Intraparotid Kikuchi-Fujimoto Disease with Persistence of Disease: A Case Report

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Abstract

Educational Objective: At the conclusion of this presentation, the participants should be able to understand an uncommon presentation of Kikuchi-Fujimoto disease as an intraparotid gland tumor with persistence of disease.

Objectives: 1) Describe a rare presentation of Kikuchi-Fujimoto disease (KFD) as a parotid gland tumor. 2) Describe an atypical disease course for KFD.

Study Design: Case report with histopathologic and radiological analysis along with a literature review.

Methods: A case report with histopathologic and radiological details is described from a university hospital. Background, incidence, disease course, and treatment options are presented through a literature review.

Results: We present the case of a 25 year old Filipino female, who at the age of 9 was diagnosed with a left parotid mass. She underwent a parotidectomy four years later. She had two re-excisions for recurrences at the ages of 15 and 17. The last excision was followed by ipsilateral facial paresis which slowly improved. She moved to the United States at the age of 19. She had another recurrence and the tumor was evaluated in Arkansas with a neck biopsy. The final diagnosis was Kikuchi-Fujimoto disease. The patient presented to us for evaluation of her ipsilateral growing left parotid tumor with facial deformity and some oropharyngeal discomfort.

On physical examination, her cranial nerve VII was 2/6 on the House-Brackmann scale with asymmetric movement of her lower branches. Flexible laryngoscopy revealed bulging of the oropharynx down to the level of the vallecula on the left. Recent MRI demonstrates a large left parotid mass with extension into the mandibular space, pterygopalatine fossa, and carotid space. She continues to complain of an ipsilateral growing parotid tumor with facial deformity and oropharyngeal discomfort, but without constitutional symptoms. She will undergo rheumatologic evaluation prior to possible future surgical intervention.

Conclusions: KFD is an uncommon, idiopathic, and generally self-limiting cause of lymphadenopathy. It commonly presents with cervical lymphadenopathy in Asian females, and typically runs a benign course that resolves spontaneously in weeks to months. The disease is often misdiagnosed at first and mistaken for lymphoma or systemic lupus erythematosus. We present an atypical case with persistence of disease 16 years after initial presentation, and involvement of an uncommon site, parotid gland, with only 6 reported cases in the literature. Although treatment is surveillance, in the setting of symptoms and cosmetic deformity as in this case, surgical options may be entertained.

Background

Kikuchi-Fujimoto disease, also known as histiocytic necrotizing lymphadenitis, is an uncommon, idiopathic, generally self-limiting cause of lymphadenopathy. It commonly presents with cervical lymphadenopathy in Asian females, and typically runs a benign course that resolves spontaneously in weeks to months. The disease can be mistaken for lymphoma or lupus, and disease recurrence is unusual.

Most common clinical manifestations include:
- Cervical lymphadenopathy
- Fever
- Pharyngitis

The cause of the disease is unknown, however, infectious and autoimmune etiologies have been postulated. Kikuchi disease has been associated in certain cases with systemic lupus erythematosus. Lymphadenopathy most often resolves over several weeks to 6 months, although occasionally persisting longer. The disease recurs in about 3% of cases. Fatality is very rare.

The disease was first described in Japan and most cases have been reported from East Asia. Women are more often affected than men, and it typically occurs in young adults.

Kikuchi disease most frequently manifests as a relatively acute onset of cervical adenopathy associated with fever and a flu-like syndrome. The disease typically presents with lymphadenopathy with cervical nodes being affected in about 80% of cases. Posterior cervical nodal involvement occurs in 65-75% of cases. General lymphadenopathy involving axillary, inguinal, and mesenteric nodes are unusual. The disease rarely involves extranodal sites such as the parotid glands.

Figure 1. Frontal and lateral views of the patient demonstrating left parotid mass and facial deformity. The well healed neck scar is visible on the lateral view.

Conclusions

Kikuchi-Fujimoto Disease is an uncommon, idiopathic, and generally self-limiting cause of lymphadenopathy. It commonly presents with cervical lymphadenopathy in Asian females, and typically runs a benign course that resolves spontaneously in weeks to months. The disease is often misdiagnosed at first and mistaken for lymphoma or systemic lupus erythematosus. We present an atypical case with persistence of disease 16 years after initial presentation, and involvement of an uncommon site, parotid gland, with only 6 reported cases in the literature. The disease was self limited in the prior cases with parotid involvement. Although treatment is normally surveillance with supportive care, in the setting of symptoms and cosmetic deformity as in this case, other therapeutic options may be entertained. In our case, the patient was not deemed to be a surgical candidate due to the extent of her disease. She was however treated with low dose radiation with promising early response. She will need to further follow up to determine role of radiation therapy in this rare disorder.