
Kimura Disease: CT and MR Findings

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Summary: The lesions of Kimura disease showed slightly high and very high intensity on T2-weighted MR, and low and intermediate intensity, respectively, on T1-weighted images. The degree of enhancement also differed between the two cases. These discrepancies may be attributable to differing degrees of fibrosis and vascular proliferation.

Index terms: Salivary glands, computed tomography; Salivary glands, magnetic resonance

Kimura disease is a rare entity that occurs primarily in Asian subjects (1, 2), characterized histopathologically by a lymph-folliculoid granuloma with infiltration of the mass and the surrounding tissues by eosinophils (3), often with concomitant peripheral blood eosinophilia and elevated serum IgE. We report two cases of Kimura disease and describe the appearance of the lesions on computed tomographic (CT) and magnetic resonance (MR) imaging, including MR with gadolinium enhancement.

Case Reports

Case 1

A 39-year-old man presented with a 3-year history of swelling and itching in the right parotid region. The peripheral blood count showed eosinophilia of 12%, and serum IgE was elevated to 1674 U/mL.

Sonography revealed an inhomogeneous low-echoic mass behind the right parotid gland and failed to show the interface between the mass and the gland. On CT, a soft-tissue-density mass was seen in the right parotid region. The mass extended subcutaneously over the sternocleidomastoid muscle. The margin between the mass and the normal parotid gland was indistinct. The density of right parotid gland was as low as that of the normal left parotid gland. On a CT sialogram done 3 months later, most of the right parotid gland showed a normal acinar pattern. The mass did not produce a distinct filling defect, but under-

filling of the acini was observed at the interface between the mass and the gland (Fig 1A). On MR done at 1.5 T, the T1-weighted images (500/18/2 [repetition time/echo time/excitations]) showed a low-signal mass and enlarged cervical lymph nodes. The right parotid gland was shown a high signal intensity like normal glands (Fig 1B). On T2-weighted images (2000/80/1), the mass displayed a signal of slightly higher intensity than that of the parotid gland. A high-signal area with a low-signal rim was detected in the mass. The enlarged cervical lymph nodes had high signal intensity (Fig 1C). On gadolinium-enhanced T1-weighted images, the mass and enlarged lymph nodes enhanced intermediately (Fig 1D).

At surgery, the mass and the right parotid gland were resected with regional lymph nodes. The high-signal area displayed in the mass on T2-weighted images proved to be lymph nodes. On histopathologic examination (Fig 1E), both the mass and the parotid gland showed extensive infiltration by eosinophils and lymphocytes, with germinal center formation, surrounded by thick fibrotic tissue. Most of glandular acini and ducts were destroyed by the granulomatous lesion, although some were spared. Histologically, a diagnosis of Kimura disease was made.

Case 2

A 17-year-old boy presented with a right-sided mass at the angle of the mandible. The peripheral blood count showed eosinophilia of 10%, and serum IgE was elevated to 9640 U/mL.

When he was 14 years old, the patient underwent resection of a mass adjacent to the right submandibular gland, and a diagnosis of Kimura disease was made. Post-contrast CT obtained at that time showed a well-enhanced mass adjacent to the right mylohyoid muscle and the submandibular gland. The right submandibular gland was spared. The right posterior auricular, bilateral superficial, and submental lymph nodes were enlarged and enhanced on CT and MR. At the present examination postcontrast CT showed a homogeneously enhanced mass in the same region. On MR, the T1-weighted images (500/20/2) displayed an intermediate signal mass adjacent to the right

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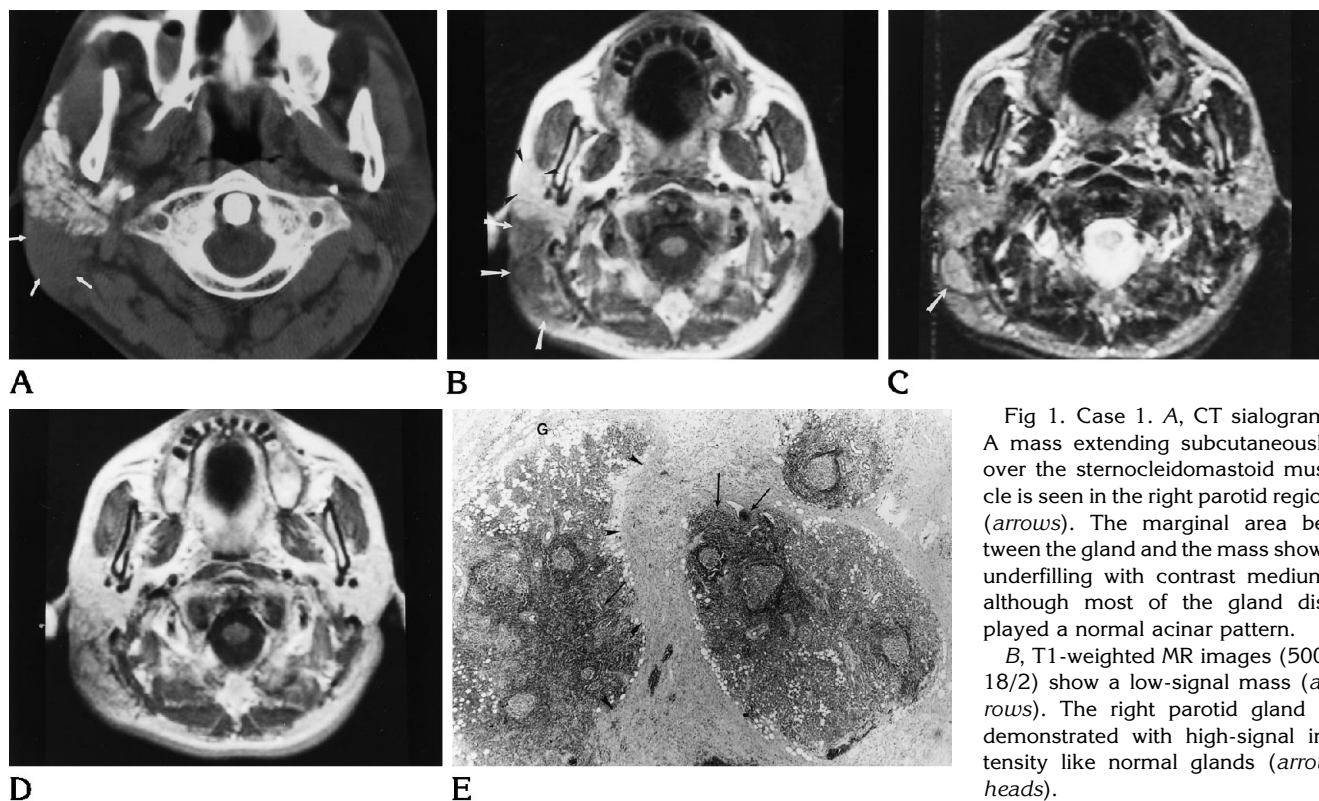


Fig 1. Case 1. A, CT sialogram. A mass extending subcutaneously over the sternocleidomastoid muscle is seen in the right parotid region (arrows). The marginal area between the gland and the mass shows underfilling with contrast medium, although most of the gland displayed a normal acinar pattern. B, T1-weighted MR images (500/18/2) show a low-signal mass (arrows). The right parotid gland is demonstrated with high-signal intensity like normal glands (arrow heads).

C, On T2-weighted images (2000/80/1) the mass shows an area of slightly higher signal intensity than the parotid gland. At surgery, the high-signal region with a low-signal rim proved to be an enlarged lymph node (arrow).

D, On gadolinium-enhanced T1-weighted images, the mass was well enhanced.

E, Micrograph of section from surgical specimen (hematoxylin and eosin, magnification $\times 8$). The interface between the mass and the gland shows extensive infiltration by eosinophils (arrows) and lymphocytes, with germinal center formation surrounded by thick fibrotic tissue (arrow heads). Some glandular acini are spared (G), whereas many acini and ducts are destroyed and replaced by lymphofolliculoid granuloma.

submandibular gland (Fig 2A). On T2-weighted images (3000/90/1), the mass and lymph nodes had a very high signal intensity (Fig 2B). On gadolinium-enhanced T1-weighted images, the mass showed marked enhancement (Fig 2C). Enlarged lymph nodes also enhanced.

At surgery, the mass was resected with some enlarged lymph nodes. On histopathologic examination (Fig 2D), the mass showed marked infiltration by eosinophils and lymphocytes and proliferation of many small vessels. The degree of fibrosis was less marked than in case 1.

Discussion

The histologic features of Kimura disease are proliferation of folliculoid structures with infiltration by mainly eosinophils, some plasma cells, lymphocytes, and mast cells. Some degree of vascular proliferation and fibrosis also is observed (3). The disease occurs primarily in Asian subjects and has male preponderance (greater than 80%) (1-3). Although the patients range in age from 3 to 71 years, 78% of the

cases occur in the second and third decades (1). The common clinical features of Kimura disease are an asymptomatic mass and local lymphadenopathy. Most lesions occur in the head and neck, especially in the parotid and submandibular regions. Other, less-frequent sites of involvement are axilla, groin, popliteal region, and forearm. The lesions commonly are found in soft tissues, but major salivary glands and lymph nodes frequently are involved. Characteristically, there is accompanying peripheral blood eosinophilia, which may reach 10% to 70%. Increased serum IgE concentrations (800 to 35 000 U/mL) also are observed (1-7). No one method is ideal for treating the disease. Some authors have found radiation therapy to be the most effective. However, many clinicians are reluctant to treat with radiation, because the disease is benign and mainly occurs in the young. On the other hand, complete resection is difficult because of the diffuse nature of the dis-

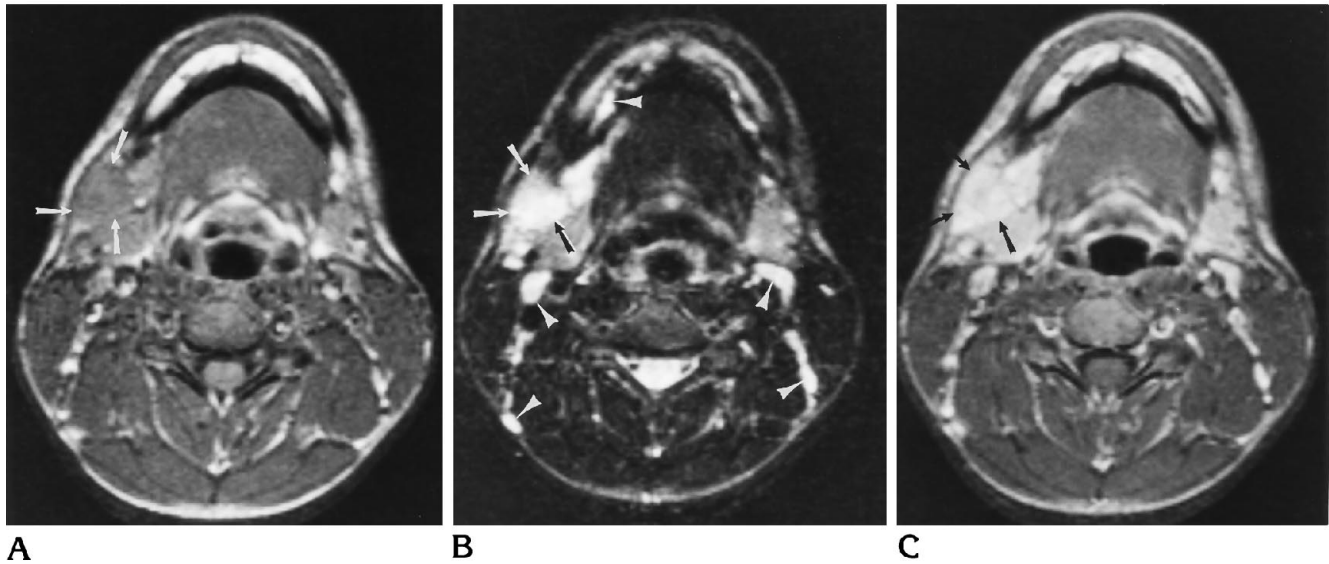


Fig 2. Case 2. A, On T1-weighted MR images (500/20/2), the mass (arrows) had a higher signal than muscle.

B, T2-weighted images (3000/90/1) displayed the mass (arrows) and lymph nodes (arrowheads) with very high signal intensity. The mass was adjacent to but not within the submandibular gland.

C, The mass was markedly enhanced (arrows).

D, Micrograph of section from surgical specimen (hematoxylin and eosin, magnification $\times 13.2$). The mass shows several germinal centers. Many small blood channels are demonstrated in the fibrotic tissue (arrows).



D

ease. Steroids and oxyphenbutazone cause rapid shrinkage of the lesion but have only a transient effect.

There are some reports on the radiologic characteristics of Kimura disease. According to three descriptions of its MR appearance (without gadolinium enhancement) (4–6), the signal appearances in Kimura disease vary. Our case 1 showed a slightly high-signal mass on T2-weighted images, whereas in case 2, a signal of very high intensity was observed. In case 1, there was significant fibrotic tissue, whereas in case 2, the degree of fibrosis was relatively mild. Further, case 2 displayed more small vascular channels in the mass than case 1. These histologic findings suggest that the signal difference on T2-weighted images may have been attributable to different degrees of fibrosis, and

the difference in vascular proliferation may have caused different degrees of enhancement. Smith et al (7) reported that on CT sialography, Kimura disease appeared as a diffusely enlarged parotid gland with a normal acinar pattern combined with a soft-tissue mass with an ill-defined border extending from the periphery of the gland. They considered that these CT appearances were useful in diagnosing Kimura disease, because this combination of features is unusual. In our case 1, the larger part of the parotid gland displayed a normal acinar pattern on CT sialography. However, underfilling with contrast medium was observed at the interface between the mass and the gland. This area showed higher signal intensity on T2-weighted MR than the normal gland, and the degree of enhancement was just as pronounced as that of

the mass. On histopathologic examination, salivary glands involved by the lesion exhibited destruction of glandular acini and ducts. This finding could explain the underfilling with contrast material. The narrow window setting on CT sialography might have obscured the underfilling area in the case of Smith et al (7).

It is difficult to diagnose Kimura disease on the basis of the radiologic appearance alone. This disease is manifested as an enhanced mass with subcutaneous extension and enlarged lymph nodes. Enhanced lymphadenopathy is observed in cervical tuberculous lymphadenitis. Most such cases have shown a central low density and peripheral rim enhancement (8), whereas enlarged lymph nodes enhance homogeneously in Kimura disease. Although enlarged lymph nodes also were observed in cases of acute sialadenitis, these acute inflammatory conditions give rise to clinical symptoms such as pain. Regarding malignant lesions, these tend to extend subcutaneously, and they display a filling defect with an ill-defined border on CT sialography (9). The CT sialographic and MR appearances in our case 1 mimicked those of an aggressive malignant tumor.

In conclusion, there are no accepted characteristic features of Kimura disease on CT or MR.

Both examinations showed a salivary gland mass and enlarged lymph nodes in the neck. The diagnosis is based on biopsy and on the patients' unique clinical features, such as eosinophilia and a high plasma IgE level.

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