A patient with chronic bilateral periauricular swelling

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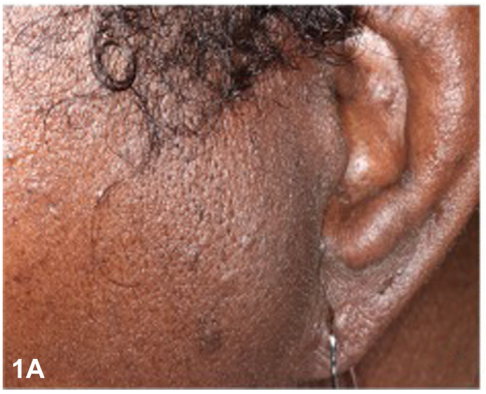
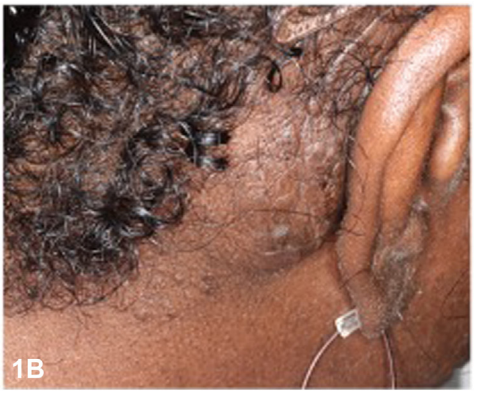
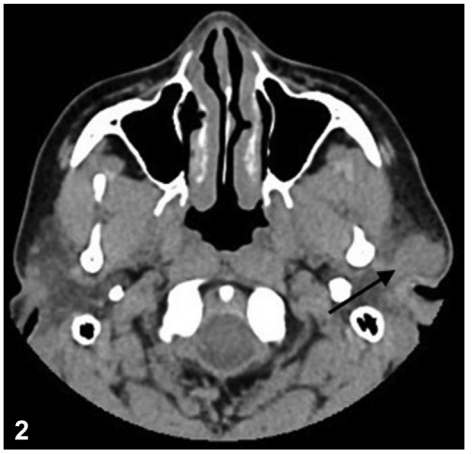
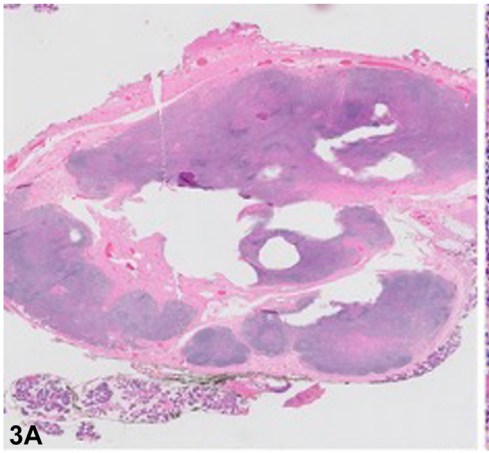
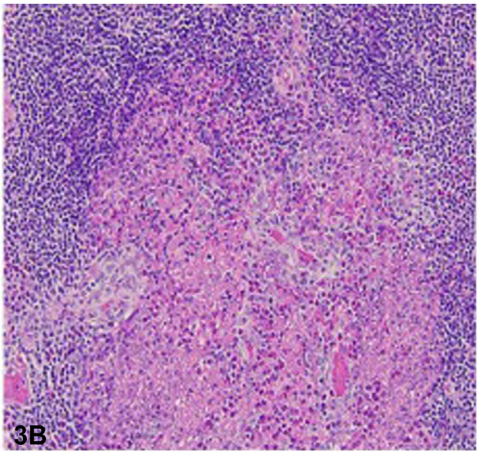
Key words: clinicopathological correlation; Kimura disease; peripheral eosinophilia.











A 49-year-old female with a history of atopic dermatitis and asthma was referred to our dermatology clinic for chronic pruritic swelling of both sides of her face. Review of symptoms was negative for fever, weight changes, and night sweats. Physical examination revealed ill-defined, nontender, subcutaneous bilateral periauricular swelling (Fig 1, *A* and *B*). Laboratory testing revealed high immunoglobulin E (Ig E), 5724 IU/mL. Computed tomography of the head and neck showed bilateral enlarged lymph nodes in the preauricular and parotid spaces (Fig 2). Excisional biopsy of the left intra-parotid lymph node confirmed the suspected diagnosis (Fig 3, *A* and *B*). Auramine rhodamine, Giemsa, and Grocott's methenamine silver staining were negative for organisms.

Question 1: What is the most likely diagnosis?

A. Kimura disease

B. Scrofuloderma

C. Rosai-Dorfman disease

D. Hodgkin lymphoma

E. Autosomal dominant hyper IgE syndrome

Answers:

A. Kimura disease — Correct. Kimura disease is a rare chronic inflammatory disorder of unknown etiology that often presents with painless lymph nodes or subcutaneous lymphoid masses in the head and neck region. The disease occurs predominantly in Asian individuals, and men are more commonly affected than women. Elevated serum IgE level and peripheral eosinophilia are helpful clues.[1]

B. Scrofuloderma — Incorrect. Scrofuloderma results from involvement of the skin overlying a contiguous focus of tuberculosis, usually in lymph nodes or bones. It classically presents as unilateral suppurative nodule overlying a cervical lymph node with fistula and sinus tract formation.

C. Rosai-Dorfman disease — Incorrect. Rosai-Dorfman disease is a multisystem histiocytic proliferative disorder of adolescence wherein patients often present with painless bilateral cervical lymphadenopathy, low grade fever, weight loss, polyclonal gammopathy, and internal organ involvement. Cutaneous lesions can be seen in 10% of the patients and are often nonspecific, but peripheral eosinophilia and elevated IgE levels would not be expected.[2]

D. Hodgkin lymphoma — Incorrect. Patients with Hodgkin lymphoma present with fever, weight loss, and generalized lymphadenopathy. Complete blood count will often show anemia and leukocytosis with or without peripheral eosinophilia.

E. Autosomal dominant hyper IgE syndrome — Incorrect. Autosomal dominant hyper IgE syndrome is a primary immunodeficiency characterized by a triad of severe atopic dermatitis, recurrent cold staphylococcal abscess, and sinopulmonary infections. Presentation occurs in early childhood and is often associated with high IgE levels. There are many associated genetic mutations with signal transducer and activator of transcription 3 (STAT3) being the most common.[3]

Question 2: Which of the following histopathological features are most likely to be seen in Kimura disease?

A. Dense lymphohistiocytic infiltrate with emperipolesis and sinusoidal expansion

B. Suppurative granulomatous dermatitis with caseous necrosis

C. Follicular lymphoid hyperplasia and dermal proliferation of blood vessels with dense lymphocytic infiltrate and eosinophilic microabscesses

D. Total effacement of nodal architecture by large hyperchromatic lymphocytes with Reed-Sternberg cells

E. Pauci-inflammatory non-caseating granulomas

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Funding sources: None.

IRB approval status: Not applicable.

Patient consent: Patient gave consent for their photographs and medical information to be published in print and online with the understanding that this information may be publicly available.

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JAAD Case Reports 2023;32:106-8.

2352-5126

https://doi.org/10.1016/j.jdcr.2022.12.004

Answers:

A. Dense lymphohistiocytic infiltrate with emperipolesis and sinusoidal expansion — Incorrect. Dilated sinuses with diffuse histiocytic infiltrate are frequently observed in Rosai-Dorfman disease with positive immunohistochemical staining for CD68 (cluster of differentiation) and S100 supporting the diagnosis. Emperipolesis, the presence of an intact cell within the cytoplasm of another cell, is a useful feature but not required for the diagnosis.[2]

B. Suppurative granulomatous dermatitis with caseous necrosis — Incorrect. Granulomatous dermatitis with caseous necrosis represents tuberculosis. Large abscesses with acid fast bacilli are frequently seen in patients with scrofuloderma.

C. Follicular lymphoid hyperplasia and dermal proliferation of blood vessels with dense lymphocytic infiltrate and eosinophilic microabscesses — Correct. Follicular lymphoid hyperplasia and proliferation of post capillary venules with stromal fibrosis and eosinophilic microabscesses are the characteristic histology features of Kimura disease.[1]

D. Total effacement of nodal architecture by large hyperchromatic lymphocytes with Reed-Sternberg cells — Incorrect. Nodular lymphoid aggregates composed of atypical lymphocytes with Reed-Sternberg cells are classic findings of Hodgkin lymphoma.

E. Pauci-inflammatory non-caseating granulomas — Incorrect. Nodular aggregates of epithelioid histiocytes with sparse lymphocytic infiltrate are typically seen in sarcoidosis.

Question 3: Which of the following is a promising long-term treatment option for this patient?

A. Corticosteroids

B. Cyclosporine

C. Surgery

E. Radiation therapy

F. Dupilumab

Answers:

A. Corticosteroids — Incorrect. Although most patients respond to oral corticosteroids, the recurrence rate is high once the treatment is discontinued, and there are various adverse effects associated with long-term therapy.[4]

B. Cyclosporine — Incorrect. Cyclosporine is a highly effective immunosuppressant medication, but long-term use is associated with renal toxicity and increased risk of malignant neoplasms.

C. Surgery — Incorrect. Although surgical excision can be performed for diagnostic and treatment purposes, it is associated with high morbidity and disease recurrence. Surgery followed by radiation therapy is associated with lower disease recurrence than either treatment alone and can be reserved for treatment-resistant cases.[4]

D. Radiation therapy — Incorrect. Radiotherapy was occasionally used in patients with treatment-resistant disease or who had recurrence after surgery. It is rarely used nowadays, given the long-term carcinogenesis risk and the availability of effective and safe long-term treatment options.

E. Dupilumab — Correct. Dupilumab is an interleukin 4 (IL-4) alpha antagonist that inhibits IL-4 and IL-13 signaling and has been shown to be an effective long-term treatment option in patients with Kimura disease. It would also be helpful for our patient's comorbid conditions, atopic dermatitis, and asthma.[5]

Conflicts of interest

None disclosed.

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