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CT of Kimura Disease

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Kimura is credited with first describing a condition—eosinophilic granuloma of the soft tissue—that subsequently has been given his name. Since then, more than 500 cases have been reported in review series, mainly in the Chinese and Japanese literature [1]. The disease occurs only in Orientals, and more than 80% of these patients are male. Presentation may occur over a wide age range, but it is most common in the second and third decades. Symptoms may fluctuate during periods of many years. The presenting feature is usually swelling in the region of the salivary glands and local lymphadenopathy. Locally, erythema and induration of the overlying skin may be present, along with a papular or follicular rash. Less common sites include all extracranial soft-tissue areas of the head and neck, with rare involvement of the axilla, groin, popliteal region, forearm, hand, and spermatic cord [2, 3]. The lesion is usually solitary, but multiple and disseminated lesions have been reported.

Characteristically the peripheral blood eosinophilia reaches 30%–40%. Increased serum IgE has also been observed. These findings suggest an allergic or parasitic cause, but searches for *Ankylostoma*, *Ascaris*, and many other organisms have been unsuccessful [1]; so, the etiology remains unknown. Histologic examination shows heavy lymphocytic and eosinophilic infiltration with vascular proliferation and fibrosis. Lymphoid tissue is hyperplastic.

Other lesions similar to Kimura disease have been described. Of these, angiolymphoid hyperplasia with eosinophilia (ALHE) is the most common. This condition differs in that (1) it affects non-Orientals, frequently women in an older age group; (2) there is involvement of more superficial subcutaneous tissues; (3) generally no lymphadenopathy is present; (4) peripheral eosinophilia is less marked; (5) vascular proliferation is more prominent; and (6) fibrosis is not a feature. Less common lesions with similar histologic features form a spectrum of disorders, and there is continuing debate in the literature as to whether these should all be classified as ALHE or whether they are distinct entities. For the present, Kimura disease has been placed under this general heading [4–6].

Kimura disease must not be confused with eosinophilic

granuloma, which is never associated with peripheral eosinophilia.

Previous radiologic investigations of Kimura disease were confined to radiographs and were essentially unhelpful.

Case Report

A 24-year-old man presented with a 19-year history of a right-sided mass at the angle of the mandible without facial-nerve dysfunction. Biopsy on two previous occasions (several years before) revealed "lymph node tumor" on the first occasion and "follicular hyperplasia" on the second, at which time a peripheral blood eosinophilia was noted. The size of the mass had fluctuated intermittently over the years. The peripheral blood count showed an eosinophilia of 64% with an absolute count of $12,000 \times 10^9/l$.

CT scans (Fig. 1) showed a swollen right parotid gland with an irregular mass at the superficial and inferior aspect of the gland and bilateral cervical lymphadenopathy. Surgery revealed a diffuse fibrotic, vascular, infiltrating process involving the right superficial lobe of parotid, surrounding lymph nodes, submandibular gland, and deep cervical lymph nodes.

A right superficial parotidectomy was performed. The right submandibular gland was resected along with the affected superficial lymph nodes. Further nodes in the inferior deep cervical chain and accessory lymph nodes were removed, necessitating partial resection of the sternomastoid muscle and interruption of the cervical branch of the facial nerve.

Histology confirmed involvement by the disease process of all resected tissue. Sections of the parotid showed fibrofatty tissue infiltrated by aggregates of lymphocytes that form follicles, sheets of eosinophils, and proliferation of small blood channels surrounded by fibrosis. Sections of the submandibular gland showed mixed serous and mucinous acini with focal lymphocytic infiltrates surrounding the ducts. Connective tissue and lymph nodes from the right cervical region also showed changes of Kimura disease.

Discussion

The CT appearance of Kimura disease consists of three features: (1) diffusely enlarged parotid gland with a normal acinar pattern on CT sialography (CTS); (2) a soft-tissue mass

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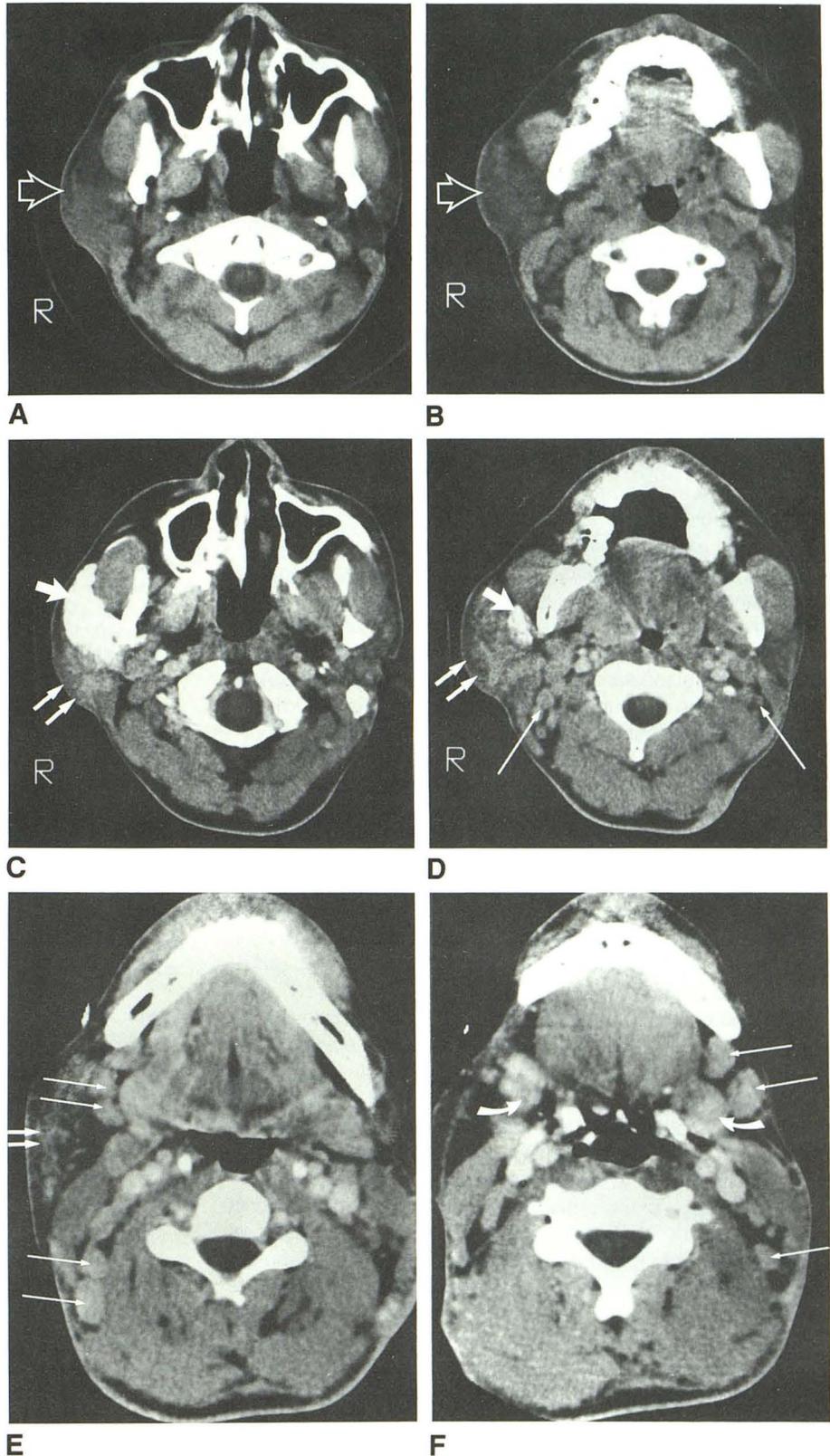
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Fig. 1.—A and B, Unenhanced scans show a soft-tissue mass in right parotid region (open arrow).

C and D, Scans with IV and parotid sialographic contrast at similar locations to A and B. Parotid acinar filling by sialographic contrast shows enlargement of gland and produces a very dense but normal contrast pattern within most of the parotid (short arrows), including the inferior pole as seen in D (short arrow). Rest of mass lies around lateral and inferior aspect of gland extending from periphery of the gland. It shows some patchy enhancement from IV contrast (medium-length arrows). Multiple deep cervical lymph nodes show uniform enhancement (long arrows).

E and F, Scans at level of submandibular glands show infiltration of connective tissue (short arrows in E) extending inferiorly from mass shown in D. Submandibular glands do not appear involved (curved arrows in F). Multiple lymph nodes in submandibular and deep cervical locations are shown (long arrows).



with an ill-defined border extending from the periphery of the gland; and (3) extensive lymphadenopathy showing uniform enhancement with IV contrast. This unusual combination may be mimicked most closely by a lymphoproliferative disease,

particularly malignant lymphoma. Other diseases differ in one or more features of their CT appearances.

Diffuse enlargement of salivary glands is seen in chronic sialadenitis, but CTS products patchy underfilling of the ducts.

Calculi, dilatation of the parotid duct, and retention cysts may also occur. Sialadenitis associated with autoimmune disease also produces a nonuniform and nonspecific pattern of underfilling on CTS. Parotid tumors may enlarge the gland but produce distinct filling defects on CTS within the gland and usually enhance with IV contrast. Involvement of the paraglandular soft tissue may occur with tumors, especially when they are recurrent or malignant, but the tumor mainly lies within the gland, with some extension into the surrounding connective tissue, rather than lying centered on the periphery of the gland.

Lymphadenopathy may be extensive in infective or inflammatory conditions, whereas nodes are usually less numerous in malignant disease. Lymph-node involvement by squamous cell carcinoma or by pyogenic and sometimes tuberculous infection show ring enhancement with IV contrast. Uniform enhancement similar to that seen with Kimura disease is seen with lymphoma and in some cases of tuberculosis. However, these diseases produce different changes in the salivary glands. Lymphoma usually shows as multiple nodules within the parotid gland, causing filling defects on CTS, and involvement of the salivary glands in tuberculosis is usually limited to secondary spread to the submandibular glands from adjacent involved nodes.

CTS in our case was particularly helpful in defining the limits of the glandular tissue and so separating the swollen gland from the surrounding abnormal tissue. It is not known whether IV contrast alone would have defined the two components so clearly. However, IV contrast was helpful in showing the enhancement pattern of the lymph nodes, and both routes of contrast administration yielded useful information.

There are three forms of treatment for Kimura disease: surgical debulking, radiotherapy, or drug therapy [1]. No one method is ideal. Good results have been claimed for radio-

therapy, with marked reduction in size or complete resolution of the disease. Follow-up over 1–4 years showed no recurrence. However, many clinicians are reluctant to treat a benign condition with potentially harmful ionizing radiation. Steroids and oxyphenbutazone have only a transient effect, and swelling recurs with cessation of either drug. Surgery is often difficult and, at this site, the facial nerve is at risk. The diffuse nature of the disease makes total excision impossible. However, recurrence rates appear low or zero over the short-to-medium term; in the absence of a better alternative, surgery is often the treatment of choice.

In summary, CT of the salivary glands appears to be useful in diagnosing Kimura disease because of the unusual combination of features. This method also delineates the extent of the disease necessary in planning treatment.

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