Kimura's Disease of the Earlobe

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Case Report

A 25 year-old woman of part-Hawaiian and part-Japanese ancestry presented with a 10-year history of a recurring mass in her left earlobe. The patient had both ears pierced ten years ago and within one year had developed an infection at the site of the left ear piercing. The earrings were removed, yet within another years time a tender erythematous mass developed in her left earlobe. The mass was diagnosed as a keloid and was injected with steroids six times without benefit. The mass was excised 4 times between 1992 and 1998 and recurred after each excision. The patient presented to us in 2002 with a mass in her left earlobe that had been increasing in size for the past 4 years.

Dermatologic examination revealed a firm and tender 4x4x2 cm lobular, erythematous lesion in the left earlobe. Further physical exam was unremarkable, including the absence of lymphadenopathy. Laboratory examinations were later performed and were entirely negative, including normal renal function and urine analysis, eosinophil count, ESR, and serum IgE levels. The patient has a past medical history of hypothyroidism treated with levothyroxine. Family history is non-contributory. We treated the patient with 2 injections of depo-Medrol 4 weeks apart prior to excision. During the first 12 weeks of follow-up care, the patient received 4 injections of depo-Medrol due to the continual recurrence of the mass. Noting the lack of improvement and resurgence of the mass following each post-excisional injection, the patient was referred to a rheumatologist and placed on prednisone at 20 mg once a day for 3 weeks with no signs of improvement. Thereafter, the patient was placed on cyclosporine, 100 mg three times a day with very rapid response and total resolution of the recurring mass on the left earlobe. The patient failed to fill the prescription for cyclosporine and during the 3 days lacking medication noted a resurgence of the mass. Upon resuming the cyclosporine at the aforementioned dosage, the mass resolved. Histopathologic examination of the excised mass revealed reactive lymphoid hyperplasia with scattered follicles showing prominent germinal centers and foci of vascular proliferation. A careful review of the patient's old chart revealed a pathology report from a previous excision in1998 that showed a nodular lesion with dermal and soft tissue infiltration of numerous large germinal centers and a polymorphous infiltrate of lymphoid cells, eosinophils, and small blood vessels located between the germinal centers. The clinical history and pathological findings of this patient are consistent with Kimura's disease.

Figure 1.— Firm 4x4x2 cm lobular erythematous lesion in the left earlobe.

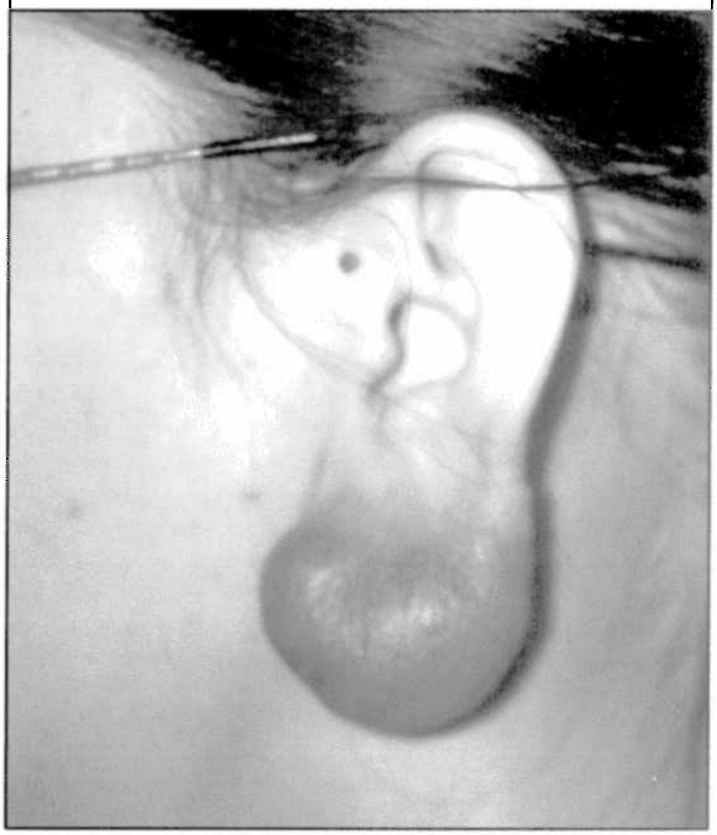

Figure 2.— Reactive lymphoid hyperplasia. Follicles with prominent germinal centers and foci of vascular proliferation.

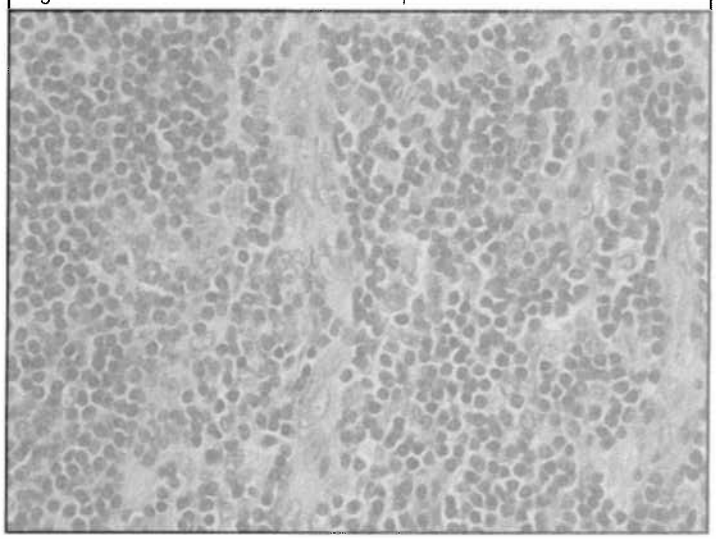

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Figure 3.— Low power micrograph showing dermal infiltration by a large germinal center in the middle and part of another germinal center in the right lower corner.

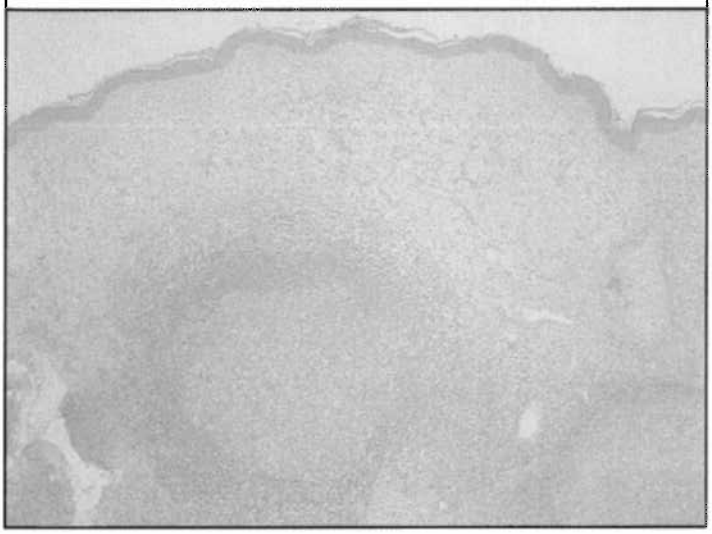
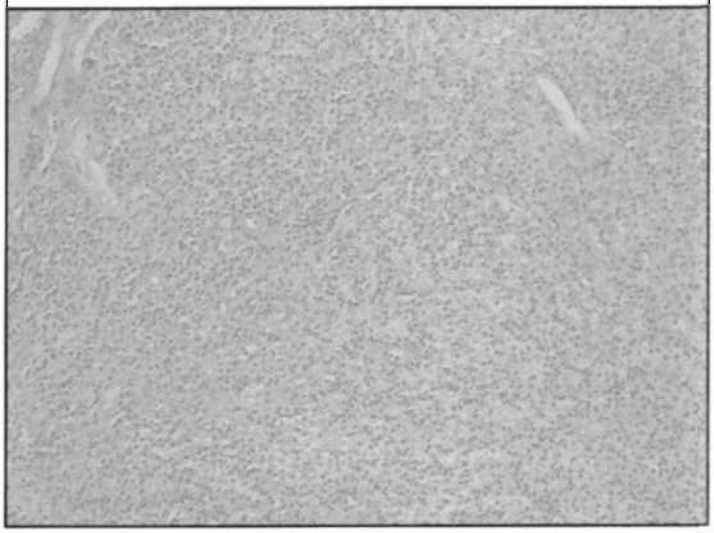


Figure 4.— A high power view of one of the germinal centers. A polymorphous infiltrate of lymphoid cell and the characteristic eosinophils and small blood vessels can be seen.



Discussion

Kimura's disease is a chronic inflammatory skin disease that usually occurs in the head and neck.[1] It usually presents with a cluster of small, painless, translucent nodules. While subcutaneous tissue and lymph nodes of the head and neck are most commonly affected, less frequent sites include the oral mucosa, salivary glands, prostate, kidney, peripheral nerves, tympanic membrane, and skeletal muscle.[2] Kimura's disease is a rare condition, with fewer than 120 cases reported worldwide in 1997.[3] There is a predisposition for Asian males, with a median age of onset of 26 years-old and a male:female ratio >3:1.[4] Kimura's disease is always benign but may mimic a neoplastic process at presentation.

Laboratory findings associated with Kimura's disease include peripheral eosinophilia and increased serum IgE.[5] CT and MRI imaging may be helpful if lymph node involvement is suspected. Renal disease may be present in 12% of these patients, characterized by proteinuria.[5]

Kimura's disease was first described in China by Kim and Szeto in 1937 as an angiolymphoid proliferative disorder of soft tissue.[6] Kimura et al. further described the condition in 1948 as an "unusual granulation combined with hyperplastic changes in lymphoid tissue."[1] The most prominent histological finding in Kimura's disease is an eosinophilic infiltration of germinal centers which results in folliculolysis. Lymphoid follicles with active germinal centers are an invariable finding. The blood vessels seen in Kimura's disease are thin-walled. These histopathologic findings can make the diagnosis of Kimura's disease when in cases where it can be difficult to do so clinically.

The differential diagnosis includes lymphoma, an infectious processes such as toxoplasmosis or tuberculosis,[5] and keloid formation. Just recently, Kimura's disease has been found to be distinct from angiolymphoid hyperplasia with eosinophils (ALHE).[7] Both of these diseases share a male predominance, predilection for the head and neck, tendency to recur, and lymphoid and eosinophilic infiltration. ALHE is more commonly found in older patients manifesting as multiple small dermal eruptions. The most important morphologic difference between Kimura's disease and ALHE lies in the nature of the blood vessels. ALHE has thick-walled blood vessels lined by plump eosinophilic endothelial cells that may contain vacuolated cytoplasm. These cells are not found in Kimura's disease.

Conclusion

The etiology of Kimura's disease is still uncertain. The most common theory is a hypersensitivity reaction mediated by T helper-2 cells.[8] Hypotheses in the past included antigenic stimulation from an insect bite, parasitic infections, and neoplastic changes.[2]

Kimura's disease may resolve spontaneously but has a tendency to recur. Treatments in literature have included surgical excision, systemic steroids, and radiotherapy. Our patients favorable response to cyclosporine given at a dose of 100 mg three times a day supports a recent case report of similar treatment of Kimura's disease.[9] The duration of treatment is, however, as of yet unknown. We plan to continue this regimen for one year, notwithstanding any unfavorable side effects.

Authors

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