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Case report

Unhabitual presentation of Kimura disease: A case report of gastric localization

Wafa Mokni[1], Ahlem Bdioui[1,2], Dorsaf Beltaifa[1,2], Marwa Krifa[1,2], Fethia Harrabi[3], Oussama Belkacem[1,2], Sihem Hmissa[1,2], Moncef Mokni[4]

[1] *Department of Pathology, Sahloul University Hospital of Sousse, Tunisia*
[2] *Research Laboratory Oncogenesis and Tumoral Progression (LR21ES03), Faculty of Medicine of Sousse, University of Sousse, Tunisia*
[3] *Department of Visceral Surgery, Sahloul University Hospital of Sousse, Tunisia*
[4] *Department of Pathology, Farhat Hached University Hospital of Sousse, Tunisia*

Abstract

Background: Kimura's disease (KD) is a rare, chronic inflammatory disorder with a benign course and an unclear cause. It primarily affects the head and neck regions. KD is often misdiagnosed due to its resemblance to other benign or malignant diseases, especially when it occurs in uncommon sites. In this report, we present a rare case of KD diagnosed in the stomach.

Methods: We report a case of KD localized in the stomach, diagnosed at the Pathology Department of Sahloul University Hospital in Sousse. A 75-year-old female presented with intestinal bleeding. Endoscopy revealed erythematous and hemorrhagic mucosa in the antral and fundic regions of the stomach. A 4 cm antral polyp was identified and subsequently resected.

Results: Initial examination of the biopsy specimen concluded with a diagnosis of a low-grade dysplastic polyp. Partial gastrectomy was performed. Histologically, the polyp was bordered by focally ulcerated epithelium without evidence of dysplasia. The axis was edematous, containing clusters of lymphoid cells dispersed within a moderate inflammatory infiltrate rich in eosinophils. Characteristic multinucleated cells with crown-arranged nuclei were observed. The lesion was well vascularized. Immunohistochemistry for CD117, Dog-1, S100 protein, SMA, and ALK was performed to exclude other diagnoses. The results were negative. After multidisciplinary consultation and expert opinion, a diagnosis of KD was established.

Conclusion: The stomach is an extremely rare localization for KD. Misdiagnosis can result in unnecessary aggressive treatments, making it essential to distinguish KD from other diseases. While KD generally has a good prognosis with no risk of malignant transformation, its high recurrence rate necessitates careful and ongoing monitoring of affected patients.

Keywords: Kimura's disease, stomach, Angiolymphoid hyperplasia with eosinophilia

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1. Introduction

Kimura's disease (KD) is a rare, chronic inflammatory disorder with a benign course and an unclear etiology. KD was first described in a Chinese study in 1937, where it was initially believed to be an "eosinophilic proliferative lymphogranuloma." In 1948, Kimura in Japan provided the definitive histological description, leading to the disease being named after him [1].

The primary regions affected are the head and neck, although KD can also occur in the groin, extremities, and trunk. Rare cases have been described in the hard palate, larynx, and median nerve. Digestive tract involvement is exceptionally rare [2]. The disease predominantly affects young Asian males and is often misdiagnosed due to its resemblance to other benign or malignant conditions, especially in uncommon sites. In this study, we present a rare case of KD diagnosed in the stomach.

***Correspondence:** Wafa Mokni. Department of Pathology, Sahloul University Hospital, Sousse, Tunisia. Email: wafa.mokni@yahoo.com

2. Case report

A 75-year-old female with a medical history of multiple sclerosis and angiodysplasia of the colon presented with intestinal bleeding. Physical examination revealed no significant findings. The patient was stable, with no signs of acute distress or abnormalities on inspection or palpation.

An endoscopy revealed erythematous and hemorrhagic mucosa in the antral and fundic regions of the stomach. A 4 cm antral polyp was identified but showed no stigmata of active bleeding. A computed tomography (CT) scan of the abdomen demonstrated an antral polypoid mass located on the posterior wall of the stomach, with no evidence of distant extension or metastasis. Routine blood tests showed normal white blood cell count and hemoglobin levels. A biopsy identified a low-grade dysplastic polyp. Partial gastrectomy was performed. Histological examination revealed that the polyp was bordered by a focally ulcerated regenerative epithelium without dysplasia. The stroma was edematous and contained clusters of lymphoid cells dispersed among a moderate inflammatory infiltrate rich in eosinophils (Fig. 1-

2).

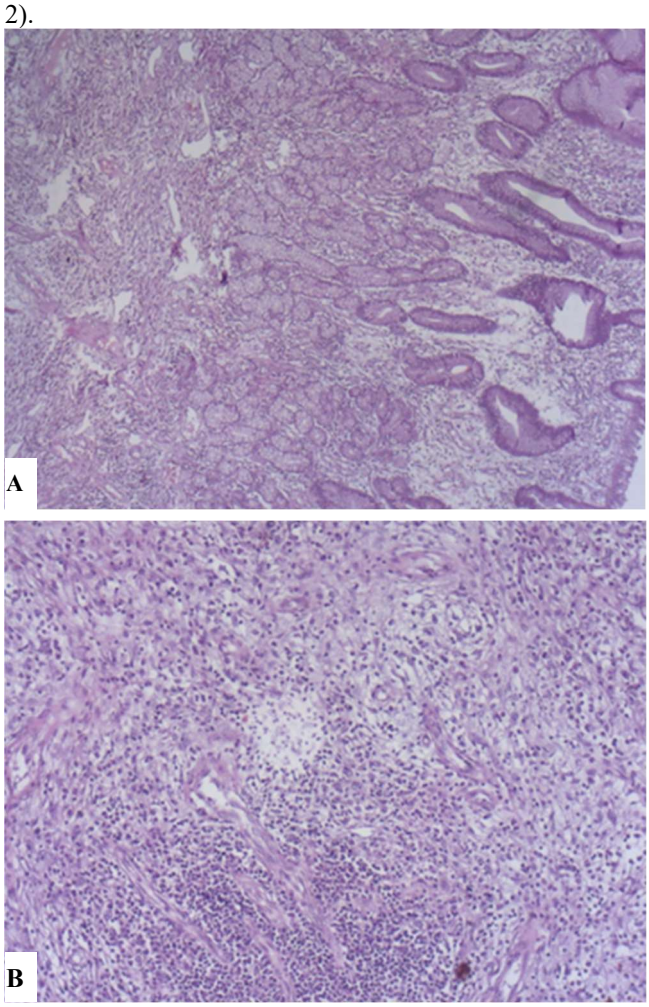
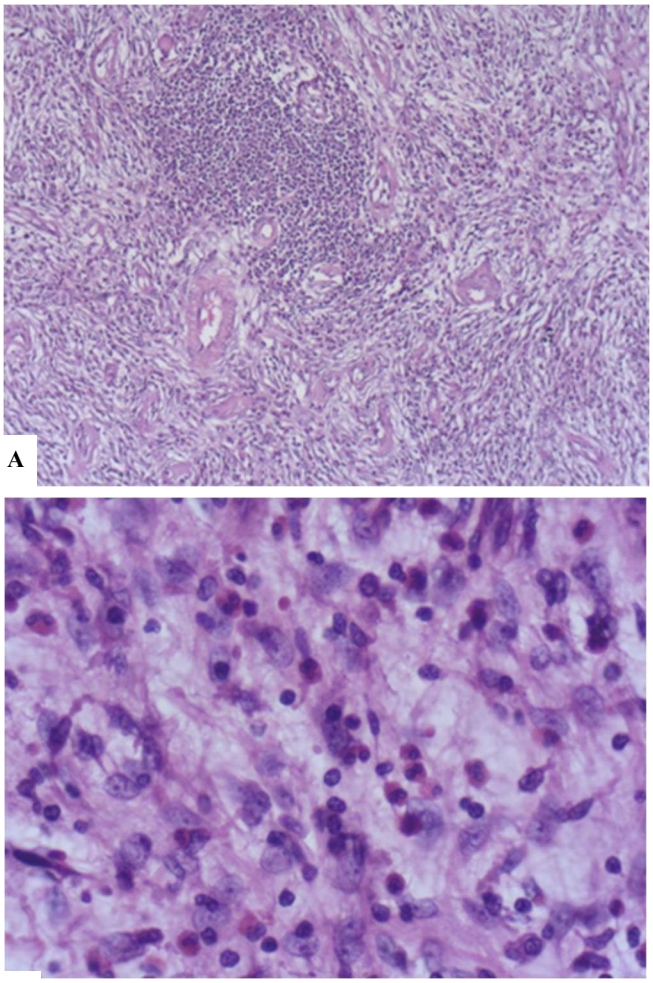


Fig.1. (A): Normal gastric mucosa with underlying sub-mucosa lesion (x50, HE). (B): the lesion is composed of inflammatory cells and hyalinized vessels (Gx200, HE).



2. (A): Lymphoid clusters (x200, HE). (B): Note the eosınophilic infiltrate (x400, HE).

Characteristic multinucleated cells with crown-arranged nuclei were observed (Fig. 3). The lesion was well vascularized. Immunohistochemistry for CD117, DOG-1, S100 protein, smooth muscle actin (SMA), and anaplastic lymphoma kinase (ALK) was conducted to rule out other diagnoses, particularly gastrointestinal stromal tumor, inflammatory myofibroblastic tumor, and inflammatory fibroid polyp (Fig. 4). After expert consultation and multidisciplinary discussion, the diagnosis of KD was confirmed.

3. Discussion

Kimura's disease (KD) is a rare, chronic inflammatory condition of unknown etiology that predominantly affects young and middle-aged Asian males, suggesting a potential genetic component [1]. KD typically manifests as solid, painless, or itchy subcutaneous lesions, primarily in the parotid and submandibular regions, often accompanied by lymph node enlargement. Multiple masses have been reported in only one case by Yu et al. [3], involving the neck, parotid region, and lymphadenopathies in the thoracic wall, axillary, and inguinal regions. Elevated serum immunoglobulin E (IgE) levels and eosinophilia are common findings in KD. The exact cause of this condition remains unclear, but several potential contributing factors have been identified, including allergic reactions, Candida infections, arthropod bites, and alterations in systemic immune responses [1].

Histopathologically, KD is characterized by pronounced reactive follicular hyperplasia with germinal centers surrounded by abundant eosinophils, lymphocytes, and mast cells. Micro-abscesses may also be observed. The vessels in KD lesions are numerous and characterized by thin walls lined with cuboidal endothelial cells. Polykaryocytic Warthin-Finkeldey-type giant cells are frequently observed [1,4]. These histological features are consistent regardless of the anatomical site of involvement.

Diagnosing KD can be challenging due to its similarities with other conditions, including malignant hematological diseases, Eosinophilic Granulomatosis with Polyangiitis (EGPA), and IgG4-Related Disease (IgG4RD). Histological confirmation is required for an accurate diagnosis. In a study involving 24 cases of KD conducted by Zhang et al., the accuracy of core-needle biopsy was found to be low (2/14, 14.3%) [5], indicating that surgical specimens are sometimes necessary for confirmation. In our case, the correct diagnosis was made using a partial gastrectomy specimen. Misdiagnosis can lead to unnecessary aggressive treatments, making it essential to distinguish KD from other diseases.

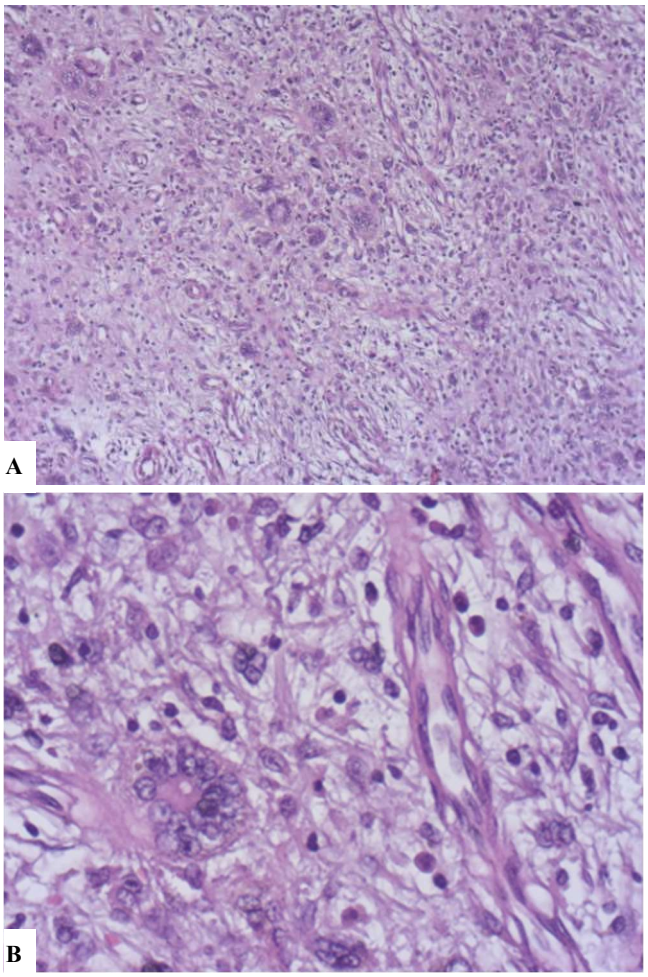


Fig.3. (A+B): Characteristic polykaryocytic Warthin-Finkeldey cells (A: Gx200; B: Gx400, HE).

EGPA is a severe, multisystem disease characterized by asthma, eosinophilia, and necrotizing vasculitis. Both KD and EGPA involve eosinophilic infiltration amidst a dense mixed inflammatory infiltrate. However, KD is marked by an increased number of vessels and varying degrees of stromal fibrosis. In contrast, EGPA is diagnosed by the presence of necrotizing vasculitis affecting small to medium-sized vessels and eosinophilic granulomas. Although granulomas are not typically associated with KD, they are well-documented in the disease. Additionally, renal involvement has been reported in KD, with approximately 16% of patients developing proteinuria and glomerulonephritis, ranging from minimal change disease (MCD) to more aggressive forms like mesangioproliferative glomerulonephritis [4].

Angiolymphoid hyperplasia with eosinophilia (ALHE) can also be confused with KD. Previously, ALHE was considered synonymous with KD. Currently, it is recognized as a type of endothelial neoplasm related to inflammatory stimulation. KD, on the other hand, is a chronic inflammatory disorder [2]. While ALHE shares some similarities with KD, it can be distinguished by its histological features and its typical presentation as a skin lesion, although rare occurrences in the gastrointestinal tract have been noted for both conditions. A rare case of ALHE in a 63-year-old male was reported by Berney et al., who presented with severe gastrointestinal hemorrhage. The nodule found and biopsied in this case consisted of inflammatory cells, primarily lymphocytes and eosinophils, infiltrating the submucosa and extending into the muscularis propria [7]. However, we did not identify any gastric KD cases in our literature review, making it an extremely rare localization.

In our case, other less common differential diagnoses must also be considered, such as inflammatory fibroid polyps and inflammatory myofibroblastic tumors. Immunohistochemistry may be useful in distinguishing these conditions.

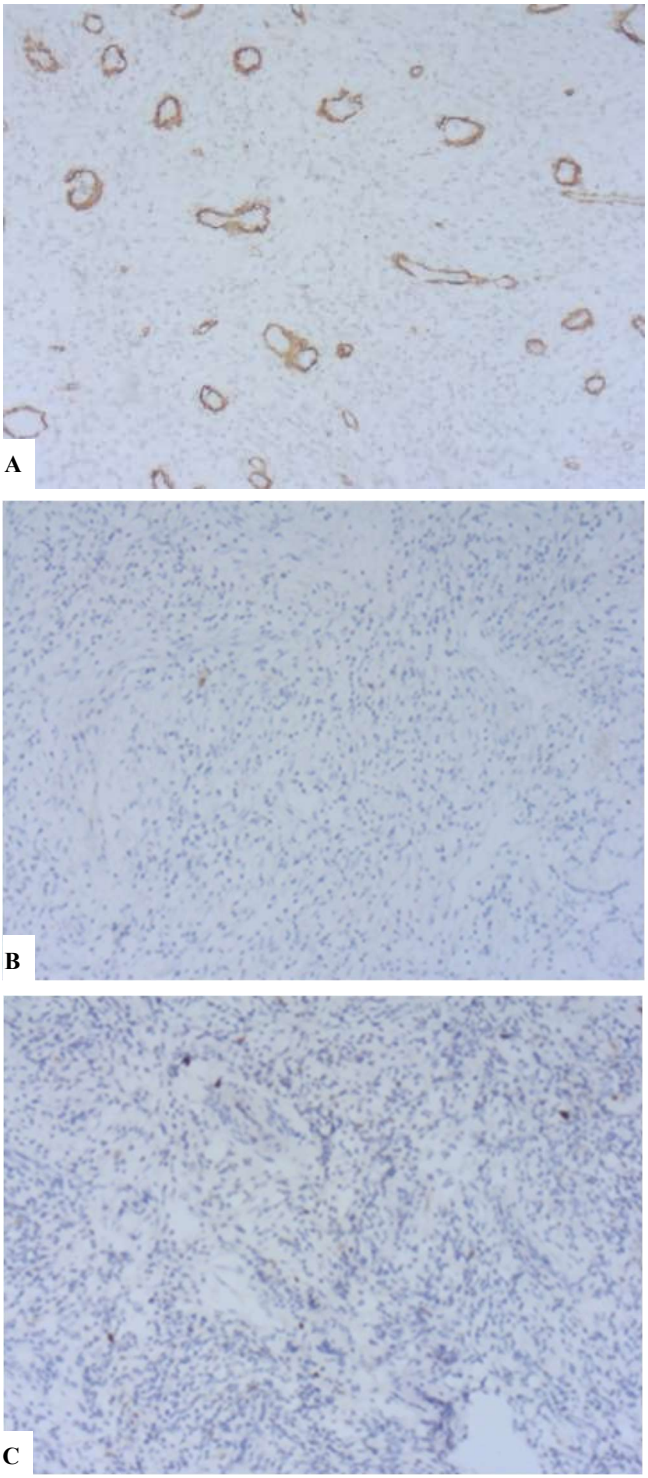


Fig.4. Representative immunohistochemistry slides: negativity for SMA (A), C-kit (B) and ALK (C) immunostaining (x50, IHC).

Effective treatments for KD include surgery, glucocorticoids, immunosuppressants, and low-dose radiotherapy. Despite the availability of treatments, KD has high recurrence rates, reaching up to 62% [1,3]. Factors contributing to a higher risk of recurrence include a peripheral blood eosinophil count exceeding 50%, serum total IgE levels above 1000 U/mL, and the presence of multiple lesions beyond the salivary glands [8]. Some studies have shown that eosinophil counts are linked to the therapeutic response in KD patients and may serve as a predictive marker for the disease [3].

4. Conclusion

Most patients with KD are young and middle-aged Asian males, and specific genes may play a role in the pathogenesis of this disease. KD is a rare and complex condition requiring a multidisciplinary approach for accurate diagnosis and effective management. A combination of clinical, biological, and histological investigations is essential to differentiate KD from other similar conditions and to avoid unnecessary aggressive treatments. While KD generally has a good prognosis with no risk of malignant transformation, its high recurrence rate necessitates careful and ongoing monitoring of affected patients. This study emphasizes the need for increased awareness of KD among physicians and pathologists to ensure accurate diagnosis and treatment, as its features overlap with many other diseases and tumors, and any organ can be involved.

Consent of patient

Written informed consent was obtained from the patient for participation in this study.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Conflict of interest

The authors declare that there are no conflicts of interest that could be perceived as influencing the impartiality of the research presented.

Authors' contribution

The authors participated equally.

Availability of data and materials

All data underlying the manuscript are available as part of the article.

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