

CASE REPORT**Kimura's disease in gall bladder and mesenteric lymph nodes : An Unusual Presentation**Garima Verma¹, Shaitan Singh Rathore², Vijay Verma³, S.R. Negi⁴, Amit Kumar Sharma⁵**Abstract**

subcutaneous tissues, the major salivary glands and lymph nodes primarily in the head and neck area. Kimura's disease was first reported in China in 1937. The cause of Kimura's disease is unknown and many theories have been proposed. We report a case of Kimura's disease in gall bladder and mesenteric lymph nodes in an Indian female presented with cholelithiasis and ascites. Histopathological examination allowed us to make a definitive diagnosis. The treatment of choice was surgical removal.

Keywords: Kimura's disease, Eosinophilia, Cholelithiasis.

INTRODUCTION

Kimura's disease is a chronic eosinophilic inflammatory disorder which presents with a slowly enlarging soft tissue mass and regional lymphadenopathy most commonly seen in the head and neck region. Salivary gland involvement has also been reported.¹ Although Kimura's disease mainly affects the head and neck, involvement of the extremities and inguinal lymph nodes has been reported.^{2,3} Peripheral eosinophilia is common and serum IgE levels are often elevated.⁴⁻⁷ Histologically, it is manifested by an abnormal proliferation of lymphoid follicles and vascular endothelium, and eosinophilic infiltrates involving the interfollicular areas and sinusoidal areas.³ Here we report a case of kimura disease in 40 year old Indian female presenting with ascites and cholelithiasis.

¹Assistant Professor, ⁴Professor, Department of Pathology. ²Professor, ³Assistant Professor, ⁵PG Student, Department of Surgery, Dr. S. N. Medical College, Jodhpur (Rajasthan).
Corresponding address: Dr. Vijay Verma
Department of Surgery, Dr. S.N. Medical College, Jodhpur. e-mail: vermavijay71@yahoo.com

CASE REPORT

A 40 year female presented to us with 6 months history of pain right hypochondrium and distension abdomen. Clinical examination findings revealed shifting dullness suggestive of ascites only. USG showed cholelithiasis with ascites. CECT was planned for further workup. CT findings were cholelithiasis with moderate free fluid and mesenteric lymphadenopathy. So patient was planned for open cholecystectomy and mesenteric lymph node biopsy. Laboratory investigations comprised high TLC (13.53*10) along with marked eosinophilia (48%). Mesenteric lymphadenopathy with oedematous bowel loops was seen along with contracted gall bladder intraoperatively. About 500 cc ascitic fluid was drained. The gall bladder and mesenteric lymph node were sent for histopathological examination. Ascitic fluid was also sent for biochemical examination and cytology. HPE of the excised swelling showed lymph node architecture with marked hyperplasia's of germinal centers. Extensive infiltration by mature eosinophils was present with occasional areas of eosinophilic abscess, sinusal and paracortical sclerosis was present. Plenty of plasma cells and mast cells were also seen.

After surgery patient was advised to come for regular follow up. She has not so far developed any new lesions or complications.

DISCUSSION

Kimura's disease was first described in China in 1937, but it was not referred to as Kimura's disease until its description in the Japanese language literature in 1948.^{8,9} The cause of this rare disease remains unknown, but it is thought to be an immune-mediated disorder and not neoplastic. Kimura's disease is endemic to Asians, has a male predominance, and presents with painless soft tissue nodules and lymphadenopathy in the head and neck region. Common sites of involvement are the parotid glands and the epitrochlear, axillary, and inguinal nodes. Although the masses enlarge slowly, patients remain otherwise asymptomatic. Pruritus and dermatitis may occur, and skin lesions can present as reddish brown papules or as subcutaneous nodules. Rare sites of involvement include the kidneys, orbits, ears, spermatic cord, and nerves.¹⁰ Etiology is thought to be an abnormal immune response to an unknown antigenic stimulus.

The constant histologic features which are seen in this disease are preserved lymph node architecture, florid germinal centers, eosinophilic infiltration, and an increased amount of postcapillary venules. The frequent features include sclerosis, karyocytosis in both the germinal centers and the paracortex, vascularization of the germinal centers, proteinaceous deposits in germinal centers, necrosis of germinal centers, eosinophilic abscesses, and atrophic venules in sclerotic areas.^{11,12}

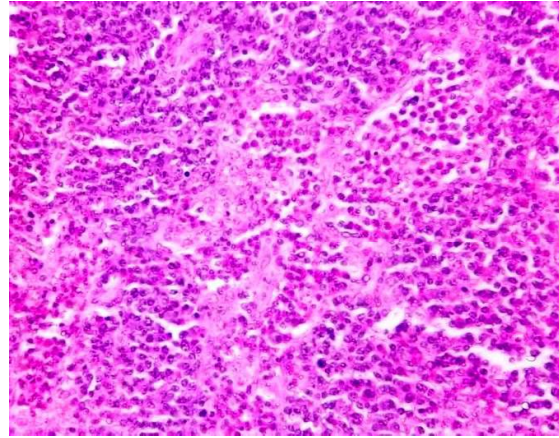


Fig. 1 Kimura's disease in mesenteric lymph node showing numerous eosinophils (10x)

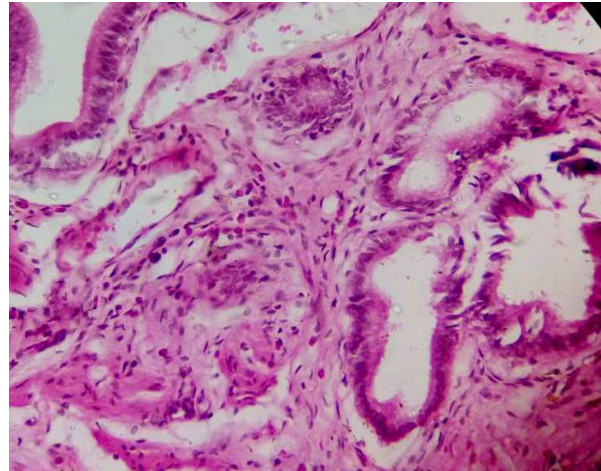


Fig. 2 Kimura's disease in gall bladder(10x)

Treatment of Kimura's disease is not specified. Surgical excision of lesion(s) is the first line therapy even though relapses are frequent. Systemic corticotherapy with prednisone is indicated for relapses and for renal involvement with good efficacy, but with a risk of relapse when the treatment is withdrawn.¹³ Radiation treatment is usually used for the local control of lesions not responsive to steroids, and total doses of 20-30 Gy have proved effective. Irradiation should be considered not only in patients resistant to steroids but also in young patients in whom the long term side effects of steroids may be more

deleterious than a limited course of irradiation which may prevent relapse.¹⁴

CONCLUSION

This case report signifies the need for increased awareness about the unusual presentation of Kimura's disease. Optimal treatment protocol and long term prognosis of patients with Kimura's disease are not established due to lack of long term follow up data in the literature and rarity of these lesions. There is no evidence that Kimura's disease of gall bladder and mesenteric lymph nodes carries an exceptionally poor outcome. Also there is no potential that the lesions will turn malignant.

DECLARATIONS

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