

Kimura Disease - A Rare Case Report

Sujata S Giriyan¹, Priya²

¹Professor and Head, Department of Pathology, Karnataka Institute of Medical Sciences, Hubli

² Post Graduate student, Department of Pathology, Karnataka Institute of Medical Sciences, Hubli

Abstract: Kimura disease is a chronic inflammatory disorder usually affecting young adults aged between 20-40years, men are affected more commonly than women^[1]. It involves subcutaneous tissues and lymph nodes predominantly in the head and neck region and is characterized by angiolymphoid proliferation and eosinophilia^[2]. It classically presents as a non-tender subcutaneous swelling in the head and neck region predominantly in preauricular and submandibular area. It may be associated with lymphadenopathy, marked peripheral eosinophilia and elevated Ig E levels^[2]. Histopathology shows hyperplastic lymphoid follicles with prominent germinal centre, hyperplasia of endothelial cells in postcapillary venule, abundant eosinophil infiltration often involving germinal centre, polykaryocytes of Warthin-Finkeldey type and Ig E deposition in germinal centre^[2]. A male patient aged 45years presented with painless swelling in the left inguinal region of 4 months duration. On USG moderate hydrocele was noted on both sides and cellulitic changes in the left inguinal lymph nodes. Multiple sections studied from the excised lymph node showed dense infiltration by plasma cells and eosinophils in the paracortex and capsule, eosinophilic microabscess in focal areas, plump endothelial cells and many blood vessels. Based on the histopathological findings the diagnosis of Kimura disease was made.

Keywords: Lymphadenopathy, Angiolymphoid proliferation, Eosinophilia

1. Introduction

Kimura Disease/Eosinophilic Lymphogranuloma is a chronic inflammatory disorder. Seen in young adults of 20-40years of age. Male to female ratio is 3:1^[3].

It involves subcutaneous tissue and lymph nodes predominantly in the head and neck region. It is characterized by angiolymphoid hyperplasia and eosinophilia^[2]. The etiology of Kimura disease is still unknown but may be due to impairment or interference with the immune system, atopic reaction to persistent antigenic stimulation by arthropod bites, virus or neoplasm^[3].

2. Case Report

A 45year male patient presented with swelling in the left inguinal region since 4 months. On examination a single swelling was seen in the left inguinal region measuring 9×5cm, soft to firm in consistency, non-tender, mobile. Bilateral scrotal swelling was also noted.

On USG moderate hydrocele was noted on both sides, multiple inguinal lymph nodes noted on both sides showing cellulitic changes. On FNAC scant material was obtained consisting of polymorphic population of lymphocytes.

Histopathology of excised left inguinal lymph node showed follicles of varying sizes with prominent germinal centre[Fig 2], paracortical region and capsule showed dense infiltration by eosinophils and plasma cells[Fig 1]. Eosinophilic microabscesses seen in focal areas, folliculolysis by eosinophils was also seen[Fig 3]. Many blood vessels with plump endothelial cells were seen[Fig 4].

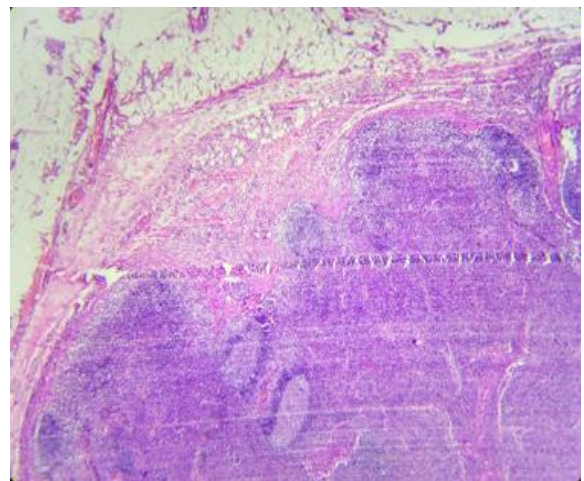


Figure 1: Eosinophil and plasma cell infiltration in the lymph node capsule, follicles with prominent germinal centre. H and E stain(10x objective)

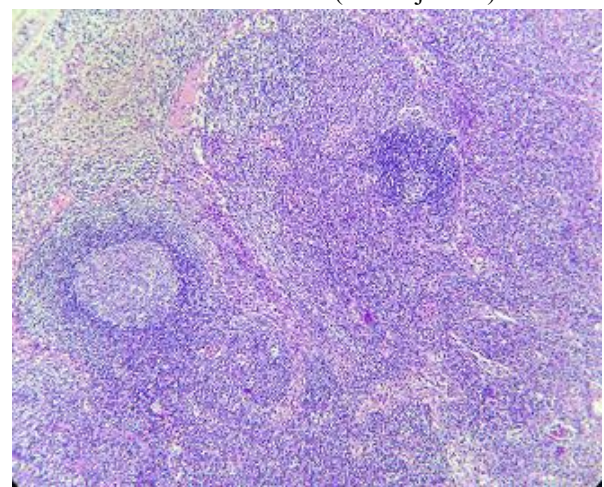


Figure 2: Lymphoid follicles with prominent germinal centre, eosinophil and plasma cells infiltration seen between the follicles. H and E stain (10x objective)

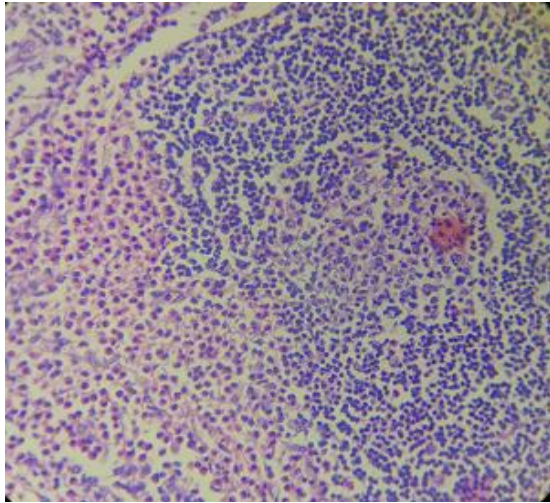


Figure 3: Eosinophil and plasma cell infiltrating the lymphoid follicle showing folliculolysis. H and E stain(40x objective)

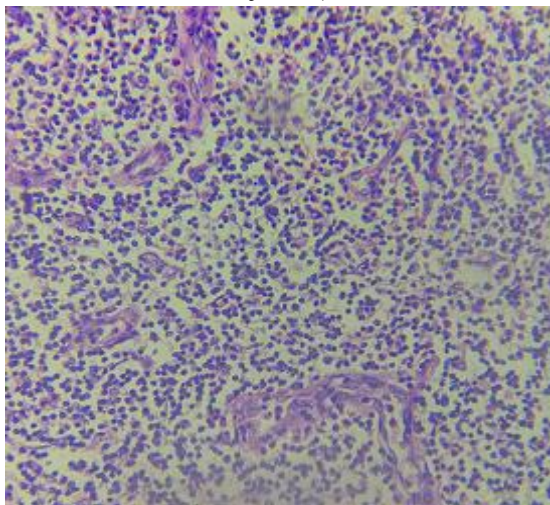


Figure 4: Vascular proliferation and lymphocytic infiltration. H and E stain(40x objective)

3. Discussion

Kimura disease is a chronic inflammatory disorder involving subcutaneous tissue and lymph nodes predominantly in the head and neck region frequently involving periauricular and submandibular region^[1]. More common among Asians, predominant in young men aged between 20-40years^[3].

The onset is insidious, manifests as a painless enlarging nodular mass located deep in the subcutaneous tissues and in almost all cases it involves regional lymphnodes. The lesions are single in 60% of cases, occasionally an isolated enlarged lymphnode may be the only presentation. Some cases can present with generalised lymphadenopathy. Peripheral blood eosinophilia and elevated serum Ig E levels are the constant features^[2].

Eosinophil infiltration is also seen in skeletal muscle, prostate, kidney in some cases. Coexisting renal disease is common, with an incidence ranging from 10% to 60% while 10% to 12% of patients may suffer from nephrotic syndrome characterized by clinically relevant proteinuria in 12% to 16% of cases^[4]. Renal impairment is probably due to

immune-complex mediated damage or due to T-helper 2 dominant immune response disorders^[4]. Kimura disease is often associated with autoimmune diseases such as ulcerative colitis and more frequently bronchial asthma^[4].

Histopathology shows markedly hyperplastic follicles with reactive germinal centre. Diffuse eosinophilia, eosinophilic microabscess and infiltration of germinal centre sometimes resulting in folliculolysis. Vascular hyperplasia mostly of postcapillary venules is seen in the mantle zone of germinal centre^[2].

Immunohistochemistry shows deposits of Ig E in the germinal centre^[5], vascular endothelial cells stain strongly with factor VIII and UEA-1 (ulex europaeus agglutinin)^[6].

Although there is no specific diagnostic feature of Kimura disease, fine-needle aspiration cytology is helpful in some cases, and definitive diagnosis can be obtained by histological examination of the excised lesion^[3]. Ultrasound, CT and Magnetic Resonance Imaging (MRI) might be diagnostic and can help staging the extent and progression of the disease as well as the lymph node involvement.

Treatment with radiation therapy shows best results, steroid therapy and excision are associated with high rate of recurrence^[5].

Differential diagnosis includes mainly angiolymphoid hyperplasia with eosinophilia(ALHE). ALHE is more common in Caucasians, females, presents with superficial skin lesions, lymphadenopathy is not seen^[5,6,7,8,9]. Others are Castleman disease, drug reactions, parasitic infestations, dermatopathic lymphadenopathy.

4. Conclusion

Kimura Disease is diagnosed based on histopathological findings of the excised lesion. Angiolymphoid proliferation and eosinophilia are characteristic findings. Above case was diagnosed as Kimura Disease on histopathology which showed the characteristic features.

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