Case Report

Indian Journal of Trauma and Emergency Pediatrics
Volume 8 Number 1, January - April 2016
https://dx.doi.org/10.21088/ijtep.2348.9987.8116.6

Kimura's Disease in A Bangladeshi Boy: A Very Rare Cause of Lymphadenopathy

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Abstract

Kimura's Disease is a rare chronic inflammatory disorder that characteristically manifests as enlargement of head and neck lymph nodes and salivary glands accompanied by eosinophilia and elevated serum IgE. It is a benign condition with unknown etiology and said to be predominantly seen in males of Asian descent. We are presenting here a clinically and histologically typical case of this disease in a 12 years old Bangladeshi boy. The diagnosis of Kimura's Disease can be very difficult and misleading; it is important not to ignore histopathological characteristics.

Keywords: Lympadenopathy; Eosinophilia; Histopathology.

Introduction

Kimura's disease (KD) is a rare benign chronic inflammatory condition presenting as multiple painless subcutaneous nodules localized mostly in the region of head and neck with coexisting lymphadenopathy and peripheral eosinophilia [1] .This quite rare condition is found almost exclusively in Asian individual in their 2nd to 4th decade of life mostly in males [2]. The first known report of Kimura's disease was from China in 1937, when Kimm & Szeto identified seven cases of the condition [3]. It first received its name in 1984 when Kimura and others noted a change in the surrounding blood vessels and referred to it as "unusual granulation" combined with hyperplasic changes in the lymphoid tissue [4]. The etiology of KD remains unknown, although an allergic reaction, trauma and an autoimmune process have all been implicated as the possible cause. The disease is manifested by an abnormal proliferation of lymphoid follicles and vascular endothelium [5].

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Case Report

Imran Hussain, a 12 years old male child, fully immunized as per EPI schedule, presented to us with swelling in the right side of the neck of 6 months duration.

Fig. 1: Patient with neck swelling

He had no history of fever, cough, fatigue, loss of weight, contact with tuberculous patient, bleeding manifestation, discharging sinus or bone and joint pain. On examination, the boy was afebrile with normal vital signs and growth, no pallor or clubbing.

In the right cervical region, posteriorly, there were three mobile, firm, non-tender lymph nodes of sizes 4.5x3, 4x2.5 & 3x2 cms with normal skin over them with no discharging sinus. Other groups of lymph nodes were not enlarged. No other abnormality including hepatosplenomegaly was noted.

The blood count showed eosinophilia (20%); peripheral smear was within normal limit except for eosinophilia. Serum IgE was very high (2000 iu/ml). Blood culture, urine and stool analysis did not show any abnormality. Skin tuberculin test was negative; same with gastric aspirate for AFB. Chest radiology and skeletal surveys were normal. Histopathological examination of excised swelling showed section of lymph node with follicular hyperplasia, paracortical expansion with prominent high endothelial venules. There was extensive infiltration of eosinophils with focal eosinophilic micro abscess. Hyalinized vessel wall, interstitial fibrosis and deposition of proteinaceous material were also present. The expanded paracortex contained plasma cells, small lymphocytes and occasional giant cell. No evidence of malignancy was seen. All these features were consistent of Kimura's disease.

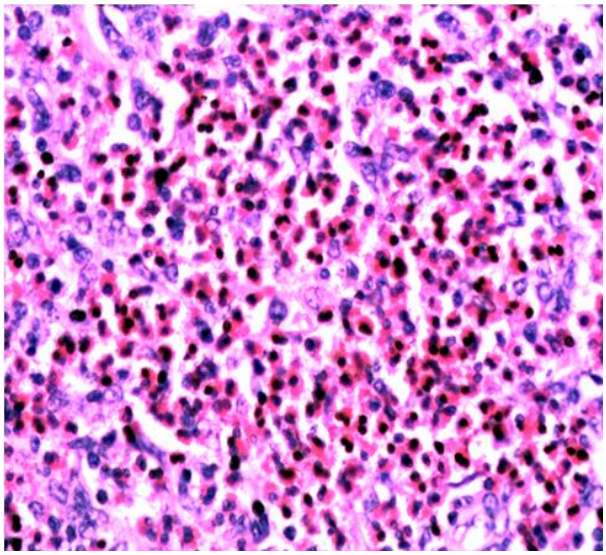

Fig. 2: Histo-pathological examination of biopsy specimen

Oral prednisolon along with cetrizine were given to the patient. The lesion began to shrink gradually within 7 days of steroid therapy followed by improvement of general well being. The patient was observed for next two weeks with continuation of same treatment and ultimately discharged 3 weeks after admission when the lesion almost disappeared with normalization of peripheral eosinophilia and immunoglobulin level. The patient was advised for follow up after one month.Follw-up examination of the patient was done about one and half month later which revealed complete disappearance of the lesion; he was advised to come for regular follow-up.

Discussion

Patients with Kimura's disease (KD) present in majority cases with nontender subcutaneous nodules and masses in the head and neck region [6]. Typical areas for the nodules are preauricular, submandibular and popliteal regions as well as oral cavity, larynx and parotid glands [7]. In addition, a presentation of Kimura's disease as a pulmonary hilar mass has recently been described [8]. Histologically, Kimura's disease was initially thought to be related to angiolymphoid hyperplasia with eonsinophilia (ALHE), however, clinically and radiologically these two entities are clearly different. Differential diagnoses of KD on imaging are that of lymph node enlargement and include: benign reactive nodes/ infectious mononucleosis/drug reactions, nodal metastasis, lymphoma/ leukemia, tuberculous adenitis, parasitic infections, etc [9]. Nearly all patients with KD demonstrate peripheral eonsinophilia and elevated levels of 1gE. In one study, the number of eosinophils was closely correlated to the sizes of the neck masses [10]. Ultrasonography is a useful modality for assessment of the neck and to aid in biopsy. The appearance of KD on other imaging modalities, including CT scan and MRI is variable and is thought to be due, at least in part, to the variable degrees of vascular proliferation and fibrosis within individual lesion. The other imaging findings also include multiple ill-defined, enhancing lesions [6]. Excisional biopsy is the gold standard to diagnose KD and to exclude malignancy.

The typical histopathologic finding is characterized by proliferation of lymphoid follicles, cellular infiltrates mainly of eosinophilic which may sometimes form eosinophilic abscess and sometimes with plasma cells, lymphocytes and mast cells. Vascular proliferation along with fibrosis is also noted [11]. Kimura's disease can lead to disfigurement secondary to growth of untreated lesions, particularly given the prediction for the head and neck. Additionally, recurrence after treatment is well described [12]. There is no consensus on the management aspect in KD so far. Observation is acceptable if the lesions are neither symptomatic nor disfiguring. However, primary prophylactic surgery is performed as a therapeutic and / or diagnostic procedure [13]. Treatment options include conservative management, steroid & immunosuppressants, surgical excision &

radiotherapy. Intralesional or oral steroids can shrink the nodules but seldom results in cure. Immunosuppressant (eg, cyclosporine) has been reported to induce remission, but the recurrence of the lesion has been observed once this therapy is stopped [14,15]. Cetrizine (an antihistamine) is an effective agent in treating its symptoms (pruritis) [6]. Radiotherapy has occasionally been used to treat recurrent or persistent KD lesions [17]. Conservative surgical excision has been considered the treatment of choice for KD; however recurrence after surgery is frequently observed [18]. To date, no malignant transformation of KD has been described in the literature. If not treated, KD can lead to disfigurement.

Conclusion

Kimura's disease is a chronic benign inflammatory condition endemic to Asians with lesions frequently persisting or recurring despite treatment. If not properly diagnosed, the cervical lymphadenopathy in KD may be initially mistaken for a malignancy. However, due to a well-obtained clinical history and histopathological awareness a proper diagnosis has been established in this case.

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STATEMENT ABOUT OWNERSHIP AND OTHER PARTICULARS
"Indian Journal of Trauma and Emergency Pediatrics" (See Rule 8)

1. Place of Publication	:	Delhi
2. Periodicity of Publication	:	Quarterly
3. Printer's Name	:	**Asharfi Lal**
Nationality	:	Indian
Address	:	3/258-259, Trilok Puri, Delhi-91
4. Publisher's Name	:	**Asharfi Lal**
Nationality	:	Indian
Address	:	3/258-259, Trilok Puri, Delhi-91
5. Editor's Name	:	**Asharfi Lal** (Editor-in-Chief)
Nationality	:	Indian
Address	:	3/258-259, Trilok Puri, Delhi-91
6. Name & Address of Individuals who own the newspaper and particulars of shareholders holding more than one per cent of the total capital	:	**Asharfi Lal** 3/258-259, Trilok Puri, Delhi-91

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