

Benign Inflammatory Lesion Mimicking Malignancy “Kimura’s Disease”: A Case Report

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ABSTRACT

Introduction: Kimura’s disease (KD) in Indonesia is quite rare. It is a distinct benign reactive process; however, it is usually mimicking malignancy. It usually affects young adults with a peak incidence in three decades, and men are mostly affected compared to women with a 3:1 ratio. KD is a chronic inflammatory disorder characterized by lymphoid hyperplasia, eosinophilia, and associated with soft tissue swelling that usually arises in head and neck regions with regional lymphadenopathy.

Case Presentation: Herein, we report a case in a 55-year-old man from East Indonesia who presented a huge lump on his left head and neck regions. The lump started about 8 years ago. Physical examination found a tumor mass of 20 x 10 cm with indistinct border and multiple lymphadenopathies left coli with the biggest size of 1.3 x 0.7 x 0.5 cm. Peripheral blood examination shows marked eosinophilia. Nasopharyngeal CT scan revealed soft tissue masses and multiple lymphadenopathies suspected of malignancy; however, thorax plain radiology did not show any sign of metastatic process. Histopathological examination shows reactive follicular proliferation, extensive eosinophilia, focal area Folliculolysis, and polykaryocytes of the Warthin-Finkeldey type.

Conclusion: Based on a complete history, laboratory findings, and histopathological examination, this case is concluded as Kimura’s disease. Although clinical findings and radiological examination suspected it as malignancy, those discrepancies were confirmed as a benign lesion by specific histopathological and laboratory findings. Commonly, most patients have a favorable prognosis and good response to therapy.

INTRODUCTION

In Indonesia, Kimura’s disease is quite rare although some literature states its endemic in Asia especially in Japan and China [1,2]. It is a chronic inflammatory disorder which is frequently mimicking malignancy. There is a marked male predominance than the female with a ratio of 3:1 [1]. The peak age onset is the third decade. The causes of Kimura’s disease are unknown yet. Commonly, it appears as a painless soft tissue mass with chronic inflammatory changes of the subcutaneous lymph nodes [3]. Up to 40% of patients present with two or more subcutaneous masses. A small subset of patients present with isolated lymph node involvement. Most patients with Kimura’s disease have peripheral blood eosinophilia (10 – 50% in differential count), and the subcutaneous lesion of Kimura disease is characterized by lymphoid infiltrate with follicle and germinal centers, abundant eosinophil and vascular proliferation of small capillaries [4,5].

Interestingly, this case report discusses an older man of 55 years old with a huge mass on his left head and neck regions. Clinical findings alone were difficult to reveal the diagnosis of Kimura’s disease, and clinicopathology assessment is needed to confirm the diagnosis. Thus, it becomes very challenging because the first clinical and radiological findings suspected malignancy lesions. After the clinicopathological conference, it was confirmed by specific histopathology findings, and the laboratory result was supported as Kimura’s disease which is often mimicking malignancy.

CASE PRESENTATION

A 55-year-old man from east Indonesia came with a chief complaint of a huge lump on his cheek for almost 8 years (**Figure 1**). It started as a small mobile lump at first at the left jaw, got bigger, and then

extended to the upper part cheek since a year ago. The past medical history like another huge lump at the other body part was denied, and another systemic disease was unknown yet. Besides a lump, the patient also said that he had heavy headache as well. Another lump or mass was also felt at the left neck.



Figure 1. A huge lump on left cheek with indistinct borders



Figure 2. Head CT Scan. Soft tissue mass regio left Zygomatoc mandibula with necrosis area

Physical examination found a tumor mass of 20 x 10 cm with indistinct borders and solid elastic consistency on the zygomatic area. On coli sinistra, there was also a small lump of 2 x 1 cm, mobile and solid. Laboratory

examination shows a significant increase in eosinophil from 1.29 103/ μ L (15.48%) to 3.92 103/ μ L in December 2019 and a slight elevation of liver function test AST 37.1U/L (11-33) and ALT 56U/L (11-50). Unfortunately, LDH status and other tumor markers were not performed yet by the clinician. Nasopharyngeal CT scan revealed soft tissue mass region zygomatocum extending to left submandible, with indistinct borders and multiple lymphadenopathies in coli sinistra with the widest size of 1.3 x 0.7 x 0.5 cm. There is also a necrosis area inside it however, there is no erosion or destructed bone around the mass (Figure 2). However, Thorax plain radiology did not show any sign of metastatic process (Figure 3). Based on clinical and radiologic findings, the clinician diagnosis a suspected malignant tumor on the left coli sinistra, and the patient was planned for incisional biopsy.

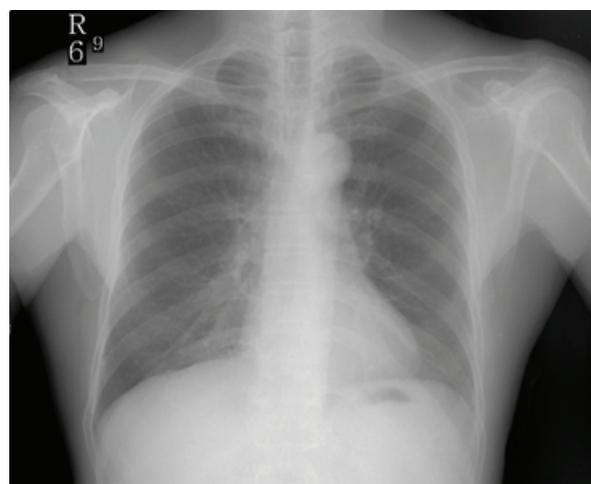


Figure 3. Thorak plain examination. There is no metastatic process.

The incisional biopsy was performed at the soft tissue coli sinistra region. The specimen was sent to a pathological laboratory, and a brown and solid tissue of 1.9 x 1 x 0.7 cm was received (Figure 4). The whole specimen was histopathologically examined.



Figure 4. Macroscopic picture in cassette

The microscopic examination with hematoxylin and eosin staining shows that the tissue consists of reactive lymphoid, vascular proliferation, and some eosinophil infiltrated centrum germinativum (**Figure 5A**). There was also a large amount of eosinophil in the perifollicular area (**Figure 5B**) and hyalinized capillary vessels (**Figure 5B, inset**). Moreover, some lymphoid follicles showed folliculolysis with apoptotic cell distribution in between (**Figure 6A**). There is also marked eosinophilia permeates in between muscles (**Figure 6B**).

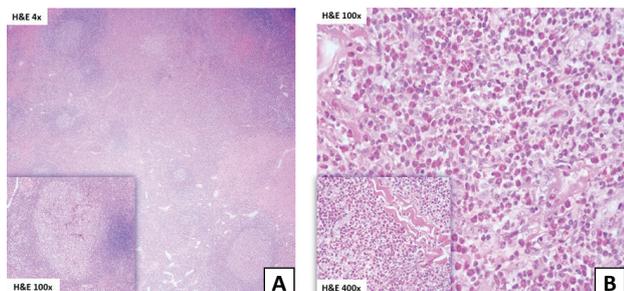


Figure 5. Microscopic findings. A. Reactive lymphoid and vascular proliferation (H&E stain, magnification 4x) and some eosinophil infiltrated the centrum germinativum (H&E stain, magnification 100x) B. Marked eosinophilia (H&E stain, magnification 100x) and hyalinized capillary vessels wall (inset, H&E stain, magnification 400x)

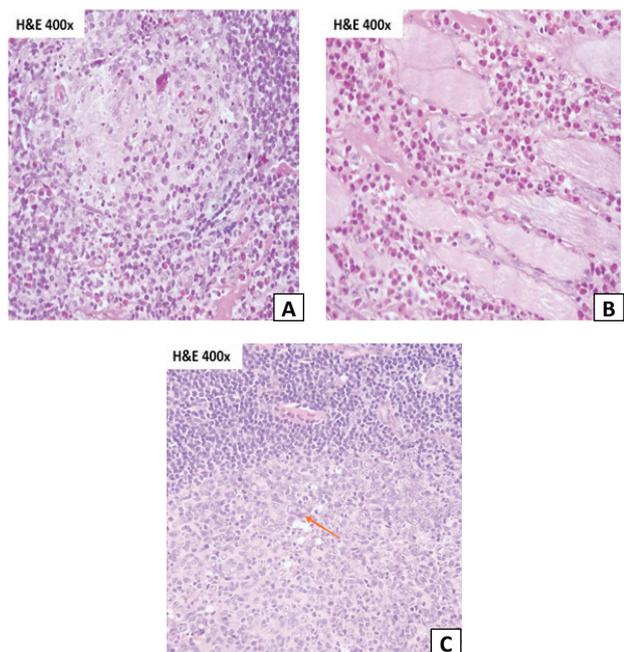


Figure 6. Microscopic findings. A. Folliculolysis with apoptotic cells distribution in between B. Marked eosinophilia permeate between muscle C. Polykaryocytes of the Warthin-Finkeldey type, characterized by the overlapping, grape-like arrangement of nuclei (red arrow)

DISCUSSION

Kimura’s disease is a benign mimicking malignant disease that is usually difficult to reveal clinically. This

unique disease predominantly affects the head and neck area of young adults with a peak age incidence in the third decade [1,2]. However, this case was an older male patient aged 55 years with a contrast epidemiologic compared with other literature [1,3–6]. Based on the anamnesis, the patient had suffered since 8 years ago, which means it is a long chronic inflammatory disease and the soft tissue lesion restricted at the head and neck area and some regional lymphadenopathy was found with normal overlying skin. On the other hand, laboratory result shows marked eosinophilia, and radiologic examination does not show any destructed bone nor metastatic process at the thorax plain radiology supported as a benign lesion. Histopathological examination revealed there were reactive lymphoid and vascular proliferation and extensive eosinophil infiltrating the centrum germinativum. In addition, this eosinophil infiltration also enters between muscle areas. Besides that, there was also folliculolysis with apoptotic cells distribution in between and scattered polykaryocytes of the Warthin-Finkeldey type, characterized by the overlapping, grape-like arrangement of nuclei. Those all above supported the diagnosis of Kimura’s disease.

Kimura’s disease is a chronic inflammatory disorder. It involves subcutaneous tissues and lymph nodes predominantly in the head and neck regions and is characterized by angiolymphoid proliferation and eosinophilia [4–6]. The diagnosis of Kimura’s disease needs clinical correlation confirmed by histopathology and laboratory findings. The other specific findings were increased mast cells and plasma cells, especially at the paracortex area. However, we had not found any eosinophilic microabscesses yet. There was also capillaries proliferation with a hyalinized wall. Kimura’s differential diagnosis of the disease is angiolymphoid hyperplasia with eosinophilia (ALHE) owing to some histologic similarities. ALHE clinically found in Caucasian women without any sign of regional lymphadenopathy and no marked increase of eosinophil on blood serum examination. However, Asian male and peripheral blood eosinophilia 10% to 50% are constant features of Kimura’s disease [4]. Essentially, Kimura’s disease is located deep in the subcutaneous tissue and in almost all cases involving the regional lymph nodes [4,5]. In contrast, ALHE is marked by papul lesion restricted at the superficial dermis.

We also had conducted a clinicopathological conference and concluded it as Kimura’s disease because, first, it was a deep-seated soft tissue lesion with concomitant involvement of lymph node locally located only at head and neck area without any other lymph node enlargement involvement and clinical “B symptom” for Hodgkin lymphoma. Then, we suggested that it did not support the diagnosis of classic HODGKIN Lymphoma especially NSCHL and MCCHL which also had eosinophil as their background and histopathology examination did

not reveal any lacunar and HRS cells [7,10]. Second, other differential diagnoses would be Kikuchi lymphadenitis or Fujimoto disease. It usually affects young women and is histopathologically characterized by histiocytes and lymphocytes [4–6]. It also has basophilic material as small clumps (hematoxylin bodies) or is deposited around blood vessels (Azzopardi effect) [4,5,10]. Third, the other lesion like cutaneous macrocytosis might also become one of differential diagnosis. However, clinically, it happens at birth or during the first 3 months of life with overlying skin showing macule or papule with Darier’s sign (stroking release histamine cause hives) and dermatographism (dermal edema). Histopathologically, it is characterized by a monomorphic population of mast cell predominant in the papillary dermis and rare eosinophils [4,5,11].

We did not perform immunohistochemistry examination to exclude other lesions as we assured that it was soft tissue mass although associated with regional lymphadenopathy with normal overlying skin (excluding macrocytosis). Another examination of the plain thorax revealed no metastatic process, and the nasopharyngeal shows no bone destruction. The treatment of Kimura’s disease includes surgical excision, steroid, and radiation [5]. Japanese literature said that almost 25% of patients with surgical therapy alone will suffer from remission [8]. Surgical therapy combined with steroid therapy can be an option. Then, radiation itself is preferred for an unresponsive lesion of steroid therapy or remission of surgical therapy [8,12]. This patient underwent incisional biopsy followed by radiation combined with low-dose steroid therapy. The lesion was reduced in size after the third radiation therapy, and there is no other new nodal involvement so far.

CONCLUSIONS

Our Kimura’s case was unique and rare because it affects an older man with a huge mass on head and neck regions and multiple regional lymphadenopathies which were clinically suspected as a malignant neoplasm. For the precise diagnosis, it needs to be confirmed by laboratory and histopathological examination. Therefore, clinical findings alone could not reveal this disease. Thus, complete history taking, clinical findings, and histopathological examination are required to reveal the precise diagnosis of Kimura’s Disease.

DECLARATIONS

Competing of Interest

The author(s) declare no competing interest in this paper.

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