Kikuchi Disease: CT and MR Findings

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Summary: Two cases of Kikuchi disease showed variable nodal enhancing features, including homogeneous enhancement and focal or extensive nodal necrosis on contrast-enhanced CT scans. At MR imaging, the area of central necrosis was isointense or hypointense on T1-weighted images and had a lower signal than nonnecrotic areas on T2-weighted images. The CT appearance of Kikuchi disease can be variable and can mimic not only lymphoma but various nodal diseases with nodal necrosis, including metastasis and tuberculosis.

Index terms: Neck, inflammation

Kikuchi disease, also known as histiocytic necrotizing lymphadenitis, is a self-limiting benign lymphadenopathy of unknown origin that predominantly affects young adults. The disease was described in 1972 by Kikuchi (1) and independently by Fujimoto et al (2). Kikuchi disease is characterized histopathologically by cortical and paracortical necrosis with a lymphoreticular infiltrate and absence of granulocytic infiltrates. Previous authors (3, 4) have reported enlarged, multiple lymph nodes that were homogeneously enhanced without evidence of gross necrosis at computed tomography (CT), as well as CT and clinicopathologic features that mimicked malignant lymphoma. We report two cases of Kikuchi disease in which enlarged nodes showed atypical enhancement, and describe the appearance of the lesions on CT scans and magnetic resonance (MR) images, including MR images obtained after contrast administration.

Case Reports

Case 1

A 35-year-old man had a 10-day history of general myalgia and fever, and a 5-day history of multiple cervical lymphadenopathy. The peripheral blood count showed mildly increased monocytes (13%) with a normal white blood cell count. Other laboratory results were negative, including serologic and autoimmune tests.

Contrast-enhanced CT of the neck revealed extensive lymphadenopathy on the right side of the neck. Enhancement was heterogeneous, and the nodes contained areas of enhancement with prominent areas of low density, suggesting gross nodal necrosis (Fig 1A). The size of the rim-enhancing nodes ranged from 0.5 cm to 2.5 cm in axial diameter on CT scans. Conglomeration of nodes and obliteration of adjacent fat plane were also seen on CT scans. Noncontrast T1-weighted MR images (500/30/2 [repetition time/echo time/excitations]), obtained at 1.5 T, showed enlarged nodes that were isointense or hypointense relative to muscle (Fig 1B); contrast-enhanced T1-weighted images showed variable enhancement, with both homogeneous enhancement and areas of low signal within the node or at its periphery (Fig 1C). On T2-weighted images (3000/80/1), the nonenhancing necrotic area was slightly higher in signal intensity than muscle, and the enhancing portions were brighter than the nonenhancing necrotic areas (Fig 1D).

A diagnosis of histiocytic necrotizing lymphadenitis was made on the basis of findings at excisional biopsy. On histologic examination (Fig 1E), the nodes showed multifocal overt necrosis with karyorrhectic debris and surrounding infiltrates of immunoblasts and histiocytes without granulocytic infiltration. The necrotic areas were revealed as fibrinoid necrotic material. After surgery, the patient received steroid medication, and, by the 2-month follow-up examination, enlargement of the cervical nodes had decreased markedly.

Case 2

A 37-year-old woman had a 1-month history of tender cervical lymphadenopathy and a 2-week history of fever. The laboratory data showed mild elevation of erythrocyte sedimentation rate, positive for IgM antibody of Epstein-Barr virus and weakly positive for antinuclear antibody.

Contrast-enhanced CT of the neck revealed multiple

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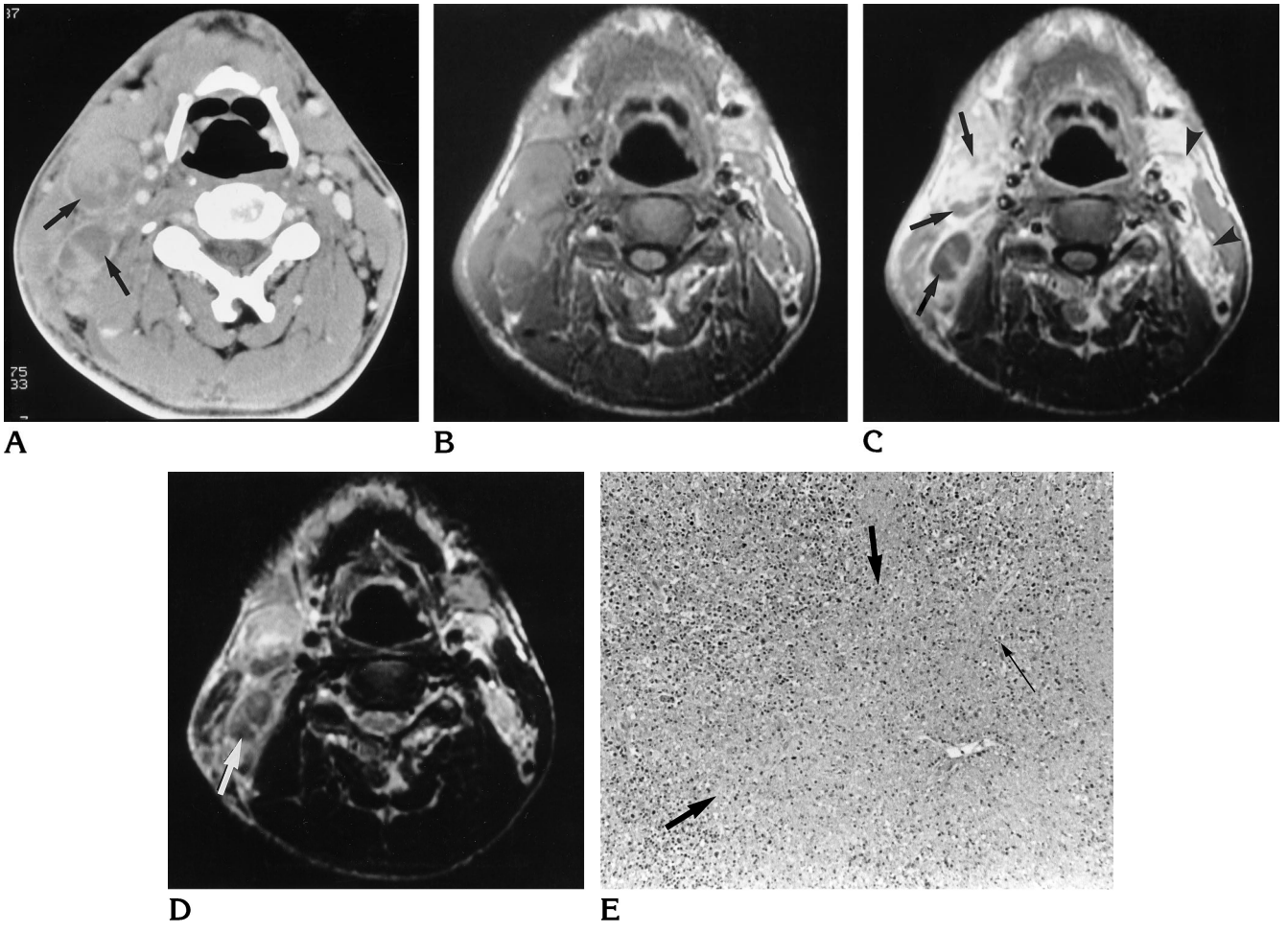

Fig 1. Case 1: 35-year-old man with 10-day history of general myalgia and fever, and 5-day history of multiple cervical lymphadenopathy.

A, Contrast-enhanced CT scan shows heterogeneous enhancement of enlarged cervical lymph nodes in right side of neck. The lymph nodes contain areas of enhancement with prominent areas of low density (*arrows*), suggesting gross nodal necrosis. Note the obliteration of perinodal fat in right posterior cervical space and adjacent superficial space.

B, On T1-weighted MR image (550/10/2), the enlarged nodes have similar or slightly lower signal intensity than muscle.

C, Contrast-enhanced T1-weighted image shows variable nodal enhancement. Areas of low signal are seen within the nodes or within the peripheral portion of the nodes (*arrows*), and homogeneous enhancement is also seen (*arrowheads*).

D, On T2-weighted MR images (4000/80/2), the nonenhancing necrotic area (*arrow*) is only slightly higher in signal intensity than muscle and lower in signal intensity than the areas that enhanced on the T1-weighted images.

E, Photomicrograph of surgical specimen of lymph node shows necrotic area (*thick arrows*) with karyorrhectic debris and nuclear dust (*thin arrow*) and no infiltration of granulocytes (hematoxylin-eosin, original magnification ×40).

left-sided internal jugular and spinal accessory lymphadenopathy. The various enhancing features included homogeneous enhancement, multifocal areas of central low density, and peripheral rim enhancement (Fig 2A and B). There was accentuation of fascial planes and increased density of fat surrounding the nodes without evidence of changes in subcutaneous tissue and skin.

Excisional biopsy was performed, and the pathologic diagnosis was histiocytic necrotizing lymphadenitis. On histologic examination, the nodes showed extensive necrosis with nuclear dust and infiltrates of immunoblasts and histiocytes without granulocytic infiltration (Fig 2C).

Discussion

Kikuchi disease is a benign form of lymphadenitis with a tendency to affect young women. The clinical features of the disease are variable, including nontender or tender cervical lymphadenopathy, fever, and leukopenia. Less frequent symptoms include weight loss, nausea, vomiting, night sweats, arthralgia, and hepatosplenomegaly (5–7). Involvement of skin and bone marrow have been reported in some cases (8, 9). The results of laboratory evaluations are

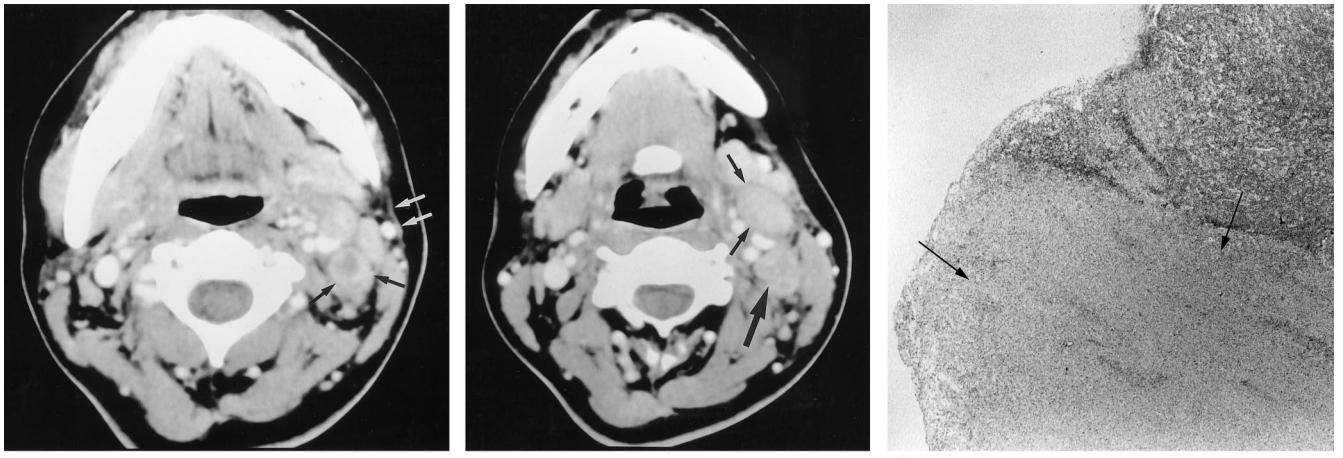

Fig 2. Case 2: 37-year-old woman with 1-month history of tender cervical lymphadenopathy and 2-week history of fever.

A, Contrast-enhanced CT scan shows a rim-enhancing node with central necrosis (*black arrows),* and adjacent homogeneously enhancing nodes in left side of neck. Accentuation of adjacent fascial plane is seen *(white arrows*).

B, CT scan at a lower level shows a homogeneously enhancing enlarged lymph node (*small arrows*) and a node with central small areas of low density (*large arrow*).

C, Photomicrograph of surgical specimen of lymph node shows extensive areas of necrosis with scattered nuclear debris (*arrows*) (hematoxylin-eosin, original magnification ×10).

usually normal or nonspecific, except for leukopenia with lymphocytosis. The pathogenesis of Kikuchi disease is still unclear, although there is some indication that it may be associated with viral infection or systemic lupus erythematosus (5, 8, 10). Although one death has been reported (11), the natural history of the disease is benign, with spontaneous resolution of the cervical lymphadenopathy usually occurring within 1 to 4 months, without treatment.

The chief pathologic features of histiocytic necrotizing lymphadenitis are variable degrees of necrosis in cortical and paracortical areas, with prominent karyorrhectic debris, proliferation of histiocytes and immunoblasts surrounding the area of necrosis, and absence of granulocytes. The pathologic features may mimic malignant lymphoma by the reactive nature of the cells and the polymorphous cellular infiltrates (4, 6).

In one case, Fulcher (3) reported that contrast-enhanced CT revealed multiple enlarged lymph nodes that showed homogeneous enhancement without evidence of necrosis, and an absence of inflammatory change in the adjacent tissue. Kim et al (4) also reported a case in which contrast-enhanced CT showed homogeneous enhancement of lymphadenopathy in the neck and mediastinum, and an accentuation of the fascial planes of the neck and mediastinum, consistent with perinodal extension. These reports indicate that the CT features of Kikuchi disease may mimic those of malignant lymphoma, owing to their homogeneously enhancing character without nodal necrosis. However, in our two cases, CT showed variable enhancing features. In case 1, peripheral rim enhancement or focal central low attenuation was seen in 80% of all enlarged nodes on CT scans. In case 2, variable enhancement of enlarged nodes and rim enhancement was present in 25% of all enlarged nodes on CT scans. In both cases, CT showed accentuation of fascial planes and obliteration of perinodal fat, consistent with previous reports (4). The peripheral rim-enhancing features of lymph nodes on CT scans suggest macroscopic necrosis. The nodal necrosis of Kikuchi disease is usually microscopic; however, the degree of necrosis is variable, and overt macroscopic necrosis can also be present (6, 12, 13). Therefore, the enhancing features of Kikuchi disease may vary significantly according to the degree of nodal necrosis, as shown in our cases. Our two cases suggest that the CT appearance of Kikuchi disease may mimic not only lymphoma but also various nodal diseases with nodal necrosis, including metastasis and tuberculosis.

The MR imaging features observed in our case 1 are of some interest. The signal intensity

of the necrotic area was only slightly higher than muscle on T2-weighted images and similar to or slightly lower than muscle on T1-weighted images, whereas the homogeneous enhancing portion showed higher signal intensity than the necrotic areas on T2-weighted images. The relatively low signal intensity of the necrotic area on T2-weighted images is different from the usual bright signal intensity of the necrotic area in metastatic nodes or benign nodal disease, such as pyogenic abscess or tuberculosis (14). When the necrosis is overt, eosinophilic fibrinoid material is present at the center of necrotic focus with abundant fragments of nuclear debris (5, 6). Although the cause of relatively low signal intensity of the necrotic area on T2-weighted images is not clear, we speculate that this may reflect restricted mobile protons within high protein content in fibrinoid material of necrotic focus, or it may be associated with high phagocytic activity of proliferated histiocytes.

In conclusion, the CT appearance of Kikuchi disease may be variable, mimicking various nodal diseases, such as tuberculosis, metastasis, and lymphoma. We observed that the necrotic area of the nodes had a lower signal than the nonnecrotic portion on T2-weighted MR images, which is different from the usual bright signal intensity of nodal necrosis seen in other nodal diseases.

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