Visible Lymphadenopathy in Kikuchi-Fujimoto Disease



n otherwise healthy 11-year-old Japanese boy presented with a 3-week history of a right neck mass. Except for mild fatigue, he did not display any associated general symptoms, including fever, night sweats, or weight loss. Physical examination demonstrated a prominent tender cervical lymphadenopathy in the posterior cervical triangle (Figure 1). Blood tests reported leukopenia and mildly elevated lactate dehydrogenase and aminotransferase levels. Over the following 2 weeks, his lymph nodes enlarged and his fatigue worsened; thus, we performed a cervical lymph-node excisional biopsy that revealed increased CD 68+ histiocytes, lymphocytes, and immunoblasts in the cortices with abundant karyorrhectic debris (Figure 2); these findings were compatible with Kikuchi-Fujimoto disease (KFD). At the 1-month follow-up, the patient's symptoms had improved without any intervention.

KFD is a rare, self-limited disease of unknown etiology that was first described and commonly observed in Asian individuals. Given its clinical and pathologic similarities with lymphoma, excluding this condition is important in the evaluation of KFD. The hallmark of KFD is lymphadenopathy on the posterior cervical triangle, which is observed in 90% of cases. In our patient, enlarged posterior cervical lymph nodes were not only palpable, but were clearly visible, which resembled the classic Winterbottom sign observed in African trypanosomiasis. Because large lymphadenopathy can be an ominous sign of lymphoma, biopsy should be considered in patients with clearly visible posterior cervical lymphadenopathy and thought given to other rare entities, such as KFD or trypanosomiasis.

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Figure 1. Visible right cervical lymphadenopathies in the posterior cervical triangle.

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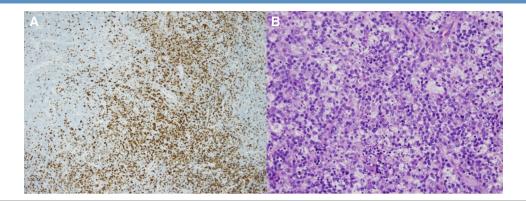


Figure 2. Cervical lymph-node excisional biopsies revealed increased CD 68+ histiocytes, **A,** lymphocytes, **B,** and immuno-blasts in the cortices with abundant karyorrhectic debris.