Original Research Paper

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Pathology

KIMURAS DISEASE PRESENTING AS INTESTINAL OBSTRUCTION– A RARE PRESENTATION.

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ABSTRACT Kimuras disease is a chronic inflammatory condition of uncertain etiology. Kimuras disease primarily involve Head and Neck region presenting as deep subcutaneous masses in the preauricular region ,forehead or scalp. Other areas that can be involved are lymph nodes, salivary glands . The disease is endemic in Asians but occurs sporadically in other racial groups and shows a distinct male predeliction . However we report a 30 year old female presenting with intestinal obstruction diagnosed with Kimuras disease.

KEYWORDS :

INTRODUCTION

Kimuras disease is a chronic inflammatory condition of uncertain etiology.First described by Kimm and Setzo in 1937 as eosinophilic hyperplastic granuloma(1) it gained prominence as kimuras disease following a report by kimura and co-workers in 1948 which elaborated on an unusual granulation combined with hyperplastic changes in lymphoid tissue(2).Kimuras disease primarily involve Head and Neck region presenting as deep subcutaneous masses in the preauricular region ,forehead or scalp(3)Other areas that can be involved are lymph nodes, salivary glands . The disease is endemic in Asians but occurs sporadically in other racial groups and shows a distinct male predeliction (3)

CASE REPORT

30 years old female presented with clinical symptoms of PAIN WHOLE ABDOMEN,Colicky in nature ,off and on for a duration of 6 days with 2 -3 episodes of vomiting. There was no history of fever,night sweats , weight loss , maleana, haematemesis, jaundice or anorexia.ON examination there was mild distension of abdomen with mild diffuse tenderness all over the abdomen.Laboratory investigations were normal with RAISED esr 30 mmh.Surgical Excision was done and ileocolic intussusceptions found on exploration was removed and specimen sent to our Laboratory for histopathologic examination . Specimen was fixed in 10 % buffered formalin for 24 hours . specimen measures 12 cm in length. Intussusceptum was found to be a solid white mass measuring 7 cm ×5cm completely occupying the lumen of intusscepien. Grossing was done .Sections from intusscepien as well as intussussceptum were taken , processed and stained by Haematoxylin and Eosin.Microscopic examination of section showed intestinal mucosa with wall showing presence of lymphoid follicles along with vascular proliferation with flat endothelial lining and variable eosinophilic infilterate forming eosinophilic abcesses at places.The diagnosis of Kimuras disease was based on histopathologic findings in conjuction with peripheral eosinophilia and elevated serum immunoglobulin E level.

DISCUSSION

Heut et al in 1989 classified the histological features of kimuras disease as constant , frequent and rare. The constant feature include preserved nodal artitecture, florid germinal centre hyperplasia, eosinophilic infiltration and postcapillary venule proliferation.This can give rise to differential diagnosis of Hodgkins lymphoma, angioimmunoblastic T cell lymphoma, Langerhans cell histiocytosis and parasitic lymphadenitis. However the lesion that bears closest resemblance to kimuras disease ,to the extent that two were once considered to be the same entity, is angiolymphoid hyperplasia with eosinophilia. Ever since ALHE was described in 1969, the misapprehension that ALHE and Kimuras were identical or at least two ends of the same disease spectrum has been perpetuated by the similarities between the two conditions namely a predilection for the head and neck region ,clinical presentation as a subcutaneous mass tendency to recur despite treatment and the presence of lymphoid infilterate with eosinophils and vascular proliferation. A path breaking study by Rosai et al (1979) (4) eventually clarified this misconception and both were established as two distinct entities. Currently kimuras disease is believed to be a chronic allergic inflammatory process of unknown origin whereas ALHE is considered to be benign vascular proliferative disorder(5). Kimuras is predominantly seen in young asian males manifesting as single or multiple asymptomatic large masses in the subcutaneous tissue or salivary glands. Regional lymphadenopathy ,peripheral blood eosinophilia and elevated serum IgE levels are commonly seen and the lesion are of shorter duration(6) whereas ALHE principally affects women in 3rd to 4th decade and has no racial predilection(7). It typically presents as multiple erythematous to brown dermal papules or nodules accompanied by tenderness and pruritis. Regional lymphadenopathy ,peripheral blood eosinophilia and elevated serum IgE levels are rare and the lesion are of longer duration. The tendency towards renal involvement and nephritic syndrome is also restricted to patients of kimuras disease and is not seen in ALHE(8).

Microscopically lesion exhibit characterstic immense eosinophilic infilterates and aggregation of eosinophils to form eosinophilic microabcessess ,lymphoid granulomas with vascular proliferation with variable degree of fibrosis(12).. Vessels are thin walled with flat endothelial lining without classic epithelioid or histiocytic cells of ALHE. Whereas in ALHE lymphoid infilteration is mild to moderate and the presence of lymphoid follicles is inconsistent with variable degree of eosinophilic infilterate and eosinophilic abcessess are usually absent. Vessels in ALHE show florid proliferation with characterstic endothelial lining of plump low cuboidal,epithelioid or histiocytoid cells having abundant acidophilic cytoplasm with vacoulization vesicular nuclei with a cobblestone like arrangement . Fibrosis is not seen(10).

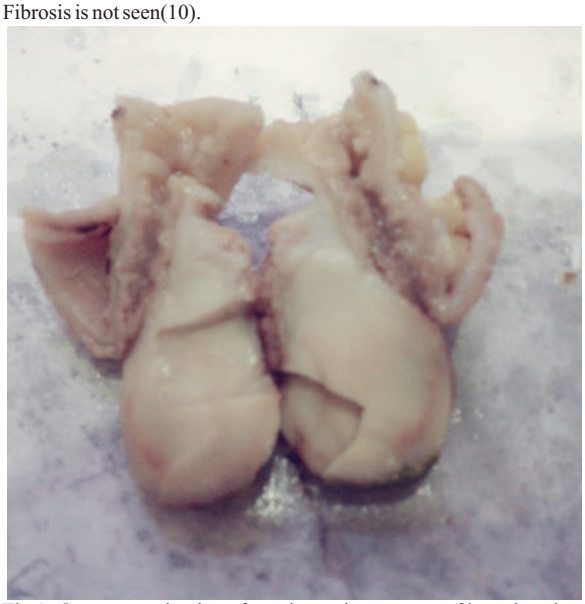


Fig 1- Gross examination of specimen shows a part of intestine along with the lesion presenting as intussusceptum.Lesion was pear shaped whistish brown in colour , solid to firm , well circumscribed measuring 8cm ×5cm.

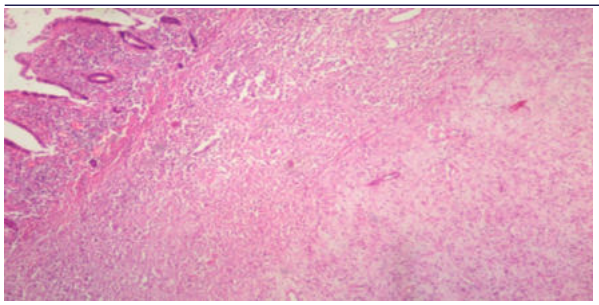
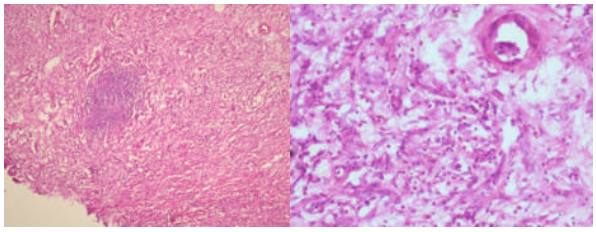


Fig 1 – shows intestinal mucosa along with angiolymphoid hyperplasia in wall. Haematoxylin and eosin 20x



CONCLUSION

The present case report highlights the need for increased awareness by all clinicians about the unusual presentation of Kimuras Disease.

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