A crick in the neck: the curious case of Kimura disease in an adolescent female

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BACKGROUND

Kimura disease (**KD**) is a rare, benign, chronic inflammatory disorder of unknown etiology. It is typically characterized by a solitary subcutaneous nodule localized to the head and neck region, with localized lymphadenopathy and occasional salivary gland enlargement. KD is described almost exclusively in Asians, mostly in adult males with a peak age of onset in the third decade of life. KD shares clinical and histopathologic features with **IgG4-related disease** (**IgG4-RD**), another chronic inflammatory disorder. IgG4-RD often exhibits fibromatous involvement in one or more organ systems, and may be confused with malignancy, infection, or other immune-mediated conditions. Correlation among clinical, serologic, radiologic, and pathologic data is required to distinguish between KD and IgG4-RD. Here we describe an adolescent female patient with a neck mass that was **originally treated as IgG4-RD, and later reclassified as Kimura disease.**

METHODS

A retrospective chart review of the patient's internal and external records.

CASE PRESENTATION

- A 14-year-old Filipino female developed a focal area of **left-sided neck swelling (Figure 1)**. She was initially treated with antibiotics for presumed infection, but without improvement.
- Her swelling gradually worsened over several months, to a maximum size of **5.5 x 3.7 x 6.3 cm**. MRI of the chest/abdomen/pelvis revealed no additional areas of involvement.

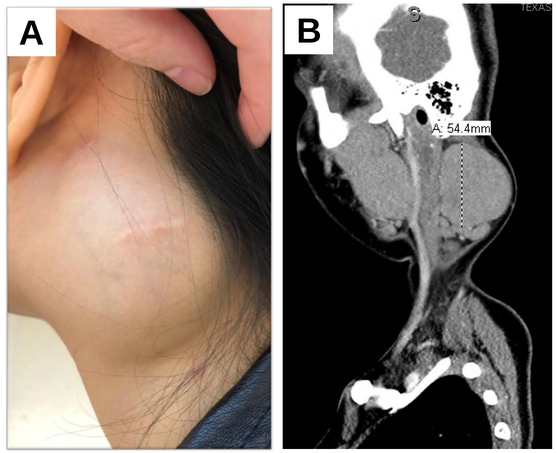

Figure 1: Left neck mass. A: Photograph of left-sided neck mass. **B**: CT scan showing large well-delineated, non-calcified homogenous, non-necrotic nodal mass.

Test (Units)	Patient Value	Reference Range
WBC (x10^3/uL)	10.5	4.5 - 13.5
Neutrophils (%)	45.6	33 - 76
Neutrophils (cells)	4788	
Lymphocytes (%)	20.3	15 - 55
Lymphocytes (cells)	2132	
Eosinophils (%)	**28.2**	0 - 3
Eosinophils (cells)	**2961**	
IgA (mg/dL)	171	66 - 295
IgE (kU/L)	**39,232**	<114
IgG (mg/dL)	1,600	641 - 1,353
IgM (mg/dL)	159	40 - 80

Table: Laboratory evaluation. Pertinent positive values are bolded.

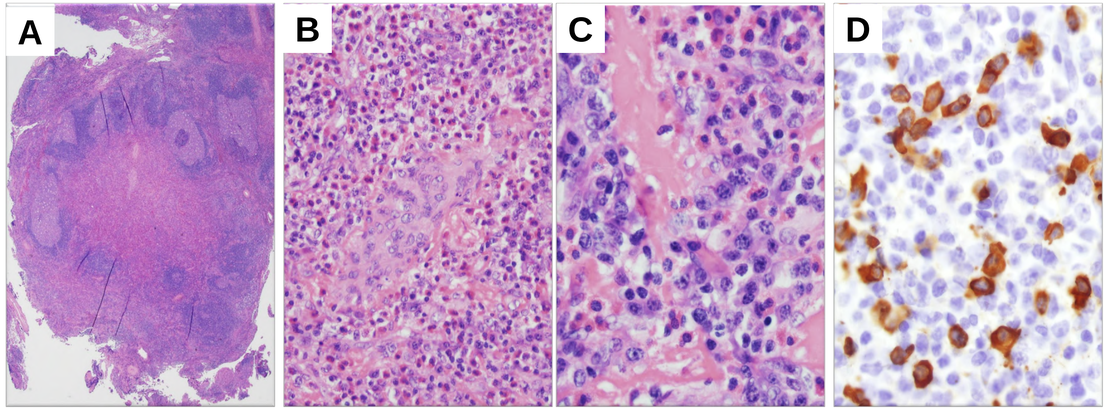

Figure 2: Histopathologic examination of tissue from left post-auricular mass. A: Hematoxylin and eosin (H&E)-stained lymphoid tissue demonstrating marked interfollicular expansion and reactive follicles (1.25x). **B**: H&E-stained tissue showing prominent eosinophilia and plasmacytosis together with plump endothelial cells (20x). **C**: H&E-stain reveals fibrotic process with elevated plasma cells and eosinophils (40x). **D**: Abundant IgG4-stained cells (>100/hpf, 40x).

CASE PROGRESSION

- **Incisional biopsy** of the mass showed reactive lymphadenopathy with follicular and interfollicular hyperplasia, increased eosinophils, focal vascular proliferation and IgG4+ cells. There was no evidence of malignancy; serum IgG4 levels were within normal.
- A working diagnosis of **IgG4-RD** was made, and the patient was started on corticosteroids with near complete response, but recrudescence with taper.
- The family moved and presented to our institution 10 months after symptom onset. She was found to have **peripheral eosinophilia** (2.96 K/uL) and highly **elevated serum IgE** (39,232 kU/L) (**Table**).
- **Re-evaluation** of her biopsy (**Figure 2**) confirmed reactive lymphoid hyperplasia with dense tissue **eosinophilia** including eosinophilic microabscesses, focal vascular proliferation, focal sclerosis and prominent **plasmacytosis with increased IgG+ cells** (100 per HPF, IgG4:IgG ratio 40%).
- Based on the patient's Asian ethnicity, focal disease without multi-organ involvement, and overall histopathologic findings, the diagnosis of **Kimura disease** was made. The mass was excised for definitive therapy (**Figure 3**). She has no evidence of renal disease, which can be present in KD (20%).

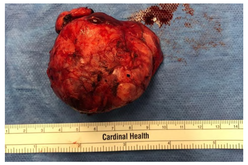

Figure 3: Gross surgical specimen.

CONCLUSION

KD shares overlapping clinical and histopathologic features with IgG4-RD, including IgG4+ plasmacytosis in affected tissue. Diagnosis requires interdisciplinary collaboration and careful examination of a patient's presenting symptoms, organ involvement, immunologic labs and histopathologic examination.