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An unusual cause of transient small bowel thickening

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A 23-year-old Swiss female presented with lower back pain lasting for four weeks accompanied by night sweats for one week. Two years earlier, she had suffered from headaches, fatigue and dysaesthesia of both arms; a CT scan of the head was then normal. On admission, she was pale and distressed, and a faint systolic murmur was noted at the left sternal border. Blood pressure was 117/55 mmHg on the right and 100/60 mmHg on the left arm. Initial laboratory testing revealed a haemoglobin of 7.8 g/dl, the C-reactive protein concentration was 175 mg/l (<10 mg/l) and the erythrocyte sedimentation rate (ESR) was accelerated to 130 mm/h. An abdominal CT scan showed hepatosplenomegaly, enlarged retroperitoneal lymph nodes and a circumscribed thickening of a small bowel loop segment measuring about 10 cm in length (Fig. 1). In addition, a faint thickening of the abdominal aortic wall was seen. Infection, chronic inflammatory or lymphoproliferative disorders were considered. However, various serologic parameters including those for HIV, EBV, CMV, and syphilis, as well as antinuclear, ANCA and anti-DNA antibodies and the rheumatoid factor were negative. Serum immune fixation was unremarkable and no pathogenic micro-organisms were detected in stool cultures. Bone marrow biopsy showed reactive changes with increased myelo- and megakaryopoiesis, plasma cells and macro-

phages. Two days after admission she complained of abdominal cramps and vomited, however, there were no clinical or radiological signs of intestinal obstruction. The C-reactive protein was now 232 mg/l. An upper gastrointestinal panendoscopy was unremarkable. Surgical exploration was scheduled but was cancelled following spontaneous relief of the gastrointestinal symptoms. Finally, MR-angiography (Fig. 2) performed 3 days after the CT scan clearly demonstrated inflammation and stenoses of the thoracic and abdominal aorta involving the supra-aortic branches and both renal arteries. In addition, a diffuse contrast enhancement of the mesenteric fat was noted while the appearance of the bowel was now unremarkable.

Based on the clinical and radiologic findings, a diagnosis of Takayasu arteritis type V was made [1]. Initial treatment consisted of 100 mg of prednisone daily, which was reduced to 50 mg after four days. The lower back pain and the night sweats disappeared rapidly and the ESR dropped to 62 mm/h. One month later a bruit in the fossa supraclavicularis was noted, an echocardiography was normal, ESR was 30 mm/h and methotrexate 15 mg per week was started. One year after the diagnosis the blood pressure difference had disappeared, but intermittent, abdominal pain and bi-frontal headaches as well as elevation of the ESR persisted. Methotrexate was first replaced by mycophenolate (2 g per day) and finally by azathioprine (100 mg per day). Over a period of another year under this treatment, the headaches and the abdominal pain improved considerably, the ESR remained slightly elevated at 20 mm/h.

Takayasu arteritis is a chronic inflammatory disease of young women characterised by non-specific constitutional symptoms in its early pre-pulselessness stages [2-4]. It primarily involves large and medium-sized arteries leading to stenoses, aneurysms or rupture. The clinical presentation is variable, as is its

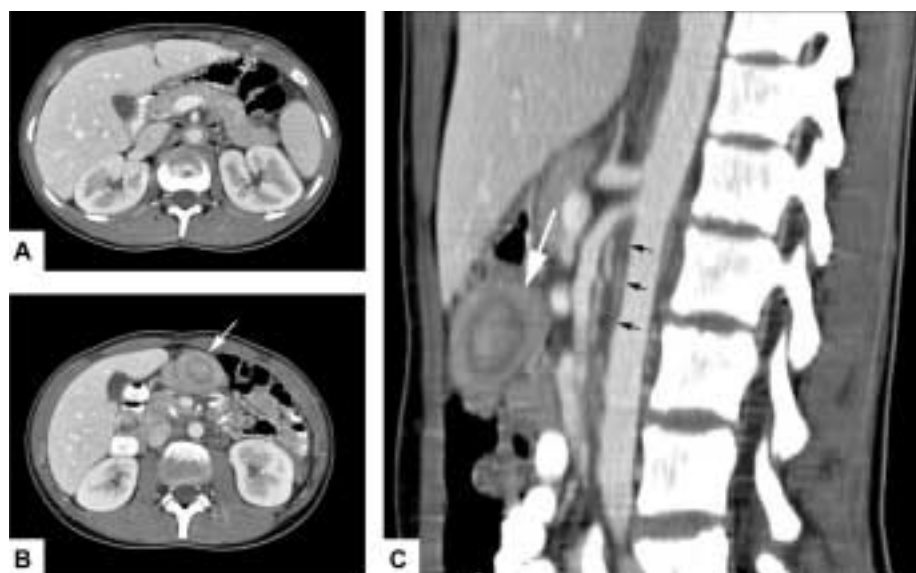
progression, response to treatment and prognosis, which depend on localisation and extent of the vasculitis. Morbidity and mortality can be substantial, however, early diagnosis and treatment may improve the outcome [4]. In the present case anaemia, hepatosplenomegaly, lymphadenopathy and bowel wall thickening initially suggested infectious or lymphoproliferative disease. The blood pressure difference, on the other hand, was a subtle sign for the presence of a vasculopathy. Indeed, in a series of 60 patients with Takayasu arteritis, blood pressure difference has been described in 17 % of the patients at onset of the disease [3]. The initial symptoms in our patient, headache, myalgia and fatigue occur in more than 50% of the patients, but gastrointestinal symptoms are rare (9%). A vasculitic involvement of the mesenteric arteries is seen in 18 % of the patients, often in the absence of symptoms possibly due to the formation of collaterals [3;5]. An association with inflammatory bowel disease, hepatobiliary disorders and lymphadenopathy has been reported only in selected cases [6;7]. However, transient bowel wall thickening most likely due to intestinal ischaemia as in the present case has not been described so far. The final diagnosis of Takayasu arteritis was established by MR-angiography, a highly sensitive and specific diagnostic procedure [8]. Taken together, Takayasu arteritis may present with abdominal symptoms, is particularly difficult to diagnose in its pre-pulseless stage, and may therefore be easily overlooked, especially in patients not stemming from Asia.

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Figure 1

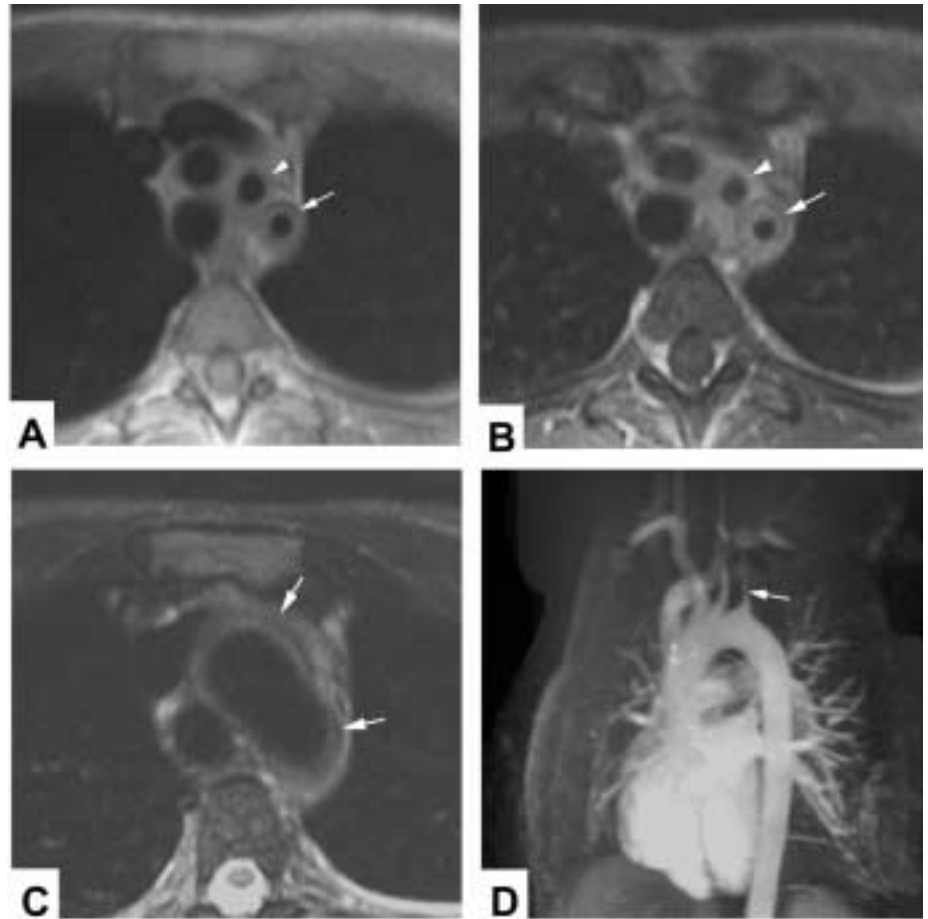
Axial contrast-enhanced CT scans at the level of the pancreas (A) and the superior mesenteric artery (B) and multiplanar reconstruction in the sagittal plane (C). Thickening of a small bowel loop (white large arrows in B and C). Enlarged para-aortic lymph nodes (small white arrows in B) and slight thickening of the aortic wall (black arrows in A and C).



No outside financial support or other financial relationships presenting a potential conflict of interest

Figure 2

Axial T₁-weighted MR images before (A) and after intravenous administration of gadolinium chelate (B): luminal changes of the left subclavian (arrow) and the left common carotid artery (arrowhead) with wall thickening and contrast enhancement. Axial T₂-weighted MR images at the level of the aortic arch: hyper-intense wall thickening (C). Sagittal multiplanar intensity projection of the three-dimensional MR angiography: extent of the luminal stenosis of the proximal left subclavian artery (D).



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