Pediatr Pol 2019; 94 (1): 54–57
DOI: https://doi.org/10.5114/polp.2019.83745
Submitted: 12.11.2018; Accepted: 12.01.2019; Published: 27.02.2019

PEDIATRIA Polska
Polish Journal of PAEDIATRICS

CASE REPORT

An unusual cause of head and neck lymphadenopathy in a paediatric patient: a case report

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ABSTRACT

Neck node enlargement is a common clinical presentation in the paediatric age group. Here we present a rare cause of neck node enlargement in a paediatric patient, called Kimura's disease (KD). KD is a rare chronic inflammatory disease with angiolymphatic proliferation. The aetiology of KD is still unknown in medical literature. A painless unilateral soft tissue mass in the head and neck region is the commonest clinical presentation. This disease should be suspected when there are painless neck nodes, hypereosinophilia, and hyper IgE, particularly in male Asian patients. The diagnosis of KD is usually confirmed by histopathological examination. There are three main treatment options in KD: surgical excision, corticosteroids, and radiotherapy. Clinicians need to be aware of KD and help to limit the number of laboratory tests and reduce the possibility of malignant lesions.

KEY WORDS:

Kimura's disease, paediatric patient, neck node enlargement, angiolymphoid hyperplasia.

INTRODUCTION

Kimura's disease (KD) is a rare clinical entity of unknown aetiology, which is occasionally seen in Asian populations. It is a benign chronic inflammatory lesion in which the patient presents with painless lymphadenopathy or subcutaneous mass in the head and neck region. The first clinical case on KD was reported by Kimm and Szeto from China in 1937 where they described seven cases of such conditions and termed them eosinophilic hyperplastic lymphogranuloma [1]. The exact prevalence of KD is not well known in medical literature. Although it is a rare disorder, few cases are reported in East and Southeast Asia, and it is extremely rare in Europe. It is often seen in young adults with a median age of 28 to 32 years, and the male-to-female ratio ranges from 3.5 : 1 to 9 : 1 [2]. There are few cases reported in India, but there are very few clinical reports from the eastern part of the country for paediatric patients. Here, we present a case of KD in 12-year-old boy presenting with post aural lymphadenopathy.

CASE REPORT

A 12-year-old boy presented with swelling behind the left ear for the preceding six months. He had no history of ear discharge. There was no associated neck swelling or any oedema in the body or swelling around the eye. On examination, there was a swelling of 4 cm × 5 cm seen behind the left ear (Fig. 1). It was firm in consistency,

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non-tender, and with no local rise in temperature. The otoscopic examination revealed a normal external auditory canal and normal ear drum. There were no other significant findings in local or systemic examination and no hepatosplenomegaly. He was afebrile without any pallor, cyanosis, or clubbing. His blood report showed mild leucocytosis with 12% eosinophils and raised absolute eosinophil count (1.25 G/l). The erythrocyte sedimentation rate was 30 mm in the first hour. Biochemical reports like blood glucose, serum urea, creatinine, bilirubin, and liver function tests were within normal limits. Serum IgE level was also elevated at 8.57×10^5 IU/l (normal range $< 11.1 \times 10^3$ IU/l). Chest X-ray was normal and fine-needle aspiration cytology (FNAC) from post-aural mass revealed a polymorphous population of lymphoid cells with the presence of mature lymphocytes, centroblasts, centrocytes, plasma cells, and immunoblasts. There were numerous eosinophils along with reactive lymphoid cells and few histiocytes with large multinucleated giant cells. Incisional biopsy was done, which showed a lesion with lymphoid aggregates with infiltration of eosinophils. There was high vascularity at inter-follicular areas with perivascular infiltrates rich in eosinophils and lymphocytes (Fig. 2). There was mild acanthosis seen in overlying dermis and fibrosis in subcutaneous tissue. A diagnosis of KD was done on the basis of histopathology. He underwent complete surgical excision. The mass was sent for histopathological examination, which revealed lymphoid aggregates with dense eosinophilic infiltrates. The inter-follicular area showed increased vascularity and perivascular infiltrates with eosinophils and lymphocytes. There was a predominant deposition of IgE within the germinal centre. The patient was started with an oral steroid, i.e. prednisolone 1 mg/kg once daily for two weeks followed by a tapered dose for six weeks. Based on the histopathological picture, peripheral hypereosinophilia, raised serum IgE level, and negative serological testing for parasitic infestations, a clinical diagnosis of KD was made. The follow up at one year did not show any evidence of recurrence.

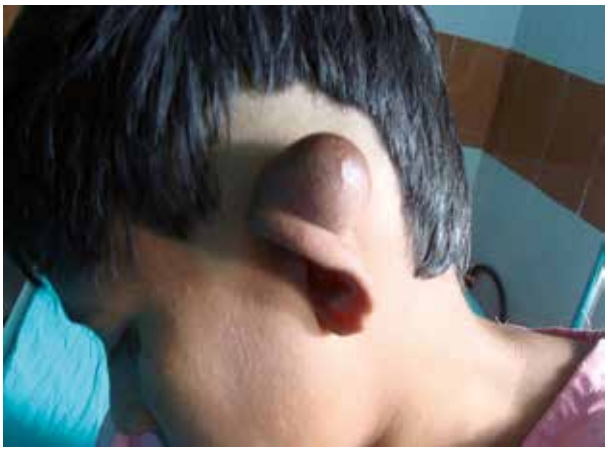

FIGURE 1. Clinical photograph of the patient showing a mass behind the left pinna

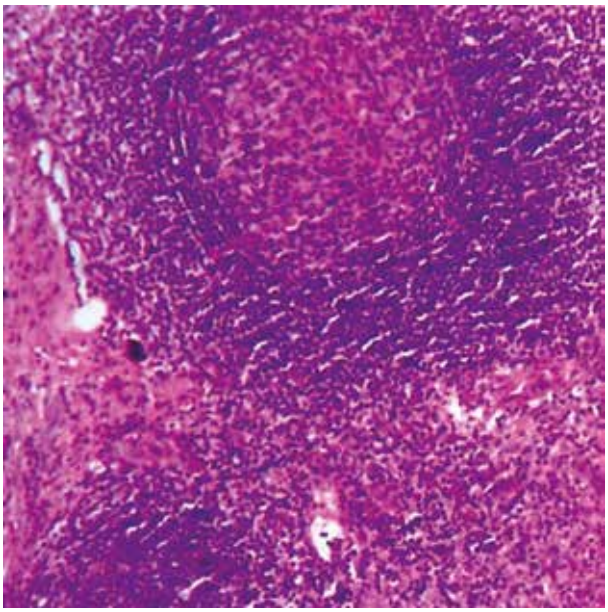

FIGURE 2. Microphotograph showing follicular hyperplasia with infiltration of a large number of eosinophils and lymphocytes (H and E 10×)

DISCUSSION

KD is a chronic, benign, and immunologically mediated disease with inconclusive aetiology. There are several aetiological factors described in the literature like trauma, parasitic infestations, *Candida*, and virus infections [3]. In our case, there was no history of trauma or chronic infections. The neoplastic origin of KD in the head and neck area could not be excluded [4]. The aetiology is thought to be an altered immune response to an unknown antigenic stimulus. It mainly affects males of Asian origin, but sporadic cases have also been reported in non-Asian populations [5]. KD often presents with painless, subcutaneous nodules and often leads to disfigurement in the head and neck region. Common sites of involvement in KD are parotid glands, axillary, epitrochlear, and inguinal nodes [6]. In our case, the mass was present in the post-auricular area, which is a rare location for KD. Most of the cases of KD were reported from China and Japan [5]. It is a male-predominant disease often seen in the second and third decade of life [7]. KD patients often show painless unilateral soft tissue mass in the head and neck region, and salivary glands and neck nodes are frequently affected. It is often associated with increase eosinophil count and serum IgE level [7]. The serum IgE level was raised in our case, which favours diagnosis of KD. Due to increased eosinophil count, this lesion is also called as eosinophilic lymphadenitis [8]. The increased eosinophil count in our case supported the diagnosis of KD. Patients presenting with peripheral eosinophilia may be mistaken for tropical eosinophilia syndrome or high-altitude-associated eosin-

ophilia, so here the patient needed a detailed head and neck examination to rule out KD. Clinically the patient did not show physical deterioration, inflammatory signs, or tumour syndrome. The diagnosis was based on clinical presentation, laboratory reports, and histopathological findings. The laboratory work-up showed increased eosinophils, which often correlate with size of the lesion and raised IgE [9]. Incisional biopsy is often advised to obtain the diagnosis. The histopathological picture comprises preservation of lymph node architecture, eosinophil infiltrates, germinal centre hyperplasia, and proliferation of postcapillary venules. There was high vascularity at interfollicular areas with perivascular infiltrates rich in eosinophils and lymphocytes. There was mild acanthosis seen in the overlying dermis, and fibrosis in subcutaneous tissue. The differential diagnoses of KD are T-cell lymphoma, atypical mycobacterial infection, eosinophilic granuloma, Mikulicz's disease, acute nonlymphocytic leukaemia, Hodgkin's disease, Langerhans histiocytosis, florid follicular hyperplasia, Castleman's disease, parasitic infections, angio-immunoblastic lymphadenopathy, and angiolymphoid hyperplasia with eosinophilia (ALHE) [10].

There is no definite standard protocol for the treatment of KD. The treatment options are surgical excision, steroid therapy, and radiotherapy. Surgery is the mainstay of treatment, where it is effective for the eradication of the mass if the whole mass is completely excised. Systemic and intralesional steroids help to reduce the size of the lesions in KD. Sometimes the mass tends to recur if steroids are stopped or discontinued [5]. There is no documentation in the medical literature regarding malignant transformation. The main concern after treatment is recurrence, which occurs in 17% to 44% of cases [11]. It has been reported that peripheral eosinophilia at levels more than 50%, serum IgE levels greater than 10,000 IU/ml, and multifocal lesions outside the salivary glands are important prognostic factors for the recurrence of KD [12]. If it is not symptomatic or disfiguring, only observation can be done. Oral corticosteroids are often used for the treatment. Sometimes intralesional corticosteroids like triamcinolone may be useful to control the localised lesion. Corticosteroids suppress the inflammation by reducing migration of the polymorphonuclear leukocytes and reversing the permeability of the capillary. This disease often recurs after cessation of the treatment. Cyclosporine, trans-retinoic acid with prednisolone, oral pentoxifylline, intravenous immunoglobulin, and photodynamic therapy sometimes induce remission. Radiotherapy is occasionally used to control persistent or recurrent disease. Conservative excision of the mass is considered as the treatment of choice for KD although there is a chance of recurrence [13]. There is a chance of recurrence also after surgical excision of the lesion although it is benign. There is a risk of recurrence in 25% of cases of KD, particularly between one to three years after treatment [14]. Corticosteroids or revision surgery can be done in the case of recurrence. Corticosteroids can be given when a patient has renal complications [9]. Radiotherapy of 20 to 30 Gy can be suggested when there is absence of local response to corticosteroids or in the case of adverse effects of corticosteroid therapy [9]. Other treatments like intralesional corticosteroid injection, cyclosporine, or photodynamic therapy are sometimes suggested. Oral pentoxifylline is prescribed in the dose of 400 mg twice daily for two months to resolve the lesions, without any side effects. It has effects on cytokines like IL-2, IL-6, IL-8, and tumour necrosis factor. The efficacy of pentoxifylline is based on aberrant chemokine levels [15]. Because KD is particularly rare in western countries and clinicians are unfamiliar with pathognomonic findings of this disorder, sometimes unnecessary diagnostic tests and different investigations are performed [16].

CONCLUSIONS

KD is a rare clinical entity in paediatric patients. It is a chronic inflammatory disease without any exact aetiology. Clinician should bear KD in mind when assessing neck node enlargement in children, particularly in Asian countries, and should diagnose it clinically on the basis of painless head and neck swelling along with marked eosinophilia and raised serum IgE. The prognosis of this lesion is usually good, and patients often need a long-term follow-up to rule out recurrence.

DISCLOSURE

The authors declare no conflict of interest.

REFERENCES

1. Rao K, Kumar S. Kimura's disease – A rare cause of head and neck swelling. Int J Otolaryngol Head Neck Surg 2014; 3: 200-204.
2. Sun QF, Xu DZ, Pan SH, et al. Kimura disease: review of the literature. Intern Med J 2008; 38: 668-672.
3. Meningaud JP, Pitak-Arnnop P, Fouret P, Bertrand JC. Kimura's disease of the parotid region: report of 2 cases and review of the literature. J Oral Maxillofac Surg 2007; 65: 134-140.
4. Pitak-Arnnop P, Bellefqih S, Chaine A, et al. Head and neck lesions of Kimura's disease: exclusion of human herpesvirus-8 and Epstein-Barr virus by in situ hybridisation and polymerase chain reaction. An immunohistochemical study. J Craniomaxillofac Surg 2010; 38: 266-270.
5. Chen H, Thompson LDR, Aguilera NS, et al. Kimura disease: a clinicopathological study of 21 cases. Am J Surg Pathol 2004; 28: 505-513.
6. Saini A, Singh V, Chandra J, Dutta AK. Kimura's disease: An unusual glandular involvement with blood and tissue eosinophilia. Indian J Pediatr 2009; 76: 647-648.
7. Viswanatha B. Kimura's disease in children: A 9 years prospective study. Int J Pediatr Otorhinolaryngol 2007; 71: 1521-1525.
8. Rosai J. Lymph nodes. In: Rosai and Ackerman's surgical pathology, Rosai J. 10th ed. Elsevier, 2011; 1805.
9. Piette EW. Kimura disease. Medscape 2016.

10. Drouillarda M, Stevea M, Ranoarivonya T, Souraud JB. Kimura's disease in a 50-year-old Tunisian man. Eur Ann Otorhinolaryngol Head Neck Dis 2017; 134: 127-129.
11. Iwai H, Nakae K, Ikeda K, et al. Kimura disease: diagnosis and prognostic factors. Otolaryngol Head Neck Surg 2007; 137: 306-311.
12. Lin YY, Jung SM, Ko SF, et al. Kimura's disease: clinical and imaging parameters for the prediction of disease recurrence. Clinical Imaging 2012; 36: 272-278.
13. Piette EW, James JA. Kimura disease clinical presentation. Medscape 2014. http://emedicine.medscape.com/article/1098777clinical#b4 (access: 26 June 2007).
14. Choi HJ. Kimura Disease Presenting as Forehead Soft Mass. J Craniofac Surg 2016; 27: 823-824.
15. Hongcharu W, Baldassano M, Taylor CR. Kimura's disease with oral ulcers: response to pentoxifylline. J Am Acad Dermatol 2000; 43: 905-907.
16. Fouda MA, Gheith O, Refaie A, et al. Kimura disease: a case report and review of the literature with a new management protocol. Int J Nephrol 2010; 2010: 673908.