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Kimura's Disease in the Arm

— A Case Report —

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= **Abstract** =

Kimura's disease is an uncommon, chronic inflammatory disease of unknown etiology. It is an important category of reactive lymphadenopathy in the oriental population. The most common sites are the subcutis of the head and neck, and parotid gland. It's clinical course is benign nature. The treatment modalities for this disease are steroid therapy, radiation therapy and surgical excision.

We experienced a case of soft tissue mass in the left arm. It was slightly tender and relatively movable. We excised the mass, which was turned out to be Kimura's disease on microscopic examination.

Key Words : Kimura's disease, Arm

INTRODUCTION

Kimura's disease is an uncommon, chronic inflammatory disease of unknown etiology[5,7]. This disease develops in the subcutaneous tissue of the head and neck, salivary glands, and inguinal regions[1,4,5,7,14]. It occurs most often in young males. Men are more commonly affected than women, and the male-to-female ratio is greater than 3:1[12].

It was confused with Angiolymphoid Hyperplasia

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with Eosinophilia(ALHE) because it resembles ALHE histologically. In 1948, Kimura et al[1,3,11] first described that the disease has different histological and clinical nature, and represented that these two diseases are different entities.

In Korea, Kimura's disease was first reported by Kim et al[9]. in 1975. From 1975 to 1992, 19 cases of Kimura's disease and ALHE have been reported[13].

We experienced a case of soft tissue mass in the left arm that was excised and pathologically diagnosed as Kimura's disease.

CASE REPORT

A twenty-two-year-old man presented with a large soft tissue mass in the left arm for two years. The mass was slowly enlarged. Two months ago, pain was developed and gradually increased even during bed resting time.

Past history and family history were not contributable.

1. Physical findings

He was healthy except for solitary soft tissue mass which was palpated in the medial aspect of left distal arm. It was 8x3cm in size, rubbery hard, relatively movable, and slightly tender. Fluctuation and local heat were not seen. Regional lymphadenopathy was noted around the main mass, especially toward the axilla. There was mild hypesthesia in the elbow joint and anteromedial aspect of the forearm. There were no general or regional skin abnormalities.

2. Laboratory fingings

1) Hematology : Hemoglobin - 12.6gm/dl, Hematocrit - 37%, WBC - 8900/mm^3, BUN/Cr - 8.6/0.8, Proteinuria - negative.

2) Simple radiography : No specific findings (Fig. 1-A, B).

3) MRI : Mass that is 10x4x3cm sized, spindle-shaped, and located at posteromedial neurovascular space of the left distal arm. It looked like encircle the basilic vein, brachial artery and vein.

This mass reveals isosignal intensity in T_1-weighted image, diffuse high signal intensity in proton and T_2-weighted image, and homogeneous enhancement in enhanced T_1-weighted image(Fig. 2-A, B).

3. Operative findings

In supine position, we made a curvilinear skin incision on the anteromedial aspect of distal arm. We found the mass between the biceps brachii muscle and the triceps brachii muscle, which was 8x4x2cm sized, elliptical in shape, and well encapsulated.

It encircled the medial antebrachial cutaneous nerve and basilic vein, and the ulnar nerve was well demarcated posteromedially from the mass, median nerve and brachial artery anteromedially. Adhesion with surrounding deep fasciae was found and made en-bloc excision difficult, but possible.

It was hypervascular, darkish-red color, and rubbery hard. Regional lymph nodes ardound the main mass were enlarged.

4. Pathologic findings

Grossly, it was a pinkish white nodular firm mass which encircled medium sized blood vessels and measures 8x4x3cm. The cut surface is pinkish white and fish flesh appeared. There was no necrosis or hemorrhage in the specimen.

Microscopic examination revealed marked hyperplasia of germinal centers which were well vascularized and contain interstitial fibrosis. There were also extensive infiltration by mature eosinophils, with formation of eosinophilic abscesses. An increase in the number of plasma cells and mast cells was noted in the interfollicular areas. There were no cellular atypism or pleomorphism(Fig. 3A-C).

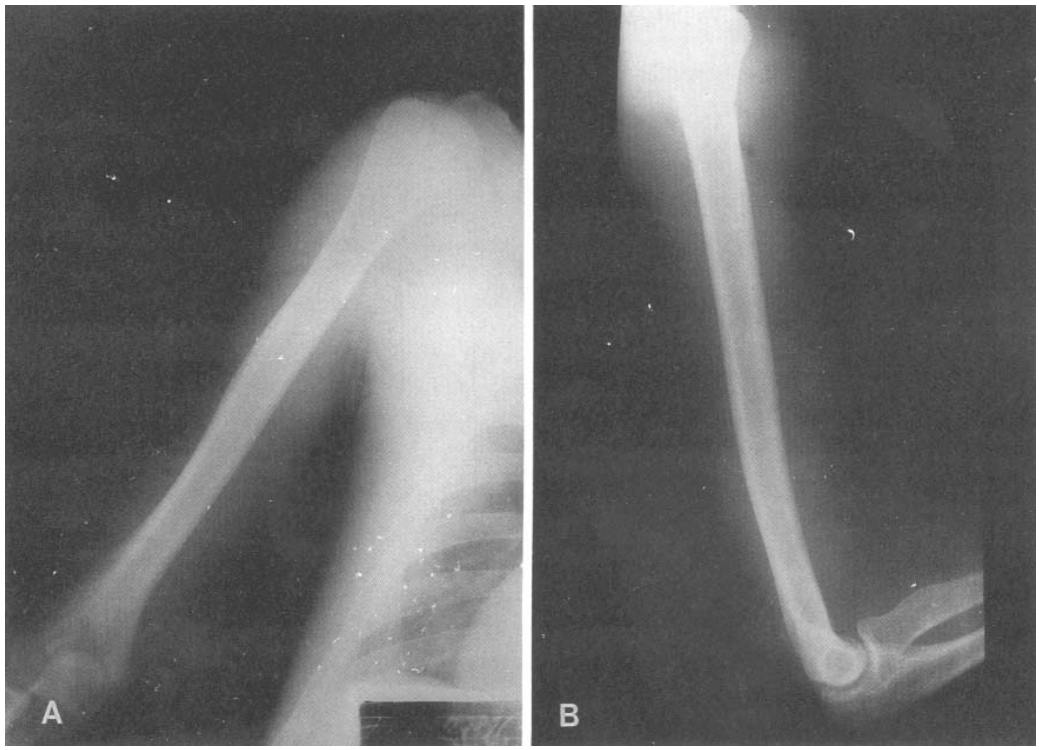


Fig. 1. The plain X-ray shows normal bone and soft tissue shadow
A. left humerus AP view.
B. left humerus lateral view.

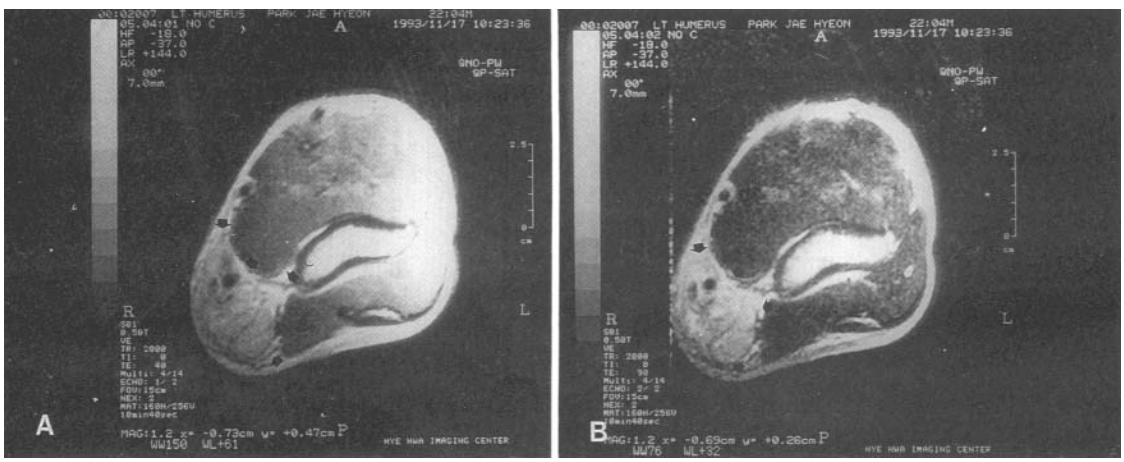
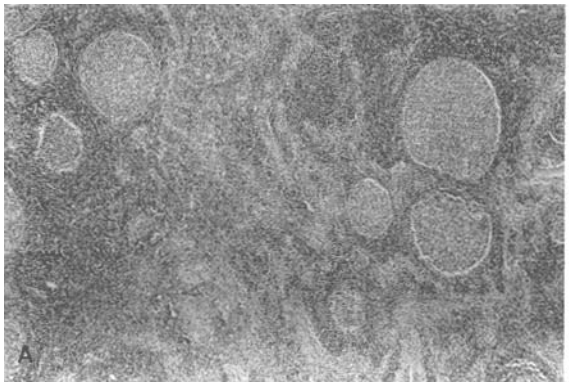
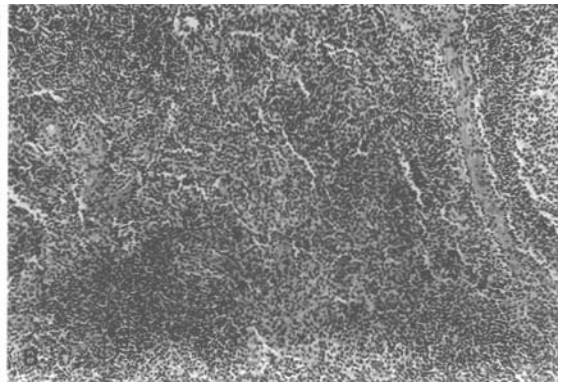


Fig. 2. A. T_1-weighted image shows that the mass is isosignal intensity.
B. T_2-weighted image shows that the mass is diffuse high signal intensity.





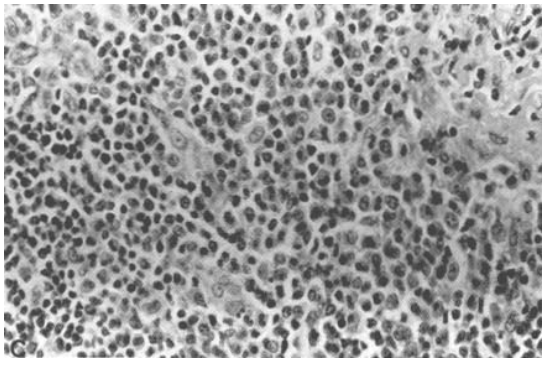


Fig. 3. A. The microscopic view shows prominent proliferation of blood vessels between the lymphoid follicles (X40, hematoxyline and eosin stain).

B. The high power view of interfollicular areas reveals prominent blood vessels proliferation(X100, hematoxyline and eosin stain).

C. The photograph shows many eosinophils mixed wiht plasma cells and lymphocytes, forming eosinophilic abscess(X400, hematoxyline and eosin stain).

5. Postoperative findings

At postoperative second day, hematoma was formed at the postoperative dead space because compression dressing was applied inadequately, so that ulnar and median nerve compression symptoms were developed. The symptoms were subsided three month later and no evidence of recurrence was identified at ten month after the operation.

DISCUSSION

Kimura's disease, a chronic inflammatory condition of unknown cause[5,7], is endemic in Orientals especially, Japan, China, and Hongkong[4,5,7,13,15]. It was first described in China by Kimm and Szeto(1937)[10], initially called "eosinophilic hyperplastic lymphogranuloma". Then considered by Kimura et al.(1948)[3,11] as "unusual granuloma with proliferation of lymphoid tissue".

Many similar cases have been reported under a variety of names such as angiolymphoid hyperplasia with eosinophilia, inflammatory angiomatous nodule, pseudopyogenic granuloma, atypical pyogenic granuloma, papular angioplasia, subcutaneous angioblastic lymphoid hyperplasia with eosinophilia, nodular angioblastic hyperplasia with eosinophilia and lymphofolliculosis, intravenous atypical vascular proliferation, histiocytoid hemangiona, and epithelioid hemangioma[4,14].

Rosai et al.(1979)[1,5,11,15] first described that ALHE and Kimura's disease represented two seperate disease entities, because of the histopathological differences. Burrall et al.(1982)[1] and, Enzinger and Weiss(1983)[1] described the difference of these two diseases as follows :

Clinically, ALHE was presented as multiple small papules or nodules in older adults with short duration(median duration : 1.5 years), and not accompanied by lymphadenopathy and peripheral blood eosinophilia. In contrast, Kimura's disease occured mainly in young men who presented with

a tumorous swelling of the subcutaneous soft tissue. Disease duration was longer than ALHE (median duration : 4 years) and frequently accompanied by regional lymphademopathy, peripheral blood eosinophilia, and elevated serum IgE[2,3,5,11,14].

In this case, regional lymphadenopathy was noted and there was no eosinophilia on two consecutive samplings. Serum IgE level was not checked.

The most important histopathologic difference is the blood vessel components of the two diseases.

In ALHE, in addition to the thin-walled blood vessels, thick-walled blood vessels with the so-called "histiocytoid" or "epithelioid" endothelial cells were characterized by hypertrophied or vacuolated endothelial cells protruding into the vascular lumen or even occluding the lumen. The blood vessels in Kimura's disease showed either flattened or cuboidal endothelial cells, and mast cells. Mast cells and plasma cells are abundant. Vacuolated endothelial cells have never been observed in this condition[1,3,12].

In Kimura's disease, eosinophilic infiltration of the germinal centers, causing progessive destruction, result in folliculolysis[5,12]. But, never in ALHE.

IgE in the germinal centers of Kimura' s lesion may be an important differential feature of Kimura's disease.

Sclerosis is another distinctive feature of Kimura's lesion, which may cause adhesion of lymph node to adjacent structures[3,7], as in our case. Sclerosis occurs most frequently in areas where eosinophilia is most marked, i.e., the paracortex.

The deposits of IgE in the germinal centers, the elevated serum IgE, and peripheral blood eosinophilia suggest that Kimura's disease is atopic in nature. So it is probably an immurologically mediated disorder[5,11,14].

Conclusively, Kimura's disease presents inflammatory process, but ALHE presnet benign tumorous condition of vascular endothelial cells[4].

Kimura's disease is usually a localized process without systemic manifestation. The only associated condition that has been reported is nephrotic syndrome with proteinuria (about 12%)[2,5,6,7,14]. In this case, proteinuria was not noted in the urinalysis.

The treatment modalities for Kimura's disease are steroid therapy[6], radiation therapy[8], and surgical excision[7,15].

The clinical course is benign and spontaneous regression is often seen[8,15].

The main treatment for persistent disease is oral corticosteroid therapy, which is remarkably effective in most cases.

If refractory and persistent even under steroid therapy, radiation therapy can be indicated. Total dose of 20 to 30 Gy with conventional fractionation scheme[8] seemed sufficient for local control in most cases. No secondary malignancy in the radiated area has so far been observed.

Disease recurrence is uncommon as treated with surgical excision[7,15]. In this case, disease recurrence was not found ten months after the operation.

CONCLUSION

We experienced a case of Kimura's disease in the left arm and surgically excised without recurrence during postoperative ten months, so we report this case with references.

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= 국문 초록 =

상완부에 발생한 Kimura씨 병

- 1례 보고 -

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Kimura씨 병은 흔하지 않은 만성 염증성 질환으로서, 그 원인에 대해서는 정확히 알려지지 않은 상태이다. 이 질환은 동양인에 있어서 반응성 임파선염을 일으키는 질환의 일종으로 분류되어 있으며, 주로 두경부의 피하조직 및 이하선 등에 호발한다. 비교적 양호한 경과를 보이며, 그 치료방법으로는 스테로이드 투여법, 방사선조사 및 수술적 절제술 등이 있다.

저자들은 좌측상완부에 발생한 경한 압통과 비교적 유동성이 있는 연부조직종양 1례에 대한 전절제 생검술을 시행하여 Kimura씨 병으로 확진하였으며, 술후 10개월 추시상 재발소견은 없었다.