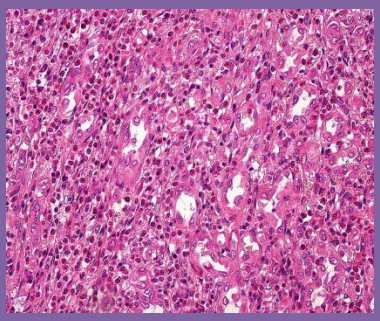
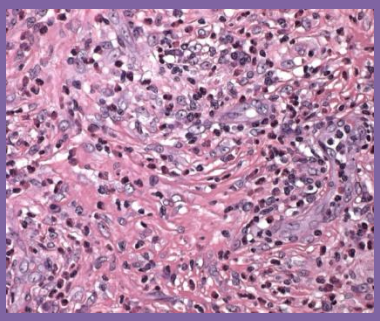
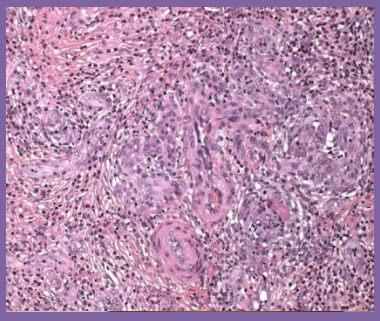
Case Presentation 3

JUVENILE TEMPORAL ARTERITIS: A CASE REPORT







This educational series for physicians is presented by the Weill Cornell Comprehensive Dermatopathology Service

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Case Contributor: Dr. Henry Spinelli

Clinical History

A 36-year old Jamaican female complained of a 1 cm enlarging, painful nodule on the left forehead. There was no history of trauma.(Figure 1) Shortly after the initial appearance of the nodule, the patient developed headaches, involving the top of the head, bilaterally, which occurred several times a week over a period of 6 months. The patient also felt unwell and fatigued. Past medical history was remarkable for seasonal allergic rhinitis and conjunctivitis. The patient was referred to Dr. Spinelli, who excised the nodule. Histologic examination revealed temporal arteritis with intimal hyperplasia, hemorrhage, inflammation and disruption of the internal elastic lamina. (Figure 2a-c) There was prominent perivascular lymphoid hyperplasia with eosinophilia, which also involved the adjacent skeletal muscle. (Figure 3) Vascular proliferation was present in the periarterial soft tissues, as well as within the temporal artery wall. The endothelial cells showed hobnailing, vacuolation of the cytoplasm and rare mitotic figures. (Figure 4) A diagnosis of juvenile temporal artertitis with features of Kimura's disease/angiolymphoid hyperplasia with eosinophilia (ALHE) was made.

The patient's headaches and malaise persisted after the removal of the nodule. At 1 month follow-up, she is without recurrence or additional symptoms.

Discussion

Vasculitis of the temporal arteries is extremely rare in patients under 50 years of age. By far the most common cause of temporal arteritis in the general population is giant cell arteritis. This vasculitis is one affecting the large and medium-sized muscular arteries and occurs predominantly in patients over 50. Other vasculitic processes may also involve the temporal artery, but are much less frequent.

Vasculitis of the temporal arteries is uncommon in patients under 50, and only approximately 40 cases have been reported in the English literature. Many of these cases are in the context of involvement of the temporal artery by a systemic vasculitis such as giant cell arteritis, polyarteritis nodosa, or Churg Strauss syndrome. In 1975, Lei et. al. described 4 cases of "juvenile temporal arteritis"(JTA), characterized by painless nodules which showed non-giant-cell inflammation of the temporal arteries. Thus far, approximately 20 cases consistent with this entity have been reported. Characteristic clinical features include age younger than 40 (although several authors have reported similar histopathologic findings in patients as old as 81), a palpable nodule (painful or painless) or swelling in the temporal area, and no systemic stigmata of vasculitis (including generalized inflammatory reaction or cranial ischemic manifestations). Peripheral blood eosinophilia is sometimes present. Histopathologic examination characteristically shows intimal proliferation, disruption of the media and extensive inflammatory infiltrate consisting of lymphocytes, eosinophils and plasma cells. Often, endothelial proliferation is present.

While some authors consider JTE a distinct disease, others believe that it is in the spectrum of Kimura's disease or ALHE. Kimura's disease and ALHE are conditions that involve the head and neck region, tend to recur despite treatment and share several histopathologic features, such as lymphoeosinophilic infiltrates of the involved tissue and vascular proliferation. In addition to direct lesional morbidity, nephrotic syndrome and T-cell dyscrasia and lymphoma can occur in the setting of both diseases. Although there have been several reports of both entities occurring in a single patient, most authors view ALHE and Kimura's disease as two distinct disorders.

Angiolymphoid hyperplasia with eosinophilia occurs in young to middle aged females, and presents with multiple small (less than 2 cm) erythematous dermal papules or nodules. The lesions are commonly superficial, localizing to the dermis or subcutis. Eosinophilia occurs in approximately 20% of patients, but other systemic manifestations are absent. Histologic examination shows a diffuse lymphoid infiltrate of diverse intensity, with a variable eosinophilic component. The vascular proliferation in ALHE is characterized by thick-walled blood vessels lined by epithelioid, hypertrophic endothelial cells with vacuolated cytoplasm and vesicular nuclei, which protrude into and sometimes occlude the lumen.

In contrast, Kimura's disease occurs mainly in young Asian males and presents with large (greater than 2 cm) masses. The lesions are deep, involving subcutaneous tissue, muscle, and salivary gland. There

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are frequent systemic manifestations, such as lymph node enlargement, eosinophilia, and elevated serum IgE. Histopathologically, Kimura's disease shows lymphoid follicle and germinal center formation with massive eosinophilic infiltrates. Vascular proliferation may be present, but the characteristic cytomorphologic alterations of the endothelium seen in ALHE are not identified.

The relationship between JTA and Kimura's disease/ALHE has not been fully elucidated. At short follow-up (all under 2 years), most cases of JTA are limited to a single lesion and neither recur nor produce subsequent systemic symptoms. This leads some authors to conclude that it is a localized, self-limited disease cured by excision. However, it is currently unclear what the long-term sequelae of the disease may be. Although four cases of JTA have shown no evidence of endothelial proliferation, most cases of JTA share histopathologic features of Kimura's disease or ALHE. There is a report of an 81-year old patient with a lesion histopathologically compatible with JTA and occurrence of ALHE in the periauricular soft tissue three years later. Furthermore, several cases with histopathology of JTA have been reported in conjunction with systemic abnormalities, such as nephrotic syndrome, eosinophilia of 31% with multiarterial involvement, and past history of lymphoma. Thus, whether juvenile temporal arteritis is an independent entity or a manifestation of Kimura's disease or ALHE involving the temporal artery is currently controversial.

Our patient presented with a painful lateral forehead nodule that had histologic findings of arteritis involving the temporal artery and headaches. As our patient's headache was generalized rather than localized, it is not suggestive of giant cell arteritis and no multinucleated cells were identified in the biopsy. The clinical presentation and histopathology of the lesion, including intimal hyperplasia, disruption of the internal elastic lamina and the prominent lymphoeosinophilic intramural and periarterial infiltrate with endothelial proliferation are diagnostic of juvenile temporal arteritis. The extent of the lymphoeosinophilic hyperplasia in the perivascular soft tissue and muscle, as well as the prominent vascular proliferation with epithelioid changes and vacuolation leads us to conclude that the lesion has features of Kimura's disease/ALHE.

Overall, there is currently no agreement as to whether juvenile temporal arteritis represents an independent entity or a manifestation of Kimura's disease/ALHE. In light of this, we believe that until more cases of this disease are reported with long-term follow-up, it should not be assumed that excision will necessarily be curative for every patient. A thorough initial work-up, close clinical follow-up and periodic laboratory studies to monitor for systemic complications may be advisable.

Table 1: CRITERIA FOR DIAGNOSIS OF GIANT CELL ARTERITIS.

-Age greater than or equal to 50 years at disease onset
-New onset of localized headache
-New onset of localized headache
-Temporal artery tenderness or decreased temporal artery pulse
- Elevated erythrocyte sedimentation rate greater than or equal to 50 mm/hour
- Biopsy sample showing necrotizing arteritis, characterized by a predominance of mononuclear cell infiltrates or a granulomatous process with multinucleated giant cells.

Reproduced from.Arthritis Rheum.1990 Aug;33(8):1122-8.

Case References

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6. Wang YH, Yin HF. One patient with Kimura's disease and angiolymphoid hyperplasia with eosinophilia also suffers from kidney injury. Beijing Da Xue Xue Bao. 2008 Aug 18;40(4):405-7.

Figure Legend

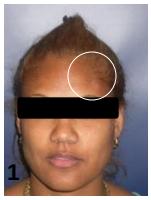

Figure 1 : Patient developed a 1 cm painful, tender nodule on left lateral forehead.

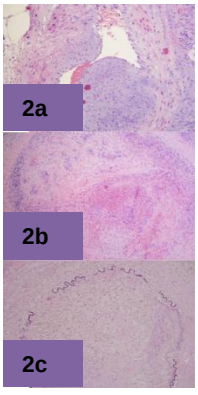

Figure 2a. 2b : The biopsy shows arteritis with intimal hypertrophy.

Figure 2c : Elastic stain highlights disruption of the internal elastic lamina.

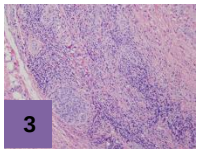

Figure 3: Extensive inflammatory infiltrate extends into the periarterial soft tissue.

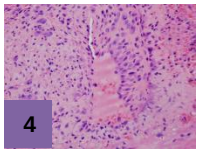

Figure 4: Hobnailing and vacuolation of the endothelial cells is present.

Under the direction of Dr. Cynthia M. Magro, the Weill Cornell Comprehensive Dermatopathology Service is a leading edge consultation service and CAP-accredited laboratory for dermatologists, plastic and general surgeons and other dermatopathologists. Dr. Magro is an internationally renowned dermatopathologist, educator and author. She is a Professor of Pathology and Laboratory Medicine at the Weill Cornell Medical College in Manhattan, and is board certified in anatomic pathology, dermatopathology and cytopathology. Dr. Magro is an expert in the diagnosis of complex inflammatory skin diseases. Her areas of expertise include cutaneous manifestations of auto-immune disease, systemic viral disease and vasculitis, atypical drug reactions, benign, atypical and overtly malignant lymphocytic infiltrates of the skin, and diagnostically difficult melanocytic proliferations.The award-winning author of *The Melanocytic Proliferation: A Comprehensive Textbook of Pigmented Lesions*, Dr. Magro has recently completed her second book, *The Cutaneous Lymphoid Proliferation, a comprehensive textbook on benign and malignant lymphocytic infiltrates*. She has co-authored over 250 peer reviewed papers and several textbook chapters. Dr. Magro frequently presents courses on inflammatory skin pathology and difficult melanocytic proliferations to the American Academy of Dermatology, the United States and Canadian Academy of Pathology, and the American Society of Clinical Pathology. Dr Magro has consistently been recognized in Who's Who in America®, Castle Connolly's renowned America's Top Doctors – New York Metro Area® edition and in the Super Doctors® list published in The New York Times Magazine.

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