Kikuchi-Fujimoto Disease in an Atypical Site

Enfermedad de Kikuchi-Fujimoto: una localización atípica

To the Editor:

Kikuchi-Fujimoto disease or histiocytic necrotizing lymphadenitis is a rare entity that is generally associated with cervical lymphadenopathy and fever. We report a case of this disease in an atypical location, the anterior mediastinum, associated with multiple pulmonary nodules.

A 27-year-old woman was evaluated in the internal medicine department for a 3-month history of fever, mainly in the evening, night sweats, and nonpruriginous maculopapular lesions. Physical examination was normal, with no evidence of cervical or supraclavicular adenopathy. Laboratory tests showed no abnormalities. Agglutination tests for Typhi and Brucella were negative, as were antibody titers against Epstein-Barr virus, rheumatic and immunological tests, and the Mantoux test. A computed tomography scan of the chest and abdomen revealed small peripherally distributed subpleural nodules (the largest measuring 9 mm in diameter) in the right lung, discrete paratracheal and bilateral hilar lymph nodes, and an enlarged subcarinal lymph node (Figure). Fiberoptic bronchoscopy was normal. Microbiology and cytology of transbronchial and bronchial biopsy samples and bronchoalveolar lavage fluid did not provide conclusive findings.

Mediastinoscopy revealed a level 4R lymph node conglomerate, which was biopsied for microbiological and pathological examination. The microbiological analysis was negative, and the pathology report showed reactive lymphadenitis with nonneutrophilic necrosis suggestive of Kikuchi’s disease. One month after surgery, the patient had no fever and was asymptomatic.

Kikuchi-Fujimoto disease, nonneutrophilic necrotizing lymphadenitis or histiocytic necrotizing lymphadenitis is a self-limiting disorder that runs a benign course. It mainly affects young Asian women generally less than 30 years of age. Of unknown origin, the disease has been associated with viruses such as parvovirus B19, human immunodeficiency virus, Epstein-Barr virus, cytomegalovirus, and herpes simplex virus type 6, and with microorganisms such as Toxoplasma, Yersinia, and Brucella species. Some authors have described an exaggerated immune response to different agents as the cause of disease.

In 70% to 90% of cases, Kikuchi-Fujimoto disease manifests as cervical lymphadenopathy, which is generally unilateral. The parotid gland is a less common location. Fever is another frequently observed symptom, occurring in 30% to 50% of cases. Other symptoms that may accompany this disease include fatigue, night sweats (as in our patient), weight loss, gastrointestinal disorders, and various skin alterations such as rash.

The diagnosis of this disease is complex as there are no conclusive laboratory or imaging test findings, though they may serve to exclude other disorders. Diagnosis can only be made by lymph node biopsy, since fine needle aspiration does not provide sufficient information.

From a pathologic point of view, the disease has 3 phases: proliferation (with an increase in mononuclear cells), necrosis (with necrosis and a predominance of histiocytes), and xanthoma. This means that diseases such as systemic lupus erythematosus, lymphomas, Hodgkin’s disease, and tuberculosis should be considered in the differential diagnosis of Kikuchi-Fujimoto disease.

Kikuchi-Fujimoto disease usually runs a benign, self-limiting course, though recurrence has been reported in some cases. Patients should be closely monitored, however, as there appears to be a clear association between lupus and Kikuchi-Fujimoto disease.

References


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