

KIMURA'S DISEASE

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Summary

This paper describes 6 cases of Kimura's disease diagnosed in the General Hospital, Kuala Lumpur. Kimura's disease is a chronic inflammatory disorder of unknown aetiology characterised by lymphoid follicles, vascular proliferation and marked eosinophilic infiltration. The clinical presentation and pathological features are outlined. A brief review of available literature and a comparative study of cases diagnosed as angiolymphoid hyperplasia with eosinophilia in the Western world are presented.

Key words: Kimura's disease, inflammation, angiolymphoid hyperplasia, eosinophilia.

INTRODUCTION

In 1948, Kimura *et al.* presented a paper entitled "On an unusual granulation with hyperplastic changes of lymphatic tissue". The condition subsequently became widely known as "Kimura's disease." However, the earliest report was from Kim and Szeto who reported 7 cases of "eosinophilic hyperplastic lymphogranuloma" in 1937.² In the English literature, the earliest report was from Wells and Whimster³ in 1969 describing 9 cases of "subcutaneous angiolymphoid hyperplasia with eosinophilia". Similar conditions have since been reported from both the East and the West under various names.

Kimura's disease is a distinctive chronic inflammatory condition of unknown etiology. It is characterised by lymphocytic and eosinophilic infiltration associated with vascular proliferation and fibrosis. The aim of this paper is to describe cases encountered in this centre and to compare them with cases described from elsewhere in the world.

MATERIALS AND METHODS

Between October 1983 and June 1985, 6 cases of Kimura's disease were diagnosed in the General Hospital, Kuala Lumpur (GHKL). The ages of the patients ranged from 15 to 50 years. There were 4 males and 2 females.

Two cases (both young males) were from West Malaysia and presented with parotid swellings which were surgically removed and processed routinely for histopathological examination. The remaining 4 cases were from Sarawak. They presented with subcutaneous swellings in the region of the head and neck.

Only histopathological slides of these lesions were received and available for study. In all instances, the histological details of the lesion, such as the nature and pattern of inflammatory infiltration, vascular changes and fibrosis were noted.

RESULTS

A summary of clinical data of all the 6 cases are listed in Table 1. Blood eosinophilia was noted in the 2 West Malaysian patients. Clinical details of the cases from Sarawak were not available.

Gross specimens from the parotid glands of the 2 West Malaysian cases were fleshy with areas of smooth lobulation along the borders. However, in areas the lesions had vague outlines and the surgical planes between lesion and normal tissue were not easily defined (Fig. 1).

Histological examination of routine paraffin-embedded tissue was carried out in all 6 cases. Microscopically, the lesions appeared similar but showed variation in the amount of lymphoid and fibrous tissue present as well as the maturity of endothelial proliferation. The details are shown in Table 2. The histological features can be described under the following headings:

1. Inflammatory infiltrate

- (i) *Lymphocytic infiltration* was a constant feature which either occurred diffusely or in clusters.
- (ii) *Lymphoid follicles* were a prominent feature in all cases (Fig. 2) except in one case in whom the duration of the lesion was not exactly known.

TABLE 1
DEMOGRAPHIC AND CLINICAL DATA OF 6 PATIENTS WITH KIMURA'S DISEASE

Case	Age (yrs.)	Sex/Race	Site	Size	Duration	Treatment
1	46	F/Ch.	Ear lobe	5.0 x 6.0 cm.	1 year	Excision
2	50	F/Mly	Cheek	6.0 x 6.0 cm.	?	Excision
3	20	M/Mly	Forehead	1.2 x 0.8 cm.	A few weeks	Excision
4	47	M/Iban	Neck	4.0 x 3.0 cm.	5 months	Excision
5	16	M/Mly	Parotid	7.0 x 6.0 cm.	3 years	Excision
6	15	M/Ch.	Parotid	10 x 10 cm.	7 years	Excision

Ch = Chinese ; Mly = Malay.

TABLE 2

THE HISTOLOGICAL FEATURES OBSERVED IN 6 PATIENTS WITH KIMURA'S DISEASE

Case	Lymphoid Follicle	Vascular Channels	Vascular Patency	Fibrosis	Eosinophils & Histiocytes	Eosinophilic Abscess
1	Moderate	Numerous with prominent endothelium	Patent & non-patent	Moderate	Eosinophils +++ Histiocytes +	Present
2	A few	Numerous with flattened endothelium	Patent Arteritis*	Marked	Eosinophils +++ Histiocytes +	Present
3	Moderate	Numerous with prominent endothelium	Patent	Mild to moderate	Eosinophils +++ Histiocytes +	Present
4	Prominent	Numerous with prominent endothelium with atypia	Mainly non patent	Minimal	Eosinophils ++ Histiocytes +	Present
5	Moderate	Moderate number	Patent	Marked	Eosinophils ++ Histiocytes +	Present
6	Moderate	Moderate number	Patent	Moderate	Eosinophils ++ Histiocytes ++	Present



FIG. 1 : Photograph showing macroscopic appearance of parotid gland with Kimura's disease. Note that the surgical plane is not easily defined.

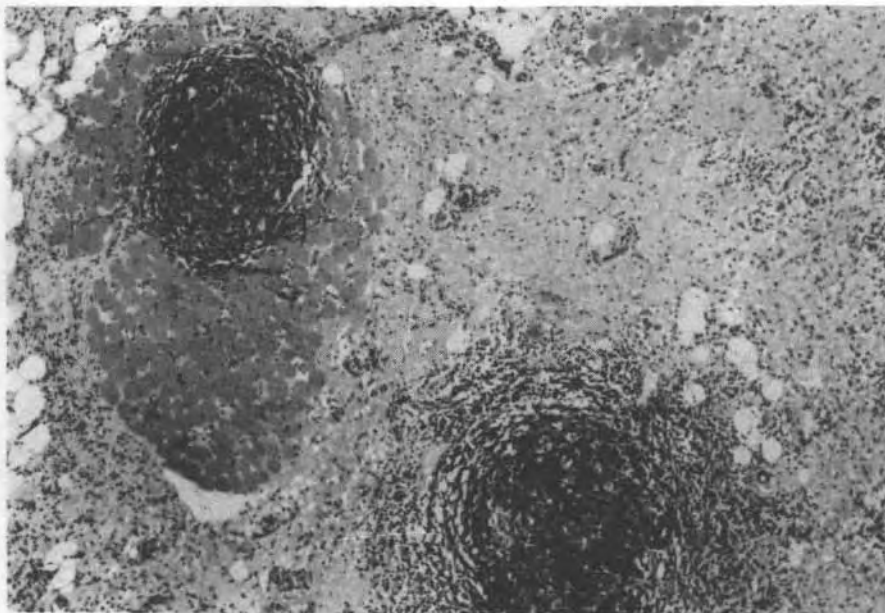


FIG. 2 : Kimura's disease. Prominent lymphoid aggregates infiltrating muscle. H&E x 100

Prominent germinal centres varying from a few to numerous were also seen in all cases.

- (iii) *Eosinophilic infiltration* was a striking feature and foci of eosinophilic abscesses were seen in all cases.
- (iv) Other cellular infiltrates included *plasma cells* and *histiocytes* (Fig. 3). Mast cells were not convincingly demonstrable in all cases. No giant cells or granulomata were seen.

2. Vascular proliferation

Vascular proliferation was a constant feature (Fig. 4) and involved both mature and immature vessels. The mature vessels were thin-walled and lined by flattened or plump endothelial cells. In 2 cases, solid masses of uncanalised endothelial cells were seen (Fig. 5). One of these cases showed an occasional vessel lined by atypical endothelial cells (Fig. 6) with eosinophilic cytoplasm and vesicular nuclei resembling histiocytes. In another similar case, areas of angiomatoid appearance were also seen. One of the 6 cases had arteritis as well as neuritis (Fig. 7).

3. Fibrosis

The degree of fibrosis varied from mild to marked. It was more pronounced where lesions were older and lymphoid follicles few.

DISCUSSION

Clinically, the lesions from our cases appeared similar to those reported from elsewhere in Asia. There was a predilection for the head and neck region and the salivary gland was a common site. The lesions can persist for many years and recurrences after surgical excision were common. The only known systemic manifestation was blood eosinophilia.

The histological appearances were fully consistent with what have been described in Kimura's disease. However, unlike the reports from Hongkong and Singapore,^{4,5} we were able to observe all 3 microscopical features. Uncanalised masses of endothelial cells were not reported from other part of Asia but were seen in 2 of our cases. One of these showed angiomatoid proliferation with endothelial hyperplasia and atypia.

We were not able to demonstrate mast cells in our cases. This was a feature noted by Tham *et al.*⁵ in 2 out of 14 cases from Hongkong.

While the above features have not been reported in cases from Asia, they have been seen in cases of angiolymphoid hyperplasia with eosinophilia (AHLE) reported in America and Europe.^{6,7,8} That Kimura's disease and AHLE are different entities have been mooted by many workers. Rosai *et al.*,⁹ after examining cases from Kawada and comparing them with his own in America, concluded that

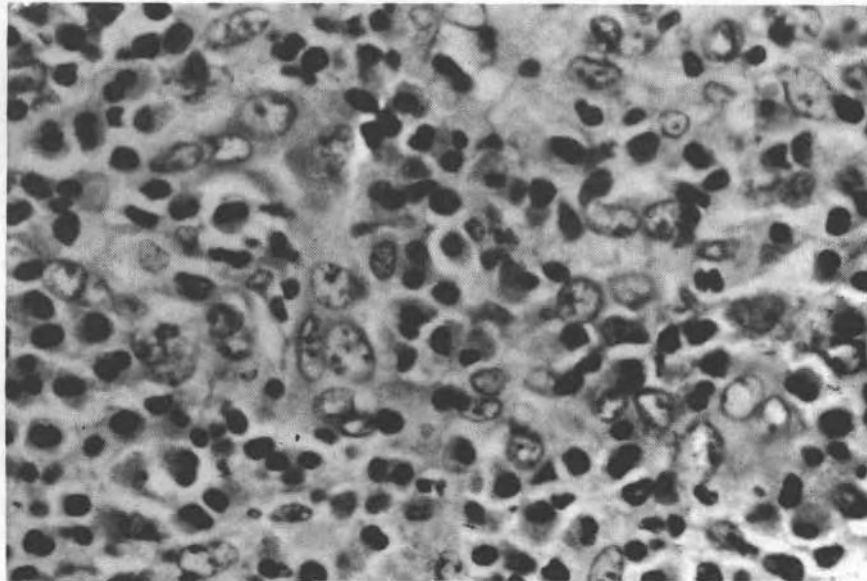


FIG. 3 : Kimura's disease showing an infiltrate of plasma cells, histiocytes and eosinophils. H&E X 550.

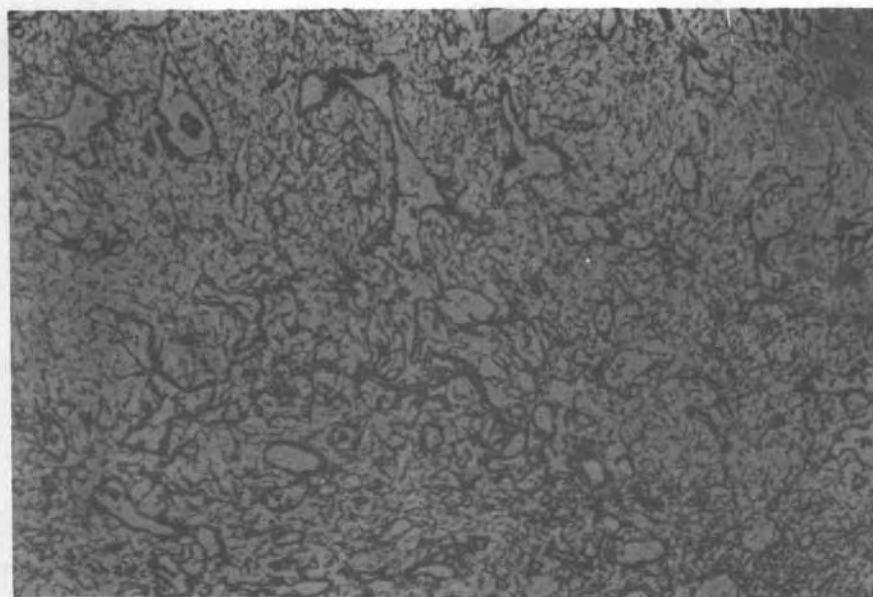


FIG. 4 : Kimura's disease showing marked vascular proliferation. Sweet & Cordon's reticulin X 150.

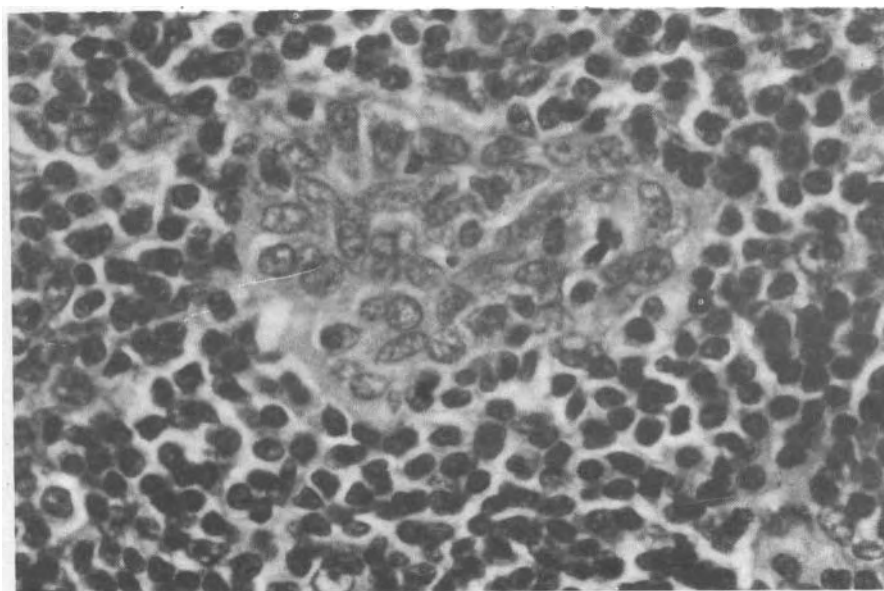


FIG. 5 : A solid mass of endothelial cells indicating an uncanalised vessel in a case of Kimura's disease. H&E X 550.

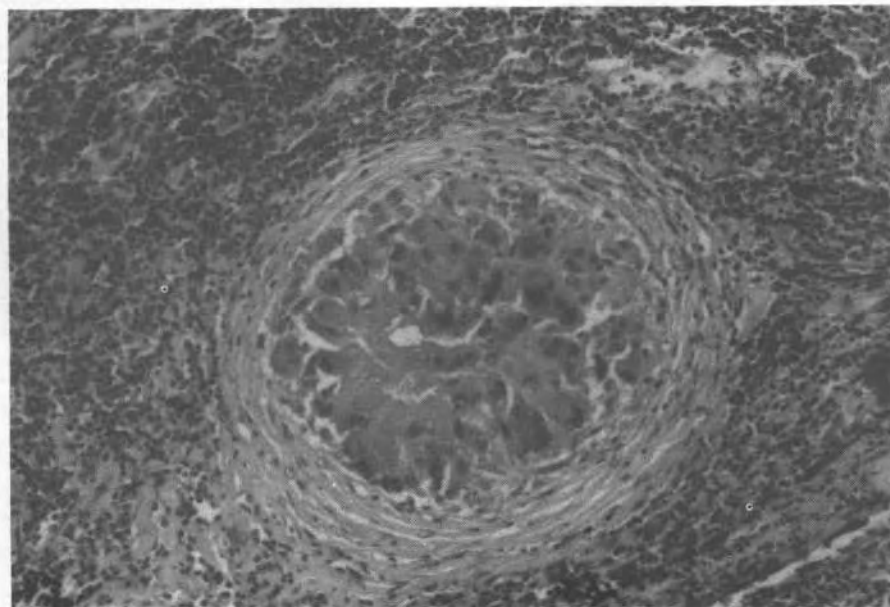


FIG. 6 : A vessel lined by atypical endothelial cells. H&E X 300.

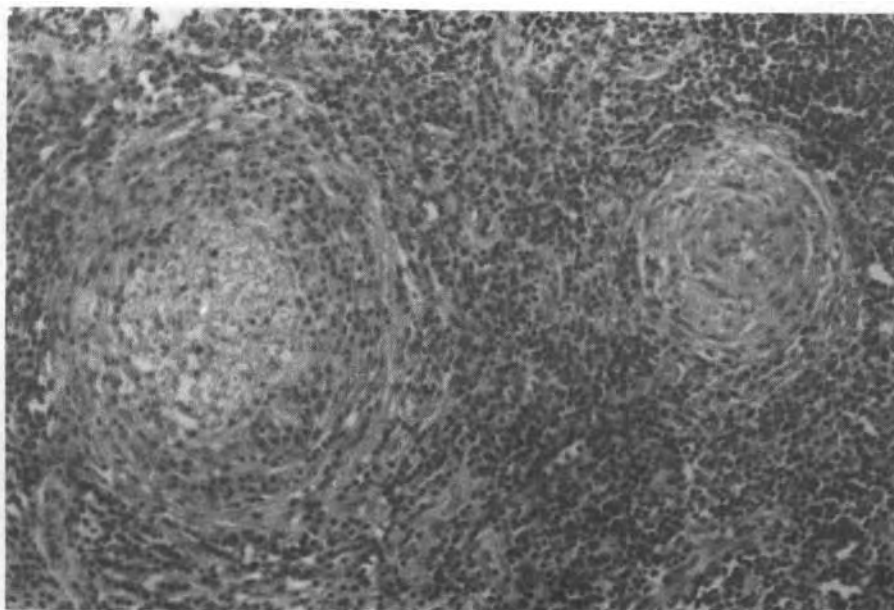


FIG. 7 : Photomicrograph showing arteritis and neuritis in a case of Kimura's disease. H&E X 150.

TABLE 3

A COMPARISON OF THE PATHOLOGICAL FEATURES OF KIMURA'S DISEASE
AND ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA

FEATURES	KIMURA'S	ALHE
Location	Deep subcutaneous, salivary gland.	Dermis or superficial subcutaneous.
Lymphocytic infiltration	Mild to marked.	Marked.
Lymphoid follicles	Always	Sometimes.
Tissue eosinophilia	Always Abscess common	Always.
Mast cells	Not seen.	Common.
Vascular change	Capillary proliferation. Canalised and uncanalised.	Angiomatoid proliferation, canalised and uncanalised, extraordinary endothelial hyperplasia.
Fibrosis	Common	Not common

there were substantial microscopical differences between the two diseases and that they therefore cannot be regarded as the same. Table 3 compares the 2 conditions.

While our cases generally differ clinically and histologically from those from America and Europe, we have observed microscopical appearances reported in AHLE but not in Kimura's disease. Possibly, the age of the lesions plays an important part in determining the variability of vascular changes, with young lesions showing uncanalised endothelial masses, angiomatoid proliferation with endothelial hyperplasia and atypia. This is not unexpected as Asian patients tend to consult doctors late, often with lesions or complaints of considerable duration such as months or years. This is generally not true in America or Europe.

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