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A Rare Location of Kimura's Disease in a Greek Male Patient with a Right Thigh's Lump (First Case in Europe): Case Report-Review of Medical Literature

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1. Preamble

Kimura's disease is a benign rare inflammatory disorder of unknown aetiology which might mimic neoplastic's lesion. Epidemiologically this condition, affects young Asian males. Early diagnosis of kimura's disease may spare the patient from unnecessary invasive diagnostic procedure (surgery-biopsy). This nosological condition primarily involves the head and neck region, and is characterized by painless subcutaneous swelling accompanied by triad regional lymphadenopathy, salivary gland involvement as well as high serum Immunoglobulin E (IgE) and eosinophilia level [1-3]. In this article, we are reporting a very rare occurrence of Kimura's disease in a young Greek man, presenting with a right thigh's palpable subcutaneous mass.

2. Introduction

Kimura's disease is a rare chronic inflammatory disorder with angiolymphatic proliferation of unknown aetiology. It was stated that an allergic reaction, trauma and autoimmune process have been implicated as possible causes. According to epidemiological perspective, this condition is very rare, only 200 cases have been reported worldwide [1]. It affects more commonly Asian young people of male gender belonging to a well-defined age-Groupe of 20 and 40 years. Regarding the sex-ratio, males are more affected in the rate of 3 /1 [3].

Despite the disease represents an endemic condition in Asians, it occurs sporadically in other racial groups. Typically, it presents as a subcute painless mass in the head and neck region especially in the periauricular, groin regions as well as in the orbit and eyelids, accompanied by regional lymphadenopathy. It may be associated with eosinophilia and increased IgE levels. The coexisting renal disease is common with an incidence ranging from 10 to 60 %, less than 10 % of patients develop nephrotic syndrome as well as more complicated course [4,5,26]. The diagnosis of Kimura's disease is not easy, it requires medical imaging modalities (CT scan, MRI, Ultrasound) followed by a surgical excision of the involved lump for pathological analysis combined to immunohistochemistry evaluation [21,22]. There are numerous therapeutic modalities, including surgical resection, radiotherapy, immunotherapy and oral corticosteroids, but the best choice is still controversial. However, in one published paper, one case was reported with spontaneous resolution. Generally, the disease prognosis is good [2,5,6]. In this article, we are reporting a rare case of of Kimura's disease in a young Greek patient presenting with a right thigh palpable lump (First case in Greece).

3. Case Report

A 50-year-old Greek man was admitted in our surgical Department presenting with a right thigh painless and palpable lump with irregular borders. His medical antecedents revealed chronic hypertension controlled with a converting enzyme-inibitor (co-renitec), hyperlipidemia internal malleoli's fracture a year prior. His laboratory's findings were INR 0,95, PT10, APTT 30.5. Glu 93, Uria 37, creatinine 1, Na 142, K 3,7, WBC 8,36, Eosinoph 25 %, Lymp 27 PLT 236, Hct 44,2 Hb 14,7. A chest -x ray was normal, an MRI of the concerned thigh revealed a subcutaneous tumour of 2,2 cm diameter, with irregular borders and inducing tentacules and oedema of the underlying muscle. This radiologic description was so suspicious of malignancy that; the patient was shifted to operator's theatre where the radical resection of the tumour was performed. The resected specimen was composed with muscle and adipose tissue , microscopic investigation stated that it was about a mixed inflammatory infiltrate extending into adjacent structures (fat cells,blood vessels,muscle fibers),consisting mainly of numerous eosinophils, and lymphocytes, with scattered lymphoid follicles with germinal centers, which are hyperplastic .The immunophenotype were CD20(+),CD3(+)B,T-lymphocytes,CD68(+) in histiocytes and Ki-67(MIB)(+).This pathologic analysis concluded that it was about a Kimura's disease in its very rare location (Figure 1-abcd).

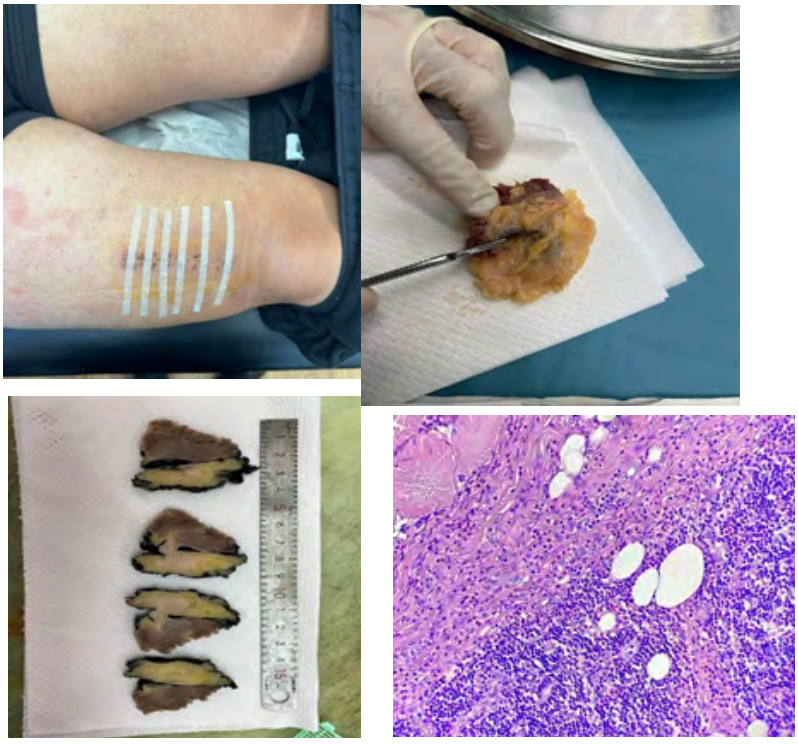

Figure-1(a-b-c-d): Location of the resected lump, resected spacemen, histologic feature.

4. Discussion

Kimura Diseases is a rare chronic inflammatory disorder, which was first described in 1937 by kim and Szeto in Chinese literature as eosinophilic hyperplastic lymphogranuloma, and has been known most often as Kimura's disease since its description in 1948 by Kimura et al in the Japanese literature [2,7]. This disease is endemic in middle-age Asian males and rarely seen sporadically in non-Asian population. However, Chen et al, concluded in their study that though rare, if clinical suspicion arises, KD should be included in any race in the differential diagnosis of any lymph node demonstrating an eosinophilic infiltrate and prominent follicular hyperplasia .The disease can be seen at any age, the 2nd and 3rd decades of life being the most common affected age. Men are more commonly affected than women, in the ratio of 3:1. It presents predominantly as subcutaneous nodules in the head and neck, often unilateral, and frequently associated with regional lymphadenopathy with or without the involvement of salivary glands. Sometimes bilateral involvement is also seen. Orbit, eyelid, palate, and pharynx have also been reported to be involved, in addition to the axilla, groin and arm. The clinical course is generally benign and self-limited. Most patients have a prolonged course with gradual increase in the swelling. Occasional spontaneous resolution is seen. The exact cause and pathogenesis of KD are still unclear, although it might be a self-limited allergic or autoimmune response triggered by an unknown persistent antigenic stimulus [8,9,10,23]. Studies have also shown that the proliferation of CD4 T cells, specifically the CD4 T-helper 2 (Th2) cells and resultant overproduction of their Cytokines, such as granulocyte macrophage colony stimulating factor, tumour necrosing factor-α, IL-4, IL-5, eoxtaxin, and RANTES trigger the production of lym-phoid follicle and high IgE. Clonal T-cell population attributes to the disease devel-opment and recurrence. The immune reaction that is believed to be the roof of KD also predisposes patients to allergic conditions like asthma, chronic urticaria, pruritus and rhinitis [9,11]. Moreo-

ver, up to 60 % of these patients exhibit renal involvement manifesting as extra-membraneous glomeru-lonephritis and nephrotic syndrome. Although there is no specific diagnostic feature of this disease, FNA is helpful in preoperative diagnosis of KD. Smears show signif-icant numbers of eosinophils in a background of lymphoid cells with occasional fragments of collagenous tissues and Warthin-Finkeldey polykaryocytes [11,12,14,19,20]. In our case, the microscopic investigation of the resected specimen revealed a mixed inflammatory infiltrate extending into adjacent structures (fat cells, blood vessels, muscle fibers), consisting mainly of numerous eosinophils, and lymphocytes,with scattered lymphoid follicles with germinal centers, which are hyperplastic. The immunophenotype were CD20(+), CD3(+)B, T-lymphocytes, CD68(+) in histiocytes and Ki-67(MIB)(+).

Hui et al, classified the histological features of KD as constant, frequent and rare. The constant features included preserved nodal architecture, florid germinal center hyperplasia, eosinophilic infiltration and post-capillary venule proliferation. Frequent features comprise sclerosis, polykaryocytes, vascularization of the germinal centers, proteinaceous deposits in the germinal canters, necrosis of germinal centers, eosinophilic abscesses and reticular IgE deposition within germinal centers. The solitary rare feature is the progressive transformation of the germinal centres. Nodal architecture is largely preserved in most cases, however, capsular fibrosis with subcasular sinusoid obliteration and soft tissue involvement is frequently present [13,15,16,24].

The diagnosis of KD is not easy and differential diagnosis includes Angiolymphoid Hyperplasia with Eosinophilia (ALHE), Hodgkin's disease, Kaposi sarcoma, eosinophilic granuloma, epithelioid hemangioma, Castleman's disease, tuberculosis, dermatopathic lymphadenopathy, lymphadenopathy of drug reactions, parasitic lymphadenitis, eosinophilic granuloma, epitheloid hemangioma,-malignancy and many more. The closest differential is ALHE. Clinically, both conditions present as soft tissue swelling arising in the head and neck region with prolonged indolent clinical course. Microscopically, both show eosinophilic infiltrates and vascular proliferations. But there are few characteristic and distinctive clinicopathologic features that differentiate the two entities. KD occurs predominantly in Asians, with a male predilection [3,17,18]. Patients usually have peripheral eosinophilic and elevated serum IgE levels. The solitary lesions are mostly in the subcutaneous tissues, frequently associated with regional lymphadenopathy and salivary gland involvement. By contrast, ALHE occurs in all racial groups with a slight female predominance. Patients present with small, superficial dermal populonodules, frequently erythematous, accompanied by bleeding, pruritis, and tumor growth. Regional lymphadenopathy, serum eosinophilia, and increased IgE levels are rare [25,27,28,33]. Histologically, KD has three components: cellular (inflammatory infiltrate including increased eosinophils and follicular hyperplasia), fibrocollagenous and vascular (arborizing vascular proliferation of the post-capillary venule, endothelial cells are usually flat and lack cytologic atypia or vacuolization) In contrast to KD, vascular proliferation is most significant in ALHE, forming aggregates or lobules comprising of plump endothelial cells with epithelioid or histiocytoid changes demonstrating cytologic atypia and vacuolization [29,31].

Imaging studies might be diagnostic and can help in staging the extent and progression of the disease as well as the lymph node involvement [21]. The diagnosis in our case was only through the histopathology examination of the excised tissue. Thus, the cytological features of reactive hyperplasia with the presence of eosinophils should at least clinch the diagnosis of KD at cytology in appropriate clinical setting. The therapeutic management of KD includes surgical excision, steroids and radiation. The surgical excision may be considered first especially for the localized lesion, even if recurrence is possible. Systemically admistered steroids show good effects on disease progression, however, withdral of steroids can often result in relapse. Radiation has been utilized for steroid resistant lesions [30,32,34].

5. Conclusion

Despite the rarity of Kumura's disease's occurrence, it must bear in mind of clinicians that this nosological entity exist, especially in the seating of subcutaneous lump's differential diagnosis in any patient no mater his origin (African, European, Asian ...) or gender (male/Female) . Morover this disease affects more people from Asia, in our case we are dealing with a Greek man without any consanguinity with Asiatic Countries in whom KD of a very rare anatomical location was diagnosed from the resected right thigh's subcutaneous mass. Therefore, we suggest pathological investigation in case of any mass presented by a patient.

6. Ethical Considerations

A written consent has been obtained from the patient, and the case is described and published without patient's identity.

7. Teaching Point

Never neglect any palpable mass presented by the patient to find out the final historical diagnosis in any age-Group.

References

1. Malhotra M, Varshney S, Singh N, Kumar S. Kimura disease: a rare case report from Himalayan Region. Indian J Otolaryngol Head Neck Surg. 2017; 69(4): 571-4.
2. Zhou P, Zhang W, Shi W, Zhang Y. Kimura disease. Dermatol Online J. 2017; 23(10): 16.
3. Kimura T, Yoshimura S, Ishikawa E. On the unusual granulation com-bined with hyperplastic changes of lymphatic tissues. Trans Soc Pathol Jpn. 1948; 37: 179–80.
4. Jiang Y, Hua QQ, Ren J, Zeng F, Sheng JF, Liao H, et al. Eosinophilic hy-perplastic lymphogranuloma: clinical diagnosis and treatment experience of 41 cas-es. Am J Otolaryngol. 2017; 38: 626-9.

5. Ye P, Ma DQ, Yu GY, Gao Y, Peng X. Comparison of the efficacy of dif-ferent treatment modalities for Kimura's disease. Int J Oral Maxillofac Surg. 2017; 46(3): 350-4.
6. Hosaka N, Minato T, Yoshida S, Toki J, Yang G, Hisha H, et al. Kimura's disease with unusual eosinophilic epithelioid granulomatous reaction: a finding pos-sibly related to eosinophil apoptosis. Hum Pathol. 2002; 33(5): 561-4.
7. Woo SH, Kim HK, Kim WS, Bae TH, Kim MK. A rare case of Kimura disease with bilateral parotid involvement. Arch Plast Surg. 2017; 44(5): 439-43.
8. Wang DY, Mao JH, Zhang Y, Gu WZ, Zhao SA, Chen YF, et al. Kimura disease: a case report and review of the Chinese literature. Nephron Clin Pract. 2009; 111(1): c55-61.
9. Rascovan N, Monteil BS, Grob JJ, Collet-Villette AM, Gaudy-Marqueste C, Penicaud M, et al. Human polyomavirus-6 infecting lymph nodes of a patient with an angiolymphoid hyperplasia with eosinophilia or kimura disease. Clin Infect Dis. 2016; 62(11): 1419–21.
10. Tanaka Y, Ueno Y, Shimada Y, Yamashiro K, Tanaka R, Urabe T, et al. Paradoxical brain embolism associated with Kimura disease mimics watershed in-farction. J Stroke Cerebrovasc Dis. 2015; 24(2): e55-7.
11. Ye X, Feng Y, Lin SX. Pulmonary embolism as the initial clinical presen-tation of Kimura disease: case report and literature review. Blood Coagul Fibrinoly-sis. 2015; 26(4): 414–8.
12. Baba A, Ojiri H, Dogru M, Tanaka Y, Takahashi S, Mogami T, et al. An unusual clinical presentation of Kimura disease manifesting with a typical cephalo-cervical lesion and an atypical subcutaneous hip mass lesion. Intern Med. 2016; 55(8): 1017-20.
13. Zhang R, Ban XH, Mo YX, Lv MM, Duan XH, Shen J, et al. Kimura's dis-ease: the CT and MRI characteristics in fifteen cases. Eur J Radiol. 2011; 80(2): 489–97.
14. Choi WJ, Hur J, Ko JY, Yeo KY, Kim JS, Yu HJ, et al. An unusual clinical presentation of Kimura's disease occurring on the buttock of a five-year-old boy. Ann Dermatol. 2010; 22(1): 57-60.
15. Osuch-Wojcikiewicz E, Bruzgielewicz A, Lachowska M, Wasilewska A, Niemczyk K. Kimura's disease in a caucasian female: a very rare cause of lymphad-enopathy. Case Rep Otolaryngol. 2014; 2014: 1–4.
16. Panduranga KM, Bhojwani KM, Bhandarkar AM, Pai RR, Rent NH. Angi-olymphoid hyperplasia with eosinophilia of root of nose: a rare phenomenon. J Clin Diagn Res. 2014; 8(3): 144.
17. Gregory X, Soon NI, Nur AR. A rare case of inguinal kimura disease. Med J Malaysia. 2018; 73(5): 326–7.
18. Gambichler T, Luther H, Bararach-Buhles M, Altmeyer P, Stucker M. Generalized Kimura's disease masquerading as nodular prurigo. Clin Exp Dermatol. 2007; 32(1): 97–9.
19. Rajpoot DK, Pahl M, Clark J. Nephrotic syndrome associated with Kimura disease. Pediatr Nephrol. 2000; 14(6): 486–8.
20. Obata Y, Furusu A, Nishino T, Ichinose H, Ohnita A, Iwasaki K, et al. Membranous nephropathy and Kimura's disease manifesting a hip mass. A case re-port with literature review. Intern Med. 2010; 49(14): 1405–9.
21. Park SW, Kim HJ, Sung KJ, Lee JH, Park IS. Kimura disease: CT and MR imaging findings. AJNR Am J Neuroradiol. 2012; 33(4): 784–8.
22. TJ Li, XM Chen, SZ Wang, MW Fan, I Semba, M. Kitano, et al. Kimura's disease. A clinicopathologic study of 54 Chinese patients. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1996; 82(5): 549–555.
23. Ranka SR, Rajput A, Kantharia CV. Kimura's disease: Rare cause of head & neck swelling. Indian Journal of Otolaryngology and Head and Neck Surgery. 2004; 56(1): 43–45.
24. Armstrong WB, Allison G, Pena F, Kim JK. Kimura's disease: two case reports and a literature review. Annals of Otology, Rhinology and Laryn-gology. 1998; 107(12): 1066–1071.
25. Szporek B, Steplewska K, Cieslik T. Kimura's disease-case re-port," Czasopismo Stomatologiczne. 2006; LIX(10): 734–739.
26. Rajpoot DK, Pahl M, Clark J. Nephrotic syndrome associated with Kimura disease. Pediatric Nephrology. 2000; 14(6): 486–488.
27. Varshney MK, Kumar A, Khan SA, Yadav CS. Kimura dis-ease of extremity: unusual manifestation in a long bone. Joint Bone Spine. 2008; 75(4): 492–494.
28. Kanazawa S, Gong H, Kitaoka T, Amemiya T. Eosinophilic gran-uloma (Kimura's disease) of the orbit: a case report. Graefe's Archive for Clinical and Experimental Ophthalmology. 1999; 237(6): 518–521.
29. Abuel-Haija M, Hurford MT. Kimura disease. Archives of Pa-thology and Laboratory Medicine. 2007; 131(4): 650–651.
30. Day TA, Abreo F, Hoajsoe DK, Aarstad RF, Stucker FJ. Treatment of Kimura's disease: a therapeutic enigma. Otolaryngology—Head and Neck Surgery. 1995; 112(2): 333–337.
31. Iwai H, Nakae K, Ikeda K. Kimura disease: diagnosis and prog-nostic factors. Otolaryngology—Head and Neck Surgery. 2007; 137(2): 306–311.
32. Aoki T, Shiiki K, Naito H, Ota Y. Cytokine levels and the effect of prednisolone on kimura's disease: report of a case. Journal of Oral and Maxillofacial Surgery. 2001; 59(10): 1238–1241.
33. Sakamoto M, Komura A, Nishimura S. Hematoserological analy-sis of Kimura's disease for optimal treatment. Otolaryngology—Head and Neck Surgery. 2005; 132(1): 159-160.
34. Hareyama M, Oouchi A, Nagakura H. Radiotherapy for Kimura's disease: the optimum dosage," International Journal of Radiation Oncology Biology Physics. 1998; 40(3): 647–651.