

Juvenile temporal arteritis

Sirs,

A 35-year-old Caucasian male presented with a 4-month history of malaise and bitemporal headache. Otherwise, the patient's medical history was unremarkable. On physical examination, a slightly painful swelling of the temporal arteries on both sides and a palpable mass in the area of the right parotid gland were noticed. Colour duplex sonography depicted multiple aneurysms of the frontal branches of the superficial temporal arteries (Fig. 1A), but no evidence for an inflammatory "halo". Sonography further showed a 2x1 cm hypoechoic mass of the right parotid gland with pronounced vascularisation. There were no pathological findings in abdominal sonography and chest x-ray. The differential blood count and the values for C-reactive protein, erythrocyte sedimentation rate (ESR) and antinuclear antibodies all were unremarkable, but an elevated serum IgE (5802 IU/ml, normal range <100 IU/ml) was conspicuous. Biopsies of the temporal artery and the right parotid gland were performed. Histological examination of the temporal artery biopsy showed luminal thrombosis and abundant cellular infiltrates of the vessel wall with a predominance of eosinophilic granulocytes, but absence of giant cells (Fig. 1B). Histology of the parotid gland was considered to be consistent with sclerosing sialadenitis.

High-dose corticosteroids were initiated, resulting in rapid clinical improvement after one week. The patient then was lost to immediate follow-up, but 24 months later he was referred for a follow-up-visit. Symptoms and signs of temporal arteritis had resolved, but cervical and axillary lymphadenopathy and tuberous cutaneous infiltrates were present. Subsequently, diagnosis of primary cutaneous T-cell-non-Hodgkin-lymphoma (gamma-delta-phenotype) was confirmed

by histology. As lymphoma associated complications did not occur, the patient was followed for about two years without treatment initiation in the oncology department.

Our case fits the criteria of juvenile temporal arteritis (JTA), *i.e.* age <50, normal ESR, histology with eosinophilic panarteritis and lack of giant cells (1). This rare condition (less than 30 cases reported in the literature) is an important differential diagnosis of aneurysms of the superficial temporal arteries in younger patients (2). Colour duplex sonography can be considered the first-choice diagnostic method, but diagnosis needs to be confirmed histologically (2).

JTA has been reported in association with angiolymphoid hyperplasia with eosinophilia (ALHE) and with Kimura's disease, respectively (1-3). These closely related entities are characterised by an eosinophilic soft tissue inflammatory reaction with variable degrees of vascular proliferation. ALHE typically presents with multiple, erythematous nodules of the skin, mainly in young women. Kimura's disease occurs mainly in young men of Asian descent with large tumours localised in the salivary glands or the subcutaneous tissue and lymphadenopathy. Eosinophilia and elevated serum IgE are frequently seen in both ALHE and Kimura's disease (1, 3). An association between JTA and T-cell lymphoma has not been described yet. However, several cases of ALHE and Kimura's disease with concurrent or subsequent development of peripheral T-cell lymphoma have been reported (3). Moreover, clonal T-cell receptor gene rearrangement was demonstrated in the tissue lesions of ALHE-patients (4, 5).

Whether or not there is a causal relation between JTA and T-cell lymphoma in the case presented remains unclear. Histology of the parotid mass did not confirm the suspected diagnosis of Kimura's disease in our patient. However, one could speculate that the parotid lesion mass was misdiagnosed and in fact was a lesion secondary to Kimura's

disease. Of note, the parotid tissue specimen was reviewed with this regard, but despite the presence of focal eosinophilic infiltrates, there was no histological evidence for Kimura's disease.

Based on our experience with the patient presented here, we recommend a careful initial diagnostic workup and regular follow-up with emphasis on signs and symptoms of lymphoproliferative disorders in patients presenting with JTA.

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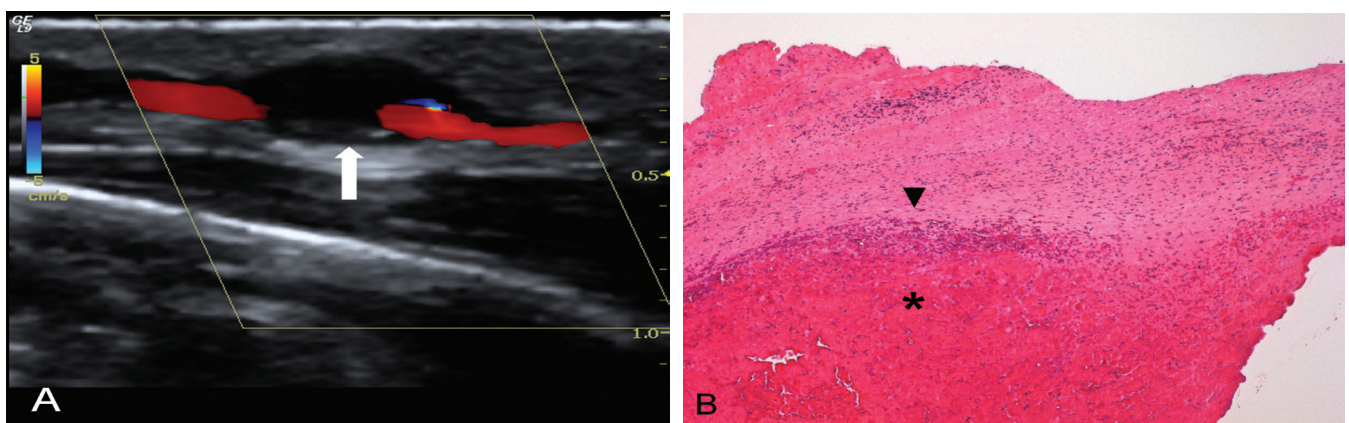


Fig. 1. (A) Aneurysms of the right temporal artery (arrow) depicted by colour duplex sonography. (B) Histology of the right temporal artery biopsy (H&E stain, original magnification x20), revealing luminal thrombosis (asterisk) and abundant cellular infiltrates of the vessel wall with a predominance of eosinophilic granulocytes (arrowhead), but absence of giant cells.