Kikuchi-Fujimoto is a rare, benign disease characterized by necrotizing lymphadenopathy. It is categorized as a reactive lymphadenopathy with paracortical hyperplasia. It usually presents with a painful enlarged cervical lymph node, but less frequently other superficial lymph nodes may also be involved. Early non-specific follicular hyperplasia with later prominent paracortical apoptosis surrounded by histiocytes and plasmacytoid monocytes with lack of neutrophils are the typical histopathological features. When the areas of necrosis and apoptosis are large, they may mimic other granulomatous lymphadenitis. The etiology of Kikuchi-Fujimoto disease is unclear. We here review a case of Kikuchi-Fujimoto disease and propose the immune-mediated pathogenesis for its etiology.

Case presentation

We present the case of a 25-year-old female of Caucasian descent who presented with acute tender unilateral (right) cervical lymphadenopathy that had started two weeks earlier without constitutional symptoms. The mass was first noticed with sore throat and cough and redness of the skin over it. The patient's past medical history was significant with having had asthma, recurrent bronchitis and multiple food allergies. A family history of diabetes was noted in her maternal grandfather. The patient described that the mass had gradually enlarged. The current unilateral neck pain was not continuous since the start, but was worsened by palpation. She denied any recent travel. On physical exam, her neck had a normal range of motion. There was a palpable, rubbery, well-circumscribed mass 2 × 1.7 × 1.5 cm close to the angle of the mandible on the right side of the neck. The mass was movable, with no adhesion to the skin or underlying soft tissue. On lab exam, white blood cell level and acute phase reactant were normal. Antinuclear antibodies (ANA) was negative. An excisional biopsy was performed. The mass appeared as a large lymph node. A portion of it was submitted for flow cytometry and the rest was submitted for histopathological review. Flow cytometry did not identify any clonal B- or T-cell populations. Histology showed sections of a lymph node with architectural distortion with areas of necrosis in both the paracortical and inter-follicular areas (Figures 1 and 2). Karyorrhexis, fibrin deposits and scattered histiocytic infiltration were noted. No neutrophils or eosinophils in the necrotic area were seen. Immunohistochemistry did not highlight any lymphoproliferative lesion. Mycobacterial and fungal stains were also negative. The diagnosis of Kikuchi-Fujimoto disease was confirmed after ruling out other possibilities.

Discussion

Kikuchi-Fujimoto disease is a rare, self-limited disease that is most frequently seen in young Asian women [1]. A literature review showed that concomitant asthma and other allergic related diseases (e.g. allergic rhinitis) with Kikuchi-Fujimoto disease have previously been seen [1–3]. Also, associations with other autoimmune diseases such as systemic lupus erythematosus (SLE) and Sjogren's syndrome have been noted [4, 5]. In up to 30% of cases, the onset of Kikuchi-Fujimoto disease starts before the onset of SLE [6].

Our current case also had a history of asthma. Even though the pathogenesis of Kikuchi-Fujimoto disease is unclear, an association between the host immune response and allergic reaction should be considered as one possible factor.

*Address for correspondence: Payam S. Pahlavan, Terre Haute Regional Hospital, 534 Antioch Cir W, Terre Haute, Indiana 47803, USA, e-mail: payamsp@yahoo.com

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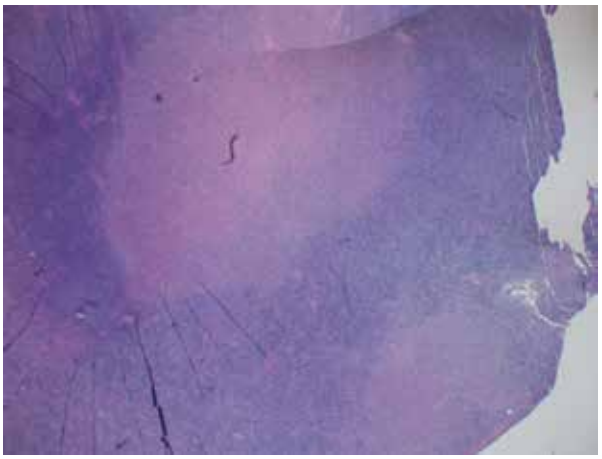

Figure 1. Lymph node with architectural distortion with pale areas (4× magnification)

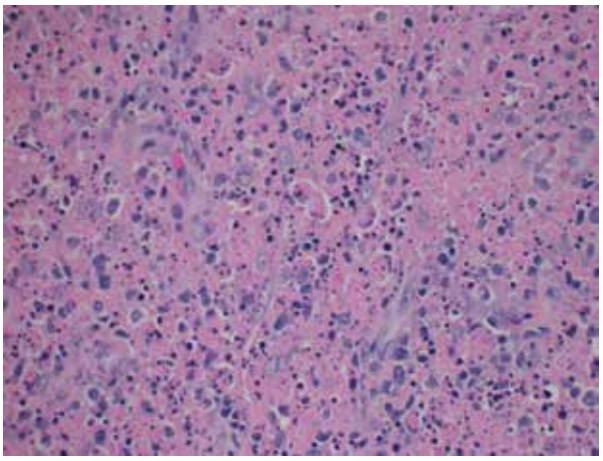

Figure 2. The pale areas composed of histocytes, eosinophilic granular material and abundant karyorrhectic debris with few plasmacytoid dendritic cells. No neutrophils present (40× magnification)

Authors' contributions

PSP — sole author.

Conflict of interest

There is no conflict of interest.

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Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; uniform requirements for manuscripts submitted to biomedical journals.

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