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CT Findings of Kimura's Disease Involving Thorax: Case Report[1]

In Jae Lee, M.D., Hyung Sim Choe, M.D., Soo Kee Min, M.D.[2], Eun Young Ko, M.D., Jae Young Lee, M.D., Hyun Beom Kim, M.D., Kwan Seop Lee, M.D., Yul Lee, M.D., Sang Hoon Bae, M.D.

Kimura's disease is a benign chronic inflammatory condition with good prognosis, commonly involving the head and neck area. With regard to thoracic manifestation, however, the information contained in the literature is limited. The diagnosis of Kimura's disease on the basis of imaging is difficult, especially in thoracic cases manifesting as lymphadenopathy. The purpose of this report is to illustrate and discuss a thoracic case of the disease manifesting as lymphadenopathy.

Index words : Chest, CT
Chest, lymphadenopathy
Kimura's disease

Kimura's disease is a benign chronic inflammatory disease of unknown etiology that occurs primarily in Orientals of young to middle age (1 - 4). Patients usually present with subcutaneous or soft tissue masses of the head and neck, often with involvement of the salivary glands and regional lymph nodes (5). The condition is characterized pathologically by the presence of lymph-folliculoid granuloma with infiltration of the mass and surrounding tissues by eosinophils (1 - 4). Since its prognosis is good, accurate diagnosis after lymph node biopsy may spare patients from unnecessary radical surgery (1, 5). Thoracic manifestation of the disease is known to be rare (6). The purpose of this report is to describe a case of Kimura's disease involving the thoracic area and demonstrated by computed tomography (CT).

Case Report

A 19-year-old man presented with a right supraclavicular mass, which first appeared two weeks earlier. Physical examination revealed a firm, non-tender, 2×3-cm mass in the right supraclavicular area. Blood tests revealed a slightly elevated white blood cell count (9600 - 13600/mm^3), though there was no significant peripheral eosinophilia (3.2%) or elevation of serum IgE (40.7 IU/ml).

Initial chest radiographs depicted anterior mediastinal widening, and contrast-enhanced CT performed on the same day revealed aggregated lymphadenopathy extending from the anterior mediastinum to the right axillary and supraclavicular areas (Figs. 1A, B). The lesion was mildly enhanced and relatively homogeneous in nature. There was, moreover, no internal focus of low attenuation, indicating the absence of necrosis, and no evidence of infiltrative change in surrounding tissue or other mass formation.

The right supraclavicular mass was excised and was found to consist of enlarged lymph nodes, with aggregation. It was well encapsulated without surrounding infiltration. Histopathologic examination showed that it consisted of enlarged lymph nodes with hyperplasia of its germinal center, accompanied by vascularization; multifocal dense eosinophilic infiltration and a focal small eosinophilic abscess were also present (Fig. 1C). Pathologic diagnosis was consistent with Kimura's dis-

[1]Department of Radiology, Hallym University Sacred Heart Hospital
[2]Department of Pathology, Hallym University Sacred Heart Hospital
Received January 14, 2003 ; Accepted March 11, 2003
Address reprint requests to : In Jae Lee, M.D., Department of Radiology, Hallym University Sacred Heart Hospital, 896, Pyungchon-dong, Dongan-ku, Anyang-city, Kyungki-do 431-070, Korea.
Tel. 82-31-380-3885 Fax. 82-31-380-3878
E-mail: ijlee2003@yahoo.co.kr

ease. Because the case was a mild one and the condition benign, medical treatment with steroid was not attempted after surgery.

About four months later, enhanced CT showed marked progression of the disease, with extensive mediastinal, axillary, supraclavicular and pericardial lymphadenopathy simulating lymphoma, metastasis or tuberculosis (Figs. 1D, E). A small mildly enhanced nodular lesion involving the right parotid gland later developed, and radiation therapy and medical treatment with

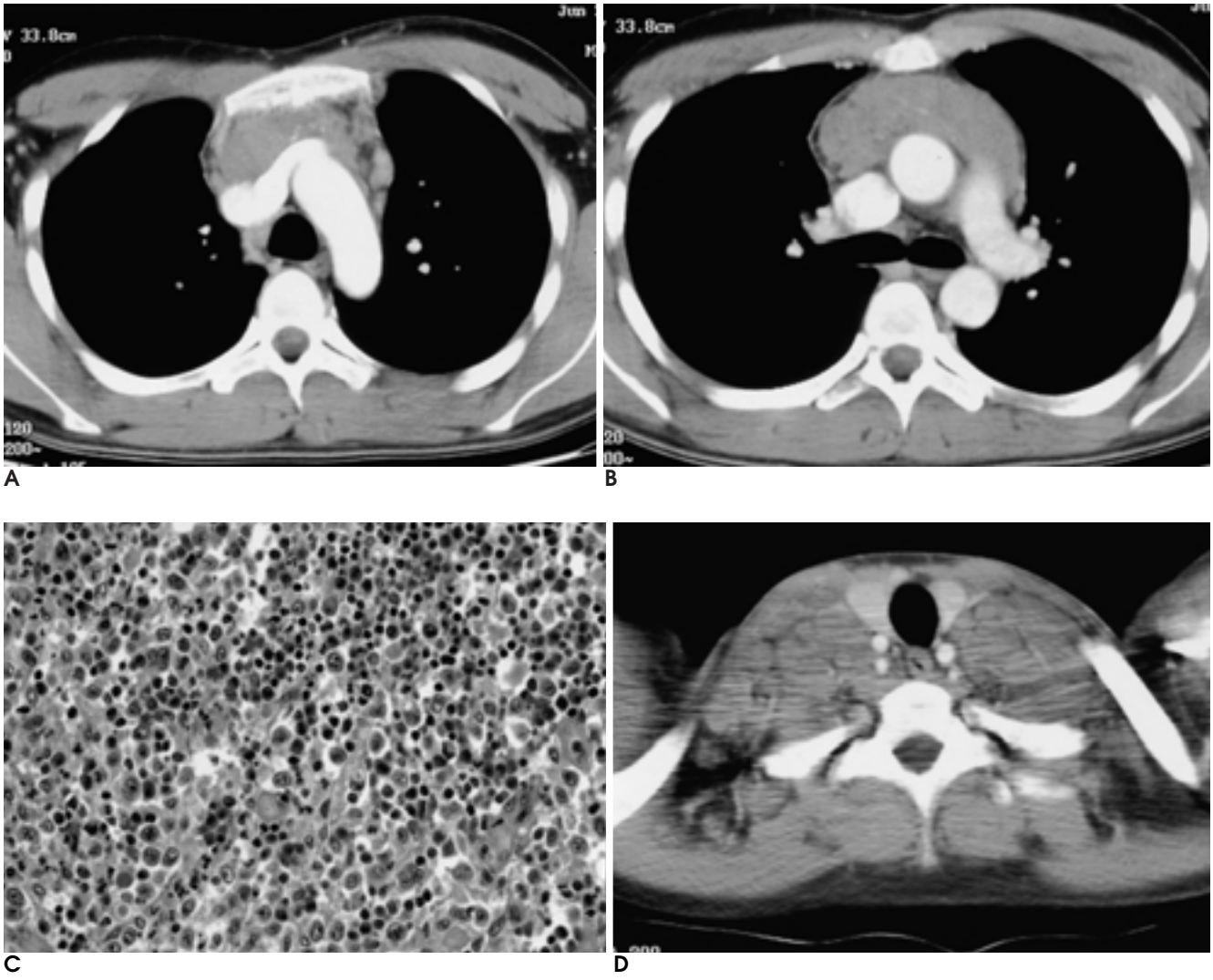

Fig. 1. 19-year-old man with Kimura's disease involving thorax.
A, B. Enhanced CT scans show mildly enhanced multiple lymph nodes in anterior mediastinal and right axillary areas.
C. Photomicrograph of section from excised lymph node reveals dense infiltration of eosinophils with focal small eosinophilic abscess (H & E stain, ×200).
D, E. Four months later without specific treatment, enhanced CT scans show marked progression of lymphadenopathy in mediastinal, axillary and supraclavicular areas.

steroid were attempted. The result was very satisfactory, with the lymphadenopathy showing marked regression.

Discussion

Kimura's disease was described in 1948 by Kimura et al. as "unusual granulation combined with hyperplastic changes in lymphoid tissue" (4). The predominant histologic feature of the condition is the proliferation of folliculoid structures, with infiltration mainly by eosinophils, some plasma cells, lymphocytes, and mast cells (3). Involved lymph nodes usually demonstrate florid reactive follicular hyperplasia, increased vascularity, and marked eosinophilia of the paracortical region (4). The disease occurs primarily in Orientals, especially those of Chinese or Japanese extraction (5). Men are more commonly affected than women, and the male-to-female ratio is greater than 3:1 (4). Although reported cases have involved patients ranging in age from 1 to 76 years, most have occurred during the second and third decades of life (3, 5, 7 - 9). The common clinical features of the disease are the presence of an asymptomatic mass and local lymphadenopathy. Most lesions occur in the head and neck, especially in the parotid and submandibular regions (1, 3, 5), though other less frequent sites of involvement are the axilla, groin, popliteal region, and forearm (3). In our case, the site of the primary lesion was the thorax, with no head involvement but later involvement of the parotid gland. Nodal involvement is characterized by eosinophilic infiltration and the presence of microabscess, increased numbers of small blood vessels, peripheral eosinophilic infiltration, and eosinophilic folliculosis (2, 5). Peripheral blood eosinophilia (10 - 70%) is frequently present (3, 4), and increased serum IgE concentrations (800 - 35,000 U/ml) are also observed (4). The IgE receptor is present in the germinal center and on follicular dendritic cells (5, 10), suggesting a close relationship with the IgE immune response, which may play an important role in the pathogenesis of the disease (5, 10). Lesions tend to recur and therapy usually consists of either local surgical extirpation or local irradiation with 25 to 30 Gy (5). Because the disease is benign and occurs mainly in the young, many clinicians are, however, reluctant to use radiotherapy (3, 5). Due to the diffuse nature of the disease, complete resection is, on the other hand, difficult (3), and transient local relief has been achieved using steroids (3, 5).

The radiologic findings of Kimura's disease involving the head and neck areas have been documented; the reported CT findings include the presence of an enhanced mass involving enlarged major salivary glands, with subcutaneous extension, and enlarged regional lymph nodes (3, 5, 9). The presence of mild enhancement is suggestive of an inflammatory process (5, 9). Nodal manifestations in Kimura's disease do not usually include peripheral rim enhancement, internal necrosis, perinodal extension or strong enhancement (1, 3, 5). Our case showed extensive, aggregated lymphadenopathy in the anterior mediastinum, axilla, upper anterior chest wall and supraclavicular area; other than this, there was no evidence of mass formation, and there was no initial involvement of head and neck areas. The lesion was relatively homogeneous and mildly enhanced, without areas of internal low attenuation suggesting nodal necrosis. According to reported descriptions of the MR imaging features of head and neck lesions, the signal intensity varied (3, 5). At T2-weighted imaging, lesions involving the mass and lymph nodes displayed slightly high to very high signal intensity, which was attributed to infiltration by eosinophils and lymphocytes, the proliferation of many small vessels, and different degrees of fibrosis (3, 5). High signal intensity at T1-weighted imaging, suggestive of methemoglobin, was noted within lesions (5). Differences in vascular proliferation may have caused different degrees of enhancement of the lesion, and flow voids (3, 5).

It is difficult to diagnose Kimura's disease on the basis of radiologic appearances alone; in patients of Oriental extraction, the existence of a salivary gland mass with cervical lymphadenopathy may suggest its presence (3, 5), though patients may occasionally present with isolated lymphadenopathy (4). In thoracic cases, diagnosis of this disease on the basis of imaging findings can be very difficult, since the radiologic appearance of nodal manifestation is similar to that of lymphoma, tuberculosis, metastasis, drug-induced lymphadenopathy, and Castleman's disease (1, 3, 5). Peripheral blood eosinophilia with elevated serum IgE levels may be helpful in differential diagnosis, though definite diagnosis can only be made by biopsy. In conclusion, although the radiologic findings of Kimura's disease are nonspecific, its possibility should be considered where an Oriental man exhibits mildly enhancing mediastinal and cervical lymphadenopathy, especially if accompanied by peripheral blood eosinophilia.

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흉부에 발생한 Kimura병의 CT 소견: 증례 보고[1]

[1]한림대학교 의과대학 성심병원 방사선과
[2]한림대학교 의과대학 성심병원 병리과

이인재 · 최형심 · 민수기[2] · 고은영 · 이재영 · 김현범 · 이관섭 · 이 열 · 배상훈

Kimura병은 예후가 좋은 양성의 만성적인 염증성 질병으로서 흉부에 생기는 경우는 드물다. 특히, 흉부의 종격동에서 임파절 비대로만 나타날 때는 감별 진단이 어렵다. 이 증례 보고의 목적은 흉부의 종격동, 쇄골상부, 액와부 등에서 임파절 비대로 나타난 Kimura병의 CT 소견을 기술하는데 있다.