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Intraparotid Kikuchi-Fujimoto disease masquerading as a parotid gland tumor

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Abstract The Kikuchi-Fujimoto disease, also known as histiocytic necrotizing lymphadenitis, is a self-limiting lesion of unknown cause first described in 1972 independently by Kikuchi and Fujimoto et al. This self-limiting disorder usually occurs in Asian women in their late 20s or early 30s. Typically, it runs a benign course and appears to resolve spontaneously 1 to 6 months after definite diagnosis. The Kikuchi-Fujimoto disease usually manifests as a localized cervical lymphadenopathy; therefore, most patients with this disease are seen in ears, nose, and throat practice. Nodal involvement other than in the neck area or extranodal involvement is rare. In this situation, however, the Kikuchi-Fujimoto disease is easily confused with other less-benign conditions. We describe a case of Kikuchi-Fujimoto disease in a 30-year-old man that presented as a parotid gland tumor. This is the third study to document intraparotid Kikuchi-Fujimoto disease in the English literature. Our report illustrates the clinical features of this unusual condition and emphasizes potential confusion with other diagnoses.

1. Case report

A 30-year-old man presented to our otolaryngology department with a 4-week history of a persistent swelling on his left preauricular area. There was no improvement after several cycles of antibiotics in other local medical departments. On physical examination, a firm, nontender mass was palpated on his left preauricular area. The mass was moderately mobile and measured 2 × 2 cm. The rest of the head and neck and systemic examination was unremarkable. The complete blood count, erythrocyte sedimentation rate, and blood chemistry values were within normal limits.

A preoperative computed tomographic scan revealed a well-defined solitary tumor within the patient's left parotid gland (Fig. 1). No evidence of abscess formation was noted. Under the clinical impression of a benign, localized parotid gland neoplasm, a limited superficial parotidectomy was performed. After exposure of the main trunk of the facial nerve, only the temporal and zygomatic branches that were immediately adjacent to the tumor were exposed. The tumor was excised with adequate margin of normal gland tissue (Fig. 2).

Histopathological analysis of the tumor revealed stellate necrosis with surrounding histiocytic inflammation (Fig. 3). Neutrophilic inflammation was absent. Stains for acid-fast bacilli, fungi, and cat scratch bacilli were negative. These features were consistent with the Kikuchi-Fujimoto disease. The postoperative period was uneventful. The patient did not experience recurrence during 14 months of follow-up.

2. Discussion

The pathogenesis of Kikuchi-Fujimoto disease remains unclear. The Kikuchi-Fujimoto disease is generally considered as an idiopathic pattern of nodal reaction to injurious

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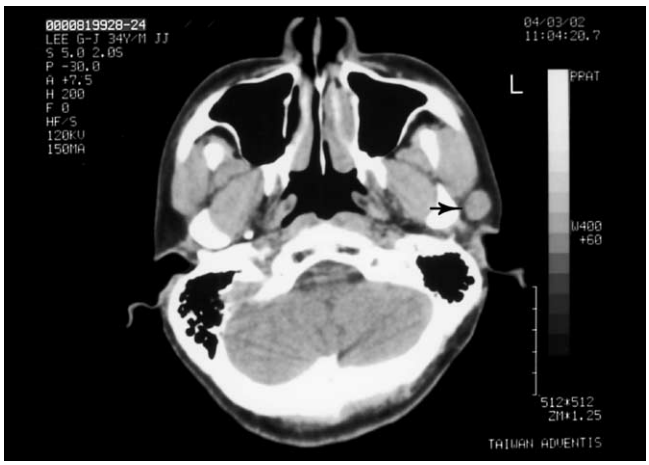

Fig. 1. Preoperative computed tomographic scan revealing a well-defined, solitary parotid gland tumor (arrow).

agents. Various microbiologic agents including adenovirus, parvovirus, and members of human Herpesviridae such as cytomegalovirus, Epstein-Barr virus, and human herpesviruses 6 and 8 have been implicated [1]. On the other hand, an autoimmune contribution to the etiology is postulated by observations that the Kikuchi-Fujimoto disease can precede or occur in association with a connective tissue disorder, especially systematic lupus erythematosus (SLE). Dorfman and Berry [2] suggested that the Kikuchi-Fujimoto disease is an attenuated form of SLE based on the histological similarities and female preponderance of both diseases.

The characteristic clinical presentation is with cervical lymphadenopathy, which is often painful or tender on palpation. The sites of cervical lymphadenopathy are common in the posterior cervical triangle and jugular carotid chain. Fever may be predominant. Dermatological findings, presented in 16% to 30% of cases, are nonspecific [3,4]. Additional complaints include nausea, diarrhea, headache, and constitutional disturbances (eg, malaise, night sweats, and weight loss). According to most English

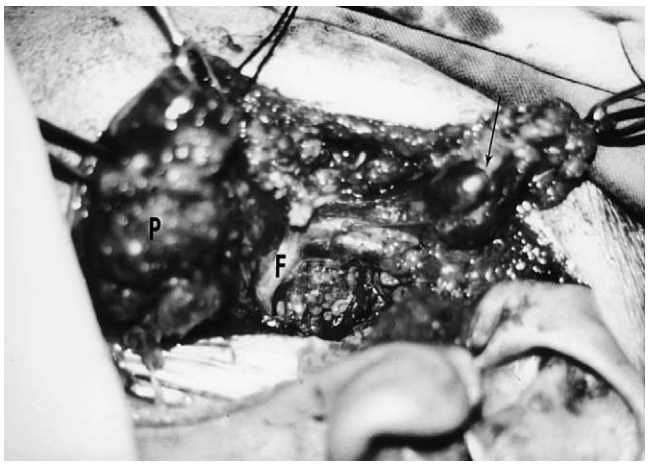

Fig. 2. Operative view during the tumor excision (F indicates the facial trunk; P, the residual parotid gland; arrow, the tumor).

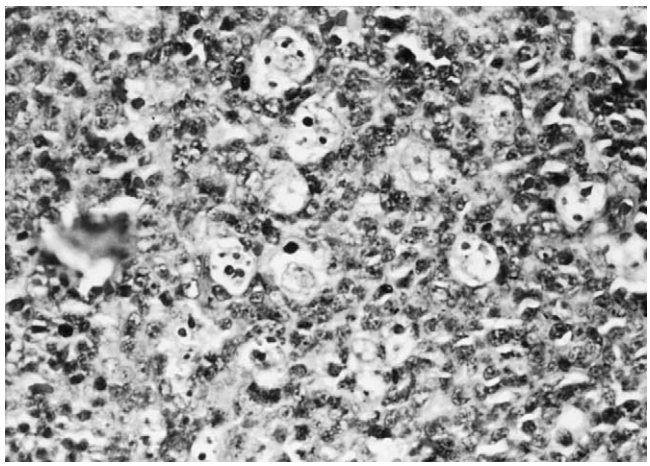

Fig. 3. Photomicrograph of surgical specimen shows the necrotizing process, with karyorrhectic nuclear debris and polymorphous aggregates of mononuclear cells. The mononuclear cell infiltrate is made of histiocytes and lymphocytes.

publications, this disease preferentially affects women in a 3 to 4:1 ratio [5,6]. However, less female predominance was reported in some Asian series [7,8]. Most patients are in their late 20s or early 30s.

Definitive diagnosis of Kikuchi-Fujimoto disease can only be made on direct histopathological examination of a lymph node biopsy. Fine needle aspiration cannot provide conclusive diagnosis of a Kikuchi-Fujimoto disease [8,9]. There are no definite laboratory tests available for the diagnosis of Kikuchi-Fujimoto disease, but negative tests are important to exclude other disorders. Also, there is no radiographic finding particular to the Kikuchi-Fujimoto disease.

The typical histopathological features of the Kikuchi-Fujimoto disease usually show a necrotizing process, with patchy or confluent areas of necrosis associated with karyorrhexis, and absence or paucity of neutrophils. In 1995, Kuo proposed 3 histopathological types of the Kikuchi-Fujimoto disease: proliferative, necrotizing, and xanthomatous [7]. These morphological variations arouse various differential diagnoses including SLE, malignant lymphoma, Hodgkin's disease, tuberculous lymphadenitis, toxoplasmosis, infectious mononucleosis, cat scratch disease, angioimmunoblastic lymphadenopathy, metastatic carcinoma, and in particular, Still's disease in children [1,2].

The Kikuchi-Fujimoto disease is generally considered a self-limiting disorder. The cervical lymphadenopathy resolves spontaneously over a period of several weeks to 6 months after definite diagnosis. Only a few fatal cases of the Kikuchi-Fujimoto disease had been reported [2,10-12]. Most authors do not recommend special treatment for the Kikuchi-Fujimoto disease. However, patients with distressing and severe symptoms could benefit from immunosuppressive treatment such as that with glucocorticoids [13]. The possible association between the Kikuchi-Fujimoto disease and SLE is well known; therefore, it is recommended that patients with Kikuchi-Fujimoto disease be

monitored on a long-term basis to check for the development of SLE [2]. Recurrence of the Kikuchi-Fujimoto disease is rarely observed [5,7,14].

3. Conclusion

This case raises interest for particular reasons. The clinical presentation of the 30-year-old man was not typical for a Kikuchi-Fujimoto disease. Neither pain nor fever was prominent at the initial stage. The intraparotid nodal involvement was exceptional. The computed tomographic scan finding favored the preoperative diagnosis to be a localized parotid gland tumor. All clinical information led to a limited superficial parotidectomy. Our report is the third to document intraparotid Kikuchi-Fujimoto disease [15,16]. The Kikuchi-Fujimoto disease is not a rare lymphadenitis among Asians. In fact, we believe that the disease is actually underrecognized and underreported outside Asia. It is those unusual features of the disease that confuse clinicians. Nodal involvement other than in the neck area or extranodal involvement could be mistaken for neoplasm. Soon before, we had encountered another case of Kikuchi-Fujimoto disease in an 8-year-old boy that presented as a submandibular gland tumor [17]. In regions where the disease is relatively prevalent, the Kikuchi-Fujimoto disease should be considered a possible cause of a tumor mass of major salivary glands. Although an early biopsy is instrumental in making a correct diagnosis of Kikuchi-Fujimoto disease, a tailored procedure is important in preventing potential complications during investigation. This self-limiting disorder requires no specific management but does long-term follow-up.

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