İstanbul Med J 2013; 14: 221-3
DOI: 10.5152/imj.2013.60

Kikuchi-Fujimoto Disease: A Rare Case Report Questioning the Diagnostic Utility of Fine Needle Aspiration Cytology

Kikuchi-Fujimoto Hastalığı: Nadir Bir Olgu Eşliğinde İnce İğne Aspirasyon Sitolojisinin Tanısal Değeri

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Abstract / Özet

Kikuchi-Fujimoto disease (histiocytic necrotizing lymphadenitis) is a rare cause of cervical lymphadenopathy which was first described in 1972. It usually affects young Asian women. The patient usually presents with systemic signs such as fever. Although the exact cause of the disease is unknown, infectious diseases and autoimmunity are suspected in many cases. We here present a biopsy-proven case of Kikuchi-Fujimoto disease in a 26 year-old Turkish woman with its FNA findings and discuss the role of USG-guided FNA in diagnostic workup.

Key Words: Kikuchi-Fujimoto disease, histiocytic necrotizing lymphadenitis, cervical lymphadenopathy

Kikuchi-Fujimoto hastalığı (histiyositik nekrotizan lenfadenit), 1972'de tanımlanmış, genelde Asyalı genç bayanlarda görülen, ateş gibi sistemik bulgularla seyreden nadir bir servikal lenfadenopati nedenidir. Etiyopatogenezi bilinmemekle birlikte enfeksiyonlar ve otoimmunite suçlanmaktadır. Biz, 1,5 aydır süren ve antibiyotik ile düzelmeyen, sağ boyunda ağrılı şişlik nedeniyle başvuran, Kikuchi-Fujimoto hastalığı tanısı koyduğumuz 26 yaşında bayan hastayı radyoloji, İİA sitolojisi ve histopatolojik bulguları ile birlikte sunmayı amaçladık. İİA sitolojisi yararlı bir tanısal yöntem olmakla birlikte bu hastalıkta tanıyı kesinleştirebilmek için çoğu kez histopatolojik inceleme de gerekmektedir.

Anahtar Kelimeler: Kikuchi-Fujimoto hastalığı, histiyositik nekrotizan lenfadenit, servikal lenfadenopati

Introduction

Kikuchi-Fujimoto disease (KFD), or histiocytic necrotizing lymphadenitis, is a rare cause of cervical lymphadenopathy affecting mainly young Asian women (1, 2). Although many conditions or etiological agents such as autoimmunity or viral infections such as Epstein-Barr virus have been proposed, the etiology and pathogenesis remains unknown. No specific laboratory tests contribute to diagnosis. Excisional biopsy of the involved lymph node is mandatory to establish a diagnosis of KFD (1). The role of fine needle aspiration (FNA) cytology in diagnosing KFD is still controversial (1, 3). In this study, we aimed to report a biopsy-proven case of KFD with its FNA findings and discuss the role of USG-guided FNA in the diagnostic workup.

Case Report

A 26-year-old young Turkish woman presented to our Ear Nose and Throat Clinic with a one-month-history of painful swelling in the right neck region which was unresponsive to antibiotics. She denied any systemic complaints except for mild fever and mild flu-like symptoms. In the physical examination, a right upper cervical mass (level 2) with moderate pain was observed. Laboratory findings including the complete blood count, liver function tests and thyroid function tests were within normal limits. Viral serology for EBV and hepatitis viruses were negative. The erythrocyte sedimentation rate was low. On MRI, a right cervical lymphadenopathy with signal increase in the central area suggestive of necrosis was observed (Figure 1). USG-guided FNA of the mass was planned. Rapid on-site evaluation was done at the Radiology Department. In cytological smears and cell block, an inflammatory infiltrate composed mainly of lymphoid cells with scattered tingible body macrophages in the background of karyorrhectic debris were seen (Figure 2A, B). The diagnosis of "highly suggestive for necrotizing lymphadenitis but lymphoid malignancy with extensive necrosis cannot be ruled out" was given. In order to obtain a final diagnosis, excisional biopsy wasperformed. Grossly, the mass was 3x3x2.5 cm and insignificant in consistency. Histopathologically, extensive areas of necrosis in a background of karyorrhectic debris was observed (Figure 3). Although we did not observe any granulomas, we performed Ziehl-Nielsen stain in these necrotic areas. It was negative for any acid fast bacilli. Immunohistochemically; myeloperoxidase was negative in necrotic areas showing that the neutrophils were absent (Figure 4A) and CD138, a plasma cell marker, was also negative. On the other hand, there were many tingible body macrophages within and around the necrosis showing immunoreactivity for CD68 (Figure 4B). Large transformed lymphocytes with immunoblast morphology were also seen around the vessels (Figure 3). The final diagnosis of KFD was made. The patient was given antiinflammatory drugs

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Received Date/Geliş Tarihi:
16.12.2012

Accepted Date/Kabul Tarihi:
14.04.2013

and is well with significant regression of her neck masses at her last follow-up two months after her operation.

Discussion

Kikuchi-Fujimoto Disease is a self-limiting condition, usually resolving within 4 months, but low recurrence rates of 3% to 4% have been reported (4). There is no specific treatment since the etiology is unknown. It should be distinguished from other diseases such as lymphoma or tuberculosis which would require specific treatment modalities. It frequently presents as a challenge even for hematopathologists. There are numerous KFD cases reported in the literature as being misdiagnosed as malignant lymphoma. The mainstay of the correct diagnosis of KFD is the morphologic examination of the lymph nodes involved (1). Typical histopathological changes are the lymphohistiocytic infiltrate with abundant karyorrhectic debris without any or few neutrophil infiltration. Other non-malignant reactive conditions such as systemic lupus erythematosus (SLE), sarcoidosis or infectious lymphadenitis such as toxoplasmosis, tuberculosis, cat scratch disease, leprosy and yersiniosis should also be considered in the differential diagnosis (5). She had no signs of SLE such as butterfly rash, joint pathology. Besides, we observed no characteristic histological features of lupus lymphadenopathy such as prominent follicular hyperplasia and interfollicular plasma cells (6). The paucity of neutrophils helped us to exclude the above-mentioned infectious lymphadenitis conditions. On the other hand, fine needle aspiration cytology is also helpful in the diagnostic workup. Typical cytological findings are polymorphous lymphoid population with abundant karyorrhectic debris

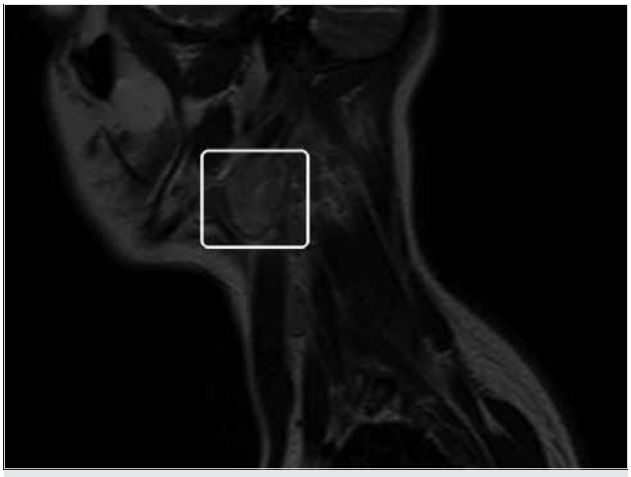

Figure 1. Right cervical lymphadenopathy with high signal intensity in central areas suggestive of necrosis

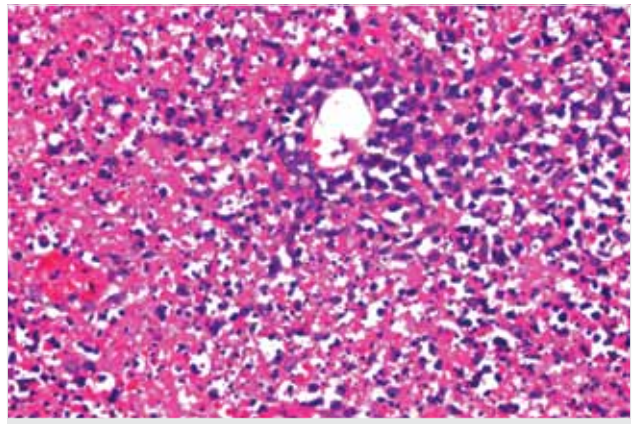

Figure 3. Histopathologically, histiocytic necrotizing lymphadenitis with perivascular immunoblasts (H&E, X400)

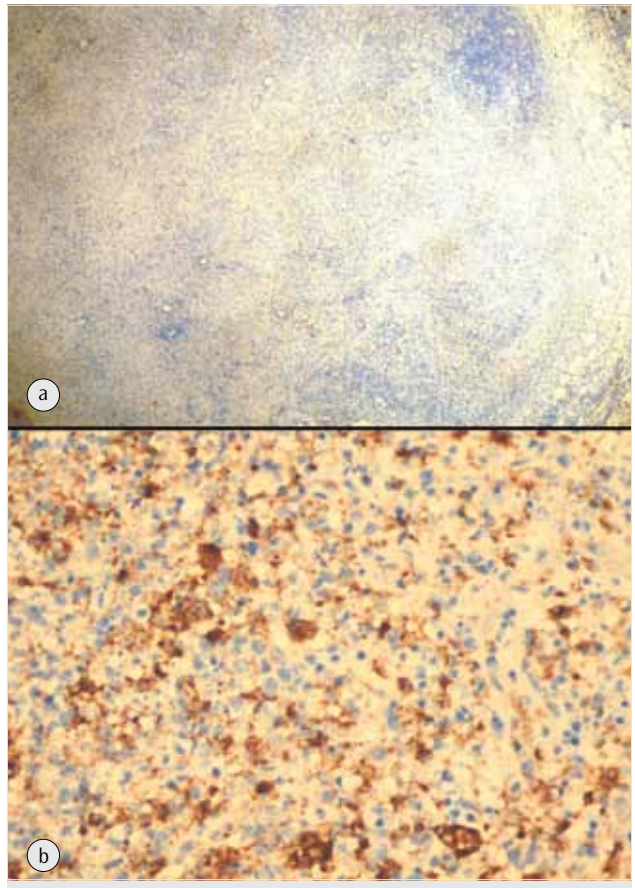

Figure 4. A) Necrosis without any or few neutrophils (MPO, X40).
B) Many histiocytes within and around the necrotic areas (CD68, X400)

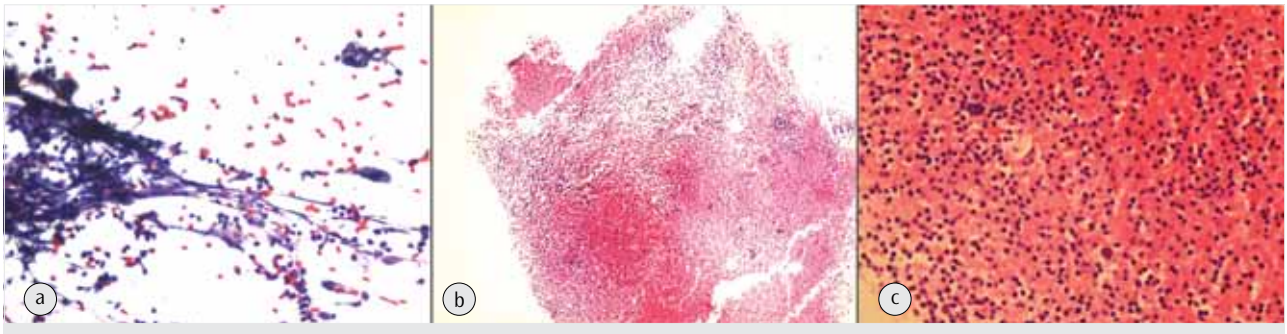

Figure 2. Lymphohistiocytic infiltrate in fine needle aspiration smears and cell block (Papanicolaou, X400)

and histiocytes, many of which showed a small size and eccentrically placed, crescent nuclei (3, 7, 8). However, the rate of making a correct diagnosis without necessitating an excision is 50% (3). Although these studies highlight the role of FNA cytology, it seems that it is still insufficient to establish a final diagnosis since there is no single cytomorphological feature pathognomonic for KFD in FNA smears (3, 7, 8).

Conclusion

Fine needle aspiration cytology could only be suggestive but not diagnostic for KFD even in a typical clinical setting (young woman with cervical lymphadenopathy) with characteristic cytological findings. Hence, the excision and histopathological examination of the lesion is mandatory.

Conflict of Interest
No conflict of interest was declared by the authors.

Peer-review: Externally peer-reviewed.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Author Contributions
Concept - N.A.Y.; Design - N.A.Y., A.M.; Supervision - A.M., E.Ü.M.; Funding - N.A.Y., A.M.; Materials - E.Ü.M., Ç.V.; Data Collection and/or Processing - N.A.Y., A.M., E.Ü.M.; Analysis and/or Interpretation - N.A.Y.; Literature Review - N.A.Y.; Writing - N.A.Y., E.Ü.M.; Critical Review - A.M., Ç.V.

Çıkar Çatışması
Yazarlar herhangi bir çıkar çatışması bildirmemişlerdir.

Hakem değerlendirmesi: Dış bağımsız.

Hasta Onamı: Yazılı hasta onamı bu çalışmaya katılan hastadan alınmıştır.

Yazar Katkıları
Fikir - N.A.Y.; Tasarım - N.A.Y., A.M.; Denetleme - A.M., E.Ü.M.; Kaynaklar - N.A.Y., A.M.; Malzemeler - E.Ü.M., Ç.V.; Veri toplanması ve/veya işlemesi - N.A.Y., A.M., E.Ü.M.; Analiz ve/veya yorum - N.A.Y.; Literatür taraması - N.A.Y.; Yazıyı yazan - N.A.Y., E.Ü.M.; Eleştirel İnceleme - A.M., Ç.V.

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