Bilateral Involvement of Juvenile Temporal Arteritis Associated with Kimura Disease

Na Ri Kim¹, Churl Hyun Im¹, Jong Wan Kang¹, Ji Hun Kim², Tae-In Park³, Han-Ik Bae³, Eon Jeong Nam¹, Young Mo Kang¹

¹Division of Rheumatology, Department of Internal Medicine, Kyungpook National University School of Medicine, Daegu, ²Division of Rheumatology, Department of Internal Medicine, Andong Medical Group Hospital, Andong, ³Department of Pathology, Kyungpook National University School of Medicine, Daegu, Korea

Juvenile temporal arteritis (JTA) is a localized nodular arteritis confined to the temporal artery without evidence of systemic inflammation, and it occurs mainly in patients younger than 50 years. From the first case report, the pathological features of JTA have been suspected to be the morphological equivalent of Kimura disease (KD), which has been supported further by the concurrent cases of JTA with KD. We present the first case of bilateral JTA accompanying KD, which was confirmed by histological and ultrasound evaluations and supports the hypothesis that JTA is a manifestation of KD. The un-excised JTA lesion was resolved completely after corticosteroid therapy with no recurrence. (J Rheum Dis 2018;25:65-68)

Key Words. Juvenile temporal arteritis, Kimura disease, Corticosteroids

INTRODUCTION

Juvenile temporal arteritis (JTA) is a localized nodular arteritis confined to the temporal artery, without evidence of systemic inflammation, and it occurs mainly in patients younger than 50 years [1]. Histologic examination of the JTA shows intimal proliferation and panarteritis with lymphocyte and eosinophil infiltrates but no giant cell, which may extend to the perivascular area [1]. From the first case report [1], the pathologic features of JTA have been suspected to be the morphological equivalent of Kimura disease (KD), which has been supported further by the concurrent cases of JTA with KD [2-5]. KD is a chronic inflammatory disorder that affects the subcutaneous tissue and lymph nodes, especially in the head and neck area, with peripheral blood eosinophilia [6]. Pathological study of the specimens from KD showed a marked reactive follicular hyperplasia with prominent follicles that were surrounded with a large number of eosinophils, lymphocytes and mast cells [6]. Fibrosis and vascular proliferation were also seen in KD [6]. Determination of the association between JTA and KD, that is, whether JTA is a manifestation of KD or whether these two diseases occur coincidentally, is important not only for understanding of the disease pathogenesis, but for the clinical decision of management. We present the first case of bilateral JTA accompanying KD, which showed a good response to corticosteroid therapy.

CASE REPORT

A 37 year old Asian Korean man visited the hospital for painless nodular lesions on bilateral temporal areas. Two years earlier, right inguinal and left antecubital masses developed with no previous history of either local infection or trauma. A few months later, a nodular lesion of the size of a corn, without cutaneous irritation symptoms, developed on the left anterior temporal area (Figure 1A). He did not have constitutional symptoms, except for occasional headache. Two weeks before hospi-
On physical examination, both lesions were firm, painless, and pulsatile on both temporal arteries. Two enlarged painless lymph nodes were palpated on the right inguinal and left antecubital areas. There was no evidence of other organ involvement. Laboratory tests showed no abnormalities except for peripheral blood eosinophilia (15.4%) and markedly elevated serum immunoglobulin E level (5,111 IU/mL). We identified the enhanced focal wall thickening at the left superficial temporal artery on computed tomographic angiography (Figure 1C). Ultrasonography with Doppler images of the lesional superficial temporal artery (Figure 1D and 1E) showed diffuse edematous wall thickening (i.e., the “halo” sign), vascular tortuosity, and irregular luminal stenosis with turbulent flow.

Histological features of both left antecubital and right inguinal lymph nodes included exuberant follicular formation and angiogenesis, with prominent eosinophilic infiltration, which were typical KD histology (Figure 2). On excisional biopsy of the nodular lesion on the left superficial temporal artery, the vascular lumen was compacted by the hyperplastic endothelium (CD34 positive), with canalicular tract formation. Disruption of internal elastic lamina and adventitial inflammation with eosinophilic infiltrates were found on the vessel wall (Figure 3). However, there were no epithelioid endothelial cells or giant cells within the lesions.

The patient was diagnosed as JTA on bilateral superficial temporal arteries, accompanying KD. The patient received treatment with corticosteroid of 0.5 mg/kg per day, followed by azathioprine therapy (1.5 mg/kg) with tapering of corticosteroid. The nodular lesion on the right superficial temporal artery disappeared 5 months after treatment. The patient had received maintenance therapy for a year. There was no recurrence of either JTA or KD during 7 years of follow-up.

**DISCUSSION**

The clinical and pathological presentation of this case was compatible with those of JTA with bilateral involvement, accompanying KD lesions. Among 21 cases identi-
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Figure 2. Inguinal and antecubital lymph nodes showed markedly increased lymphoid follicles with germinal centers and exuberant eosinophilic infiltrations, which is compatible with Kimura disease (H&E, A: ×40; B: ×200; C: ×400).

Figure 3. Biopsy of the superficial temporal artery lesions (H&E, A: ×40; B: ×100) showed intravascular proliferating structures, with eosinophilic infiltrations covered by endothelial linings (C: CD 34, ×200) and intramural and perivascular inflammatory lesions.

ified in a PubMed search for JTA [1,2,5-7,12] and the present case, most of the JTA patients were younger than 40 years old, except for two cases. Males were predominantly affected with JTA than female (15 males in 21 cases). Most JTA lesions were painless mass at the temporal area without systemic symptoms, except for headache and fatigue. These patients did not show any internal organ involvement and systemic inflammatory reaction. Histologically, JTA showed endothelial proliferation with luminal narrowing, disruption of the internal elastic lamina, and panarteritis with lymphoid follicle formation in the periarterial tissues [1]. KD showed similar pathologic features with JTA, including lymphatic eosinophilic infiltration, vascular proliferation, and multiple follicle formation, except for differences in the distribution of the involved tissues. Previous case reports of JTA accompanying KD, in which JTA either preceded [2] or developed at the same time with KD [3-5]. One case was old age JTA, and KD developed as retroauricular mass in 3 years later after JTA excision [2]. The other three cases were JTA, which co-existed with neighboring KD lesions at the concurrent periarterial specimen [3-5]. These features suggested that JTA is a manifestation of KD, which is further supported by the present case.

In contrast to classic temporal arteritis, JTA usually involves unilateral side. Only four case reports of bilateral involvement of the JTA have been identified up to now [9-12]. Common characteristics of bilateral cases and our case included occurrence at age between 20 and 40 years, male gender, and peripheral blood eosinophilia. The histological features and clinical course were not different from those with unilateral involvement. To the best of our knowledge, this is the first case of bilateral JTA associated with KD. Whether JTA is an independent entity or a presentation of KD or angiolymphoid hyperplasia with eosinophilia (ALHE) is difficult to define, until now, histologic findings have been the key to differentiate these entities. Compared to histologic findings of JTA or KD, ALHE showed vascular proliferation by thick-walled blood vessels lined by epithelioid, hypertrophic endothelial cells with vacuolated cytoplasm and vesicular nuclei [7].

The Doppler ultrasound may be a useful imaging tool for JTA. In the Doppler sonographic study of ALHE involving the temporal artery, characteristic findings included the
“halo” sign reflecting vascular wall edema around the involved artery and the sinusoidal blood flow in the lumen [13]. Although the halo sign was suggested as a specific marker for classic temporal arteritis in a recent meta-analysis [14], it should be interpreted in the context of specific clinical settings, such as JTA and ALHE.

In the previously reported JTA lesions, surgical excision was both diagnostic and curative management. The un-excised JTA lesion in the present case, however, was resolved completely after moderate dose corticosteroid therapy, without recurrence. Considering the hypothesis that JTA is a manifestation of KD and the recommended therapeutic options for multiple or recurred KD include corticosteroid and immune-suppressant therapy, medical treatment may be effective for JTA patients with accompanying KD [6]. Furthermore, in patients of JTA with typical Doppler ultrasound findings, such as the halo sign, medical treatment with corticosteroid may be considered as the first line treatment and the surgical option may be considered only when the disease is refractory to medical therapy. However, surgery may be the first choice for the treatment of single and localized JTA or KD [1,15].

**Ethical considerations**

The study was approved by the Institutional Review Board (IRB) of the Kyungpook National University Hospital (IRB no. 2011-06-001).

**SUMMARY**

There is currently no agreement as to whether JTA represents an independent entity or a manifestation of KD, with no established long-term treatment. This is the first case of a bilateral involvement of JTA accompanying KD, which was confirmed by histological and ultrasound evaluations and supports that JTA is a manifestation of KD. The un-excised JTA lesion was resolved completely after corticosteroid therapy. Medical treatment with corticosteroid may be considered as the first line treatment while surgery can be considered for treatment of focal lesions or refractory cases to medical therapy.

**CONFLICT OF INTEREST**

No potential conflict of interest relevant to this article was reported.

**REFERENCES**