Juvenile temporal arteritis

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Abstract
Juvenile temporal arteritis (JTA) is an extremely uncommon vasculitis of unknown etiology (eleven documented cases have been reported in the literature) affecting older children and young adults. In contrast to the classic form of temporal arteritis, it is not a systemic disease nor does it cause local symptoms at the temporal area. JTA has a benign clinical course and it is curable by surgical excision. The term JTA was coined by Lie and his colleagues, in 1975, when they reported four cases of an otherwise asymptomatic disease presenting with a painless nodule at the temporal region in older children and young adults. No one case had evidence of systemic disease or history of trauma to the temporal region, and the excisional biopsy of the lesions had revealed a non-giant cell granulomatous inflammation of the temporal arteries with eosinophilic infiltration, intimal proliferation and microaneurysmal disruption of the media. JTA is treated by surgical excision and does not recur.

Keywords
Juvenile temporal arteritis – Temporal arteritis – Kimura’s disease – Angiolymphoid hyperplasia with eosinophilia - Thromboangiitis obliterans with eosinophilia

Disease name / synonyms
Disease: Juvenile temporal arteritis
Synonym: Non-giant cell granulomatous temporal arteritis with eosinophilia.

Definition
JTA is a nongranulomatous nonnecrotizing eosinophilic vasculitis, presenting as painless temporal nodule in older children and young adults; it is completely unrelated to the classic giant cell temporal arteritis (Andonopoulos AP et al., 2004; Bollinger A et al., 1986; Fujimoto M et al., 1996; Lie JT et al., 1975; Lie JT, 1995; Tomlinson FH et al., 1994; Wakugawa M et al., 1999; Watanabe C et al., 2002).

Histopathology
Histologically, involved temporal artery shows remarkable thickening of the wall with constriction or obliteration of the lumen. There is panarteritis, with or without fibrinoid necrosis, characterized by significant intimal proliferation and dense, intravascular and perivascular, infiltration by numerous eosinophils, lymphocytes and plasma cells. Giant cells are not seen. Lymphoid follicles and newly formed capillaries surround the vessel. Elastica van Gieson staining may reveal focal disruption of the internal elastic lamina.

Diagnostic criteria of JTA
1- Occurrence in children and young adults;
2. Absence of associated features such as myalgia, visual disturbance, fever, anemia;
3. Manifested as painless temporal nodule;
4. Normal erythrocyte sedimentation rate;
5. Eosinophilic panarteritis and thrombosis with or without microaneurysmal disruption of the artery;
6. Intimal proliferation, disruption of the media, and extensive infiltrate consisting of lymphocytes, eosinophils and plasma cells;

Differential diagnosis
JTA should be differentiated from conditions that may occur in the temporal regions and/or be associated with non-giant cell eosinophilic temporal arteritis.

**Kimura’s disease**
It is an inflammatory disorder of unknown etiology seen in an endemic form in Orient. It usually presents as a subcutaneous mass of the head and neck region, which may persist or recur, and is associated with peripheral eosinophilia and elevated IgE. Histopathologically, early lesions show a prominent vascular proliferation and dense lymphocytic infiltration with numerous eosinophils, while late lesions reveal a nodular lymphoid hyperplasia and fibrosis (Kung IT et al., 1984). Though, it is not known whether Kimura’s disease involves a specific artery, the vascular changes of this condition may resemble those of JTA. The report of Watanabe et al. describes an elderly patient in whom JTA preceded a typical manifestation of Kimura’s disease. In this report, the authors suggest that JTA may be an accessory lesion of Kimura’s disease (Watanabe C et al., 2002).

**Angiolymphoid hyperplasia with eosinophilia (ALHE)**
It is a dermal and subcutaneous lesion occurring more frequently in the head and neck. Histopathologically, the lesion is characterized by prominent vascular proliferation with histiocyte-like endothelial cells, and a nodular and diffuse lymphocytic infiltrate with eosinophils (Olsen TG et al., 1985).

**Thromboangiitis obliterans with eosinophilia (Buerger’s disease) of the temporal arteries**
It is an unusual form of temporal arteritis in young cigarette smokers, unassociated with vascular involvement of either upper or lower extremities. This variant of Buerger’s disease mimics clinically the classic temporal arteritis and histopathologically Kimura’s disease and JTA (Lie JT et al.1988).

**Etiology**
The etiology of JTA remains unknown.

**Clinical description**
Clinically, JTA manifests as asymptomatic temporal nodule and affects older children and young adults (Andonopoulos AP et al., 2004; Bollinger A et al., 1986; Fujimoto M et al., 1996; Lie JT et al., 1975; Lie JT, 1995; Wakugawa M et al., 1999; Watanabe C et al., 2002). Bilateral JTA has been reported in one case (Lie JT, 1995). The systemic symptoms that occur in the classic temporal arteritis such as headache, myalgia, visual disturbance and fever are absent. The hematologic tests and the erythrocyte sedimentation rate are usually normal (Andonopoulos AP et al., 2004; Bollinger A et al., 1986; Fujimoto M et al., 1996; Lie JT et al., 1975; Lie JT, 1995; Tomlinson FH et al., 1994; Wakugawa M et al., 1999; Watanabe C et al., 2002). Peripheral blood eosinophilia may be present (Bollinger A et al., 1986; Fujimoto M et al., 1996; Watanabe C et al., 2002).

**Diagnostic methods**
Lesional biopsy.

**Epidemiology**
JTA is rare. Fewer than 20 cases have been reported in the literature (Andonopoulos AP et al., 2004; Bollinger A et al., 1986; Fujimoto M et al., 1996; Lie JT et al., 1975; Lie JT, 1995; Tomlinson FH et al., 1994; Wakugawa M et al., 1999; Watanabe C et al., 2002).

**Management (treatment)**
JTA is treated by surgical excision and does not recur. (Lie JT et al., 1975; Lie JT, 1995).

**Unresolved questions**
Is JTA an expression of Kimura’s disease?

**References**

**Melachrinou M and Andonopoulos AP. Juvenile temporal arteritis, Orphanet encyclopedia. January 2005:**
http://www.orpha.net/data/patho/GB/uk-junvenile-temporal-arteritis.pdf