Juvenile temporal arteritis (JTA) is a rare and controversial diagnosis. The clinical features include: age younger than 40 years old, temporal nodule with or without pain, no systemic features and no recurrences or systemic symptoms upon follow up. The histopathologic features include intimal hyperplasia, endothelial hyperplasia, disruption of the internal elastic lamina and a lymphoepithelial panarteritis that lacks germinal centers and granulomatous inflammation. The differential diagnosis consists of angiolymphoid hyperplasia with eosinophilia (ALHE) and Kimura disease. It is debated whether JTA is a distinct diagnosis or a subset of ALHE or Kimura disease. We report a case of an 18-year-old man with a clinical presentation of temporal artery pseudoaneurysm, but with histopathologic features of JTA. The patient has not sought further treatment 2.5 years after the original excision.

Key Words: juvenile temporal arteritis, angiolymphoid hyperplasia with eosinophilia, Kimura disease, temporal artery, temporal artery pseudoaneurysm

INTRODUCTION

Temporal arteritis in patients younger than 50 years of age is rare. Less than 40 cases have been reported in the literature. Within this younger population the differential diagnosis of temporal arteritis is divided into three categories: juvenile temporal arteritis (JTA), temporal arteritis associated with systemic vasculitis (Churg-Strauss syndrome and polyarteritis nodosa) and giant cell arteritis.1

JTA was first described in 1975 in a case series by Lie.2 It remains a rare diagnosis with a total of less than 20 cases reported in the literature. JTA typically presents in patients who are less than 40 years of age as a temporal nodule that may be painful or painless, and can be unilateral or bilateral. Less commonly JTA can be associated with headaches (three prior cases) and antecedent trauma (one prior case).3 While peripheral eosinophilia may be present, commonly there are no systemic features and the sedimentation rate is within normal limits. Excision is the treatment of choice, and there have been no reports of recurrences or systemic symptoms at follow up.

The histopathologic features of JTA include intimal hyperplasia, endothelial hyperplasia, disruption of the internal elastic lamina and a panarteritis containing lymphocytes and eosinophils. Several, but not all, of the cases also describe one or more of the following: periarteritis consisting of lymphocytes and eosinophils, lymphoid aggregates without germinal centers, scattered multinucleated giant cells, thromb, vascular proliferation, fibrinoid necrosis and pseudoaneurysm.1,3,4,5

We add to the scant literature of this uncommon disease with a case report of an 18-year-old man with a clinical diagnosis of temporal artery pseudoaneurysm that upon histopathologic exam had features of JTA.

CASE REPORT

The patient was an 18-year-old man with a recent history of trauma to the right temporal region, presenting with a pulsatile mass over the right temporal artery. Otherwise, he was in his usual state of health and did not have any systemic signs or symptoms. The lesion was clinically diagnosed as a superficial temporal artery pseudoaneurysm and excised.

The resected specimen consisted of a single piece of soft pink tissue that measured 1.3 x 0.8 x 0.8 cm and contained a dilated vessel with a wall thickness measuring up to 0.2 cm that is partially obstructed by soft tan tissue. Microscopic examination revealed a panarteritis that consisted of eosinophils and lymphocytes (Figure 1), intimal hyperplasia (Figure 1), extensive fibrous replacement of the media, disruption of the internal elastic lamina (Figure 2) and multifocal vascular proliferations lined by plump endothelial cells within the intima and the media (Figure 1). The surrounding soft tissue and skeletal muscle contained a lymphoepithelial infiltrate with lymphoid aggregates that lacked germinal centers, multinucleated giant cells and granulomatous inflammation (Figure 3 and Figure 4). In addition, the arterial lumen was partially obstructed by an organizing thrombus with intravascular papillary endothelial hyperplasia (Figure 5). These features were considered diagnostic for JTA, and follow up was recommended. The patient has not sought further treatment 2.5 years after the excision.
Figure 1. Intimal hyperplasia with a chronic inflammatory infiltrate consisting of eosinophils and lymphocytes and vascular proliferation lined by reactive endothelial cells (Hematoxylin & Eosin, 200x Magnification).

Figure 2. Disruption of the internal elastic lamina (Verhoeff-Van Gieson, 200x magnification).

Figure 3. Skeletal muscle involved with associated inflammation (Hematoxylin & Eosin, 200x Magnification).

Figure 4. Lymphoid aggregate without germinal center (Hematoxylin & Eosin, 200x Magnification).

Figure 5. Organizing thrombus with intravascular papillary endothelial hyperplasia (Hematoxylin & Eosin, 200x Magnification).
Table 1. Differential Diagnosis of Juvenile Temporal Arteritis.*

<table>
<thead>
<tr>
<th>Clinical Presentation</th>
<th>Juvenile Temporal Arteritis</th>
<th>Angiolympoid Hyperplasia with Eosinophilia</th>
<th>Kimura Disease</th>
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<tbody>
<tr>
<td>Age younger than 40 years old. Temporal nodules that are painful or painless, and can be unilateral or bilateral.</td>
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<td>Young to middle aged women. Single or multiple erythematous papules or nodules (&lt;2 cm) on the face or neck. Pain and pruritus.</td>
<td>Young Asian men. Pre-auricular and submandibular regions. Large nodules (&gt;2 cm) that involve the deep subcutaneous tissue/skeletal muscle, lymph nodes or salivary glands.</td>
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Systemic Signs and Symptoms

Typically none. Can have peripheral eosinophilia. | Peripheral eosinophilia, lymphadenopathy, T-cell lymphoma and renal disease. | Renal disease, T-cell lymphoma, lymphadenopathy, peripheral eosinophilia and elevated serum IgE levels. |

Arteritis

Common. Panarteritis contains lymphocytes and eosinophils. Also, intimal hyperplasia and disruption of the internal elastic lamina. | Less common. | Less common. |

Periarteritis

Less common. Periarteritis consists of lymphocytes, eosinophils and lymphoid aggregates without germinal centers. | Diffuse lymphocytic infiltrate with or without eosinophils. Secondary follicles may develop. Granulomatous inflammation is not typical. | Reactive lymphoid follicles with germinal centers surrounded by a dense infiltrate composed of eosinophils, lymphocytes and mast cells. |

Neovascularization

Less common and least prominent. | Prominent well-circumscribed vascular proliferation. Vessels are small caliber and lined by a single layer of bland plump endothelial cells. | Present, but it is less prominent than that found in ALHE. Lacks the reactive appearing endothelial cells. |

Treatment

Surgical excision | Surgical excision | Surgical excision |

Prognosis

No recurrence at follow up. | Recurrence common | Recurrence common |

*Brief Description of Table: This table compares and contrasts the features of JTA with the features of its main differential diagnosis, ALHE and Kimura disease.

DISCUSSION

JTA is a rare and controversial diagnosis. The main differential diagnosis consists of angiolymphoid hyperplasia with eosinophilia (ALHE) and Kimura disease.

ALHE usually presents as a single or multiple erythematous papules or nodules (< 2 cm) on the face or neck of young to middle aged women that are painful and pruritic. Systemic signs and symptoms reported in the literature include: peripheral eosinophilia, lymphadenopathy, renal disease and T-cell lymphoma. One study also demonstrated the same TCR gene rearrangement in both the T-cell lymphoma and the ALHE. Histologically ALHE is a dermal lesion that contains a diffuse lymphocytic infiltrate that can be accompanied by eosinophils, secondary follicle formation and rarely granulomatous inflammation. The vascular proliferation is prominent and consists of a well-circumscribed proliferation of small caliber vessels lined by a single layer of bland plump endothelial cells that can narrow the lumen of a nearby muscular artery via mass effect.
Kimura disease most commonly occurs in young Asian men as preauricular and submandibular large nodules (>2 cm) that involve the deep subcutaneous tissue/skeletal muscle, lymph nodes or salivary glands. These lesions are frequently accompanied by systemic findings such as nephrotic syndrome, T-cell lymphoma, lymphadenopathy, peripheral eosinophilia and elevated serum IgE levels. Histopathologically Kimura disease contains reactive lymphoid follicles with germinal centers surrounded by a dense infiltrate composed of eosinophils, lymphocytes and mast cells that can develop into an eosinophilic abscess. Vascular proliferation may be present, but is less prominent than the vascular proliferation of ALHE and lacks the reactive appearing endothelial cells.\(^1,5,9\)

The differences between JTA, Kimura disease and ALHE are subtle. (See Table 1) While a diagnosis can be made when they present with the classic clinical presentation and classical histopathologic findings, in a given case it may be impossible to differentiate these entities.\(^1\) General differences that aid in diagnosis include: The inflammation seen in ALHE and Kimura disease is usually found in the soft tissue surrounding muscular arteries, and rarely causes an arteritis.\(^2,11,12\) All three entities are associated with vascular proliferation, but the vascular proliferation is often more prominent in Kimura disease and ALHE than in JTA.\(^1\)

The similarities between these entities have sparked a debate as to whether JTA is a distinct diagnosis or whether all three entities are different manifestations of the same disease process.\(^1\)

The evidence supporting the theory that JTA, ALHE and Kimura disease are related consists of several case reports of JTA and Kimura disease in the same patient. One case was a 23-year-old man with an asymptomatic nodule of the left temple and peripheral eosinophilia. Histologic examination revealed JTA involving the left temporal artery. However, the surrounding soft tissue had a pattern consistent with Kimura disease. The patient was asymptomatic at follow up 8 months after surgical excision.\(^13\)

Evidence that supports the theory that JTA is an independent diagnosis consists of a case of JTA without endothelial proliferation.\(^1,3,5\) While vascular proliferation can occur in JTA, it is a common and prominent feature of ALHE and Kimura disease.\(^1,5\) The histologic sections in this case did reveal multifocal vascular proliferations lined by plump endothelial cells, however this finding did not rule out a diagnosis of JTA because the vascular proliferation was not a prominent feature and the overall clinical and histopathologic features were most consistent with JTA.

Whether JTA is an independent diagnosis or a facet of ALHE and Kimura disease is significant because it has prognostic implications. JTA does not recur after local excision and is not associated with serious systemic symptoms. In contrast, ALHE and Kimura disease have a tendency to recur after local excision and have been associated with serious systemic symptoms such as nephrotic syndrome and T-cell lymphoma.\(^5\)

Clinically this patient was thought to have a superficial temporal artery pseudoaneurysm. This diagnosis fit with the clinical presentation of prior trauma and a painless pulsatile mass in the temporal region.\(^15\) While JTA is not traditionally associated with trauma, there is one prior documented case of JTA status post a history of trauma, and three documented cases of JTA associated with a pseudoaneurysm.\(^1\) Thus, trauma may be a cause of JTA. However, an alternate theory is that the trauma did not directly cause JTA, but only brought the lesion to the attention of the patient.

The diagnosis of JTA was made after histopathologic examination. Features that led to this diagnosis include intimal hyperplasia, disruption of the internal elastic lamina, focal vascular proliferation lined by reactive endothelial cells, panarteritis with a chronic inflammatory infiltrate composed of lymphocytes and eosinophils, lymphoid aggregates without germinal centers and a lack of multinucleated giant cells and granulomatous inflammation. In addition, a chronic inflammatory infiltrate composed of lymphocytes and eosinophils did involve the surrounding soft tissue. This feature is suggestive of Kimura disease, but also has been reported in prior cases of JTA. Thus, the clinical presentation and the overall histopathologic picture were more consistent with JTA.\(^13\)

All prior cases of JTA reported in the literature have follow up times of less than two years post diagnosis, and most cases have follow up at less than one year.\(^5\) This short time interval makes it difficult to discern the natural history of JTA. This case has a slightly longer follow up time of 2.5 years, at which the patient has not sought further medical therapy and is presumed to be asymptomatic and without recurrence.

This case is an example of a lesion with the typical clinical presentation, histopathologic features and natural history of JTA that does not have significant overlap with ALHE and Kimura disease. Thus, this case supports the theory that JTA is an independent entity.

CONFLICT OF INTEREST

None.

REFERENCES


