syndrome [5], who later developed a sixth nerve palsy. More recently, Mitra and Koul published a report of a 10-yr-old girl with acute unioocular visual loss associated with papilloedema [6]. The authors describe several separate presentations over the course of 3 yr with manifestations of neuro-Behçet’s, including various cranial nerve lesions, hemiparesis and sensory disturbances, associated with active cerebrospinal fluid and magnetic resonance imaging abnormalities. A brisk response to systemic corticosteroids was noted, in keeping with the experience of our patient.

The important differential diagnoses of bilateral papilloedema in the context of Behçet’s syndrome are cerebral venous thrombosis and central nervous system infection. The latter, of particular concern in the immunosuppressed patient, may be excluded by imaging and lumbar puncture. Cerebral venous sinus thrombosis is well described in Behçet’s syndrome [7, 8] and is usually characterized by severe headache; the diagnosis can be made by magnetic resonance angiogram.

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High-resolution MRI in giant cell arteritis with multiple inflammatory stenoses in both calves

Sir, giant cell arteritis (GCA) commonly involves the cranial arteries in a segmental pattern [1–3]. In the majority of cases, the affection of the temporal arteries can be demonstrated by duplex ultrasonography [4] and is confirmed by biopsy. In a study of 246 patients with proven GCA, however, 23 had a widespread arteritis, and three of these 23 patients presented solely with symptoms due to vasculitis of the lower extremities [5]. Here we present the capacity of high-resolution MRI to depict the unusual vascular involvement in a 65-yr-old man with proven GCA.

He presented with increasing claudication of both legs for the previous 3 months. The medical history was inconspicuous, except for a well-controlled allergic bronchial asthma. No cardiovascular risk factors, such as hypertension, hyperlipidaemia, diabetes and smoking, were present. Just before admission, new onset of a bilateral headache had been interpreted as sinusitis. Additionally, he reported increasing night sweat during the last 3 months. The initial laboratory findings revealed a severe inflammatory response [ESR 85 mm/1st h, CRP 131 mg/l (normal value <5)] but otherwise normal results for the differential blood count and renal and liver function. ANA, ANCA and rheumatoid factor were undetectable. In the physical examination, both dorsalis pedis arteries were impalpable, while the superficial temporal arteries were hard and tender to palpation. Chest X-ray and ultrasound of the abdomen showed no signs of underlying malignant disease. A digital subtraction angiography (DSA) was performed, and showed multisegmental stenoses and occlusions of the lower limb arteries and the formation of multiple collaterals (Fig. 1A). High-resolution MRI of the superficial temporal artery (Fig. 1B) and right leg (Fig. 1D) revealed inflammatory changes of the vessel walls, such as thickening and contrast enhancement, underlying the stenoses and obstructions. Suspecting GCA, a biopsy of the right temporal artery was performed. The histology demonstrated a severe mononuclear infiltration of the arterial wall compatible with GCA. The patient was immediately started on a high dose of steroids (1 mg/kg body weight) and aspirin 100 mg once daily. Due to the systemic involvement, empirical treatment with five pulses of monthly intravenous cyclophosphamide was added to the therapy and switched thereafter to oral methylprednisolone (15 mg/wk). The steroids were gradually tapered to 5 mg/day according to clinical and serological assessment. The initial headache improved within 24 h and the pain-free walking distance increased within 9 months from 25 to 400 m. Angiographic re-evaluation after 6 months showed a still decreased but improved perfusion of both dorsalis pedis arteries.

After 15 months of therapy with MTX and low-dose steroids, the patient reported no further improvement of the claudication and CRP values, which were slightly elevated [8 mg/l (normal value <5)]. Re-evaluation by MRI revealed the absence of inflammation in the temporal artery (Fig. 1C) and significantly decreased inflammatory signs in the lower calf arteries (Fig. 1E).

We present an unusual case of GCA with stenoses and occlusions of the lower limb arteries leading to claudication as pertinent clinical findings. In such a case, reaching the proper diagnosis is difficult unless the superficial temporal arteries are involved. The diagnosis of extracranial involvement is often only indirect and is made on the basis of clinical findings. Ultrasound is the primary imaging procedure of choice in GCA. However, it is observer-dependent, restricted to superficial arteries and not suitable to the examination of widespread arterial involvement. PET scanning has been suggested as an alternative method to depict vascular involvement [6], but radioactivity, lower resolution and high cost are disadvantageous. Recently, the first results of high-resolution MRI vessel wall imaging of the temporal artery in GCA [7] demonstrated mural thickening and contrast enhancement in medium-sized arteries defining the extent and intensity of vascular involvement and thus active disease [8].

In our case, the involvement of the superficial temporal arteries and the lower leg arteries was correctly depicted by high-resolution MRI. A control MRI under treatment showed an improvement in vessel wall inflammation. Interestingly, at that time the patient still presented with low systemic inflammation and no further improvement of claudication. Accordingly, the MRI then showed residual contrast enhancement.

Based on the clinical and laboratory parameters and supported by the MRI finding, immunosuppressive therapy was increased.
This case demonstrates that GCA can be more widespread than commonly expected, including arteries of the lower extremity. Especially for patients without involvement of the temporal arteries or in case of negative biopsy, high-resolution MRI provides excellent additional diagnostic support and will enhance our knowledge of the extent of the arterial involvement in GCA.

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Fig. 1. (A) DSA shows multisegmented stenoses and occlusions of the lower limb arteries with the formation of collaterals. High-resolution MRI of (B and C) the right superficial temporal artery and (D and the E) the right calf. Contrast-enhanced, fat-saturated spin echo sequence clearly depicts mural thickening and contrast enhancement of the inflamed superficial temporal artery (arrow in B) and the anterior tibial artery (arrow in D). The accompanying veins (arrowheads in D and E) show a homogeneously increased signal intensity due to intravenous contrast agent within their comparatively larger lumen. Biopsy of the temporal artery confirmed the diagnosis of GCA. A follow-up examination after 15 months of corticosteroid treatment revealed the absence of mural inflammatory signs in the temporal artery (arrow in C) and significantly decreased mural inflammatory changes in the anterior tibial artery with less restricted lumen of the artery (arrow in E).