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## Early diagnosis of pediatric Takayasu arteritis (TA) not fullfiling the ACR criteria

Sirs,

TA is a chronic vasculitis of unknown etiology characterized by granulomatous inflammation of large and medium-sized vessels that affects the aorta, its main branches and the pulmonary arteries (1). Onset is most common during the third decade of life, but childhood disease has been reported.

Angiography has been the standard imaging tool for the diagnosis and follow-up of TA (2), but less invasive techniques, such as computed tomography (CT) and magnetic resonance angiography (MRA), can visualize early changes (vessel wall thickening without stenosis or dilatations, mural edema, increased mural vascularity) not detectable by conventional angiography, and may lead to earlier diagnosis (3-4).

In 1990, the American College of Rheumatology (ACR) defined specific diagnostic criteria for TA (5), and diagnosis is confirmed by the presence of at least three of the six criteria, but most of these are clinical features of the disease's late fibrotic phase and thus may not be fulfilled in the early inflammatory stage.

In this report we describe a case of TA the diagnosis of which was established by CT and MRA imaging, even though the ACR criteria were not met.

A 16-year-old girl was admitted to the Rheumatology Department of the Children's Hospital in Rome because of malaise and fever of unknown origin for several days which was unresponsive to antipyretics. She complained of sharp dorsal pain for two months that worsened with fever. Cardiac murmur was detected out but no other bruit. She did not present decreased pulsation of any peripheral pulse or significant difference in systolic pressure in the arms. C-reactive protein (CRP), erythrocyte sedimentation rate (ESR) and complement were repeatedly elevated, normocitic anemia was also detected, and ANA were negative. Echocardiography revealed a bicuspid shaped aortic valve with regurgitation and aortic root dilatation. Chest and abdominal CT revealed wall thickening of the aortic arch, of its primary branches and of the entire thoracic and abdominal aorta. MRA revealed a marked dilatation of both the ascending and descending aorta with valve strip immobility and valve regurgitation during the diastolic phase. Although our patient had just one of the clinical ACR criteria, TA with bicuspid aortic valve regurgitation was diagnosed on the basis of the MRA. The patient started oral prednisone at 2 mg/kg/dose. Steroid therapy was quickly effective but methotrexate was also added at 15 mg/m$^2$/dose for disease control. She quickly recovered, her pain improved and she had no fever for 3 months. However, she then relapsed with fever and marked increase of acute phase reactants. Infliximab was started (5 mg/kg at time 0, 2, 4 and 8 weeks and then every 8 weeks) with prompt and persistent remission.

Although less invasive cross-sectional methods such as CT and MRA can effectively demonstrate thickening of vessel walls, which may be the earliest manifestation of the disease, the arteriogram abnormality is still one of the diagnostic criterion of TA. The Pres Vasculitis study group is now working to change the classification criteria for vasculitis in children (6). For the diagnosis of TA, angiographic abnormalities detected by conventional, CT or MR angiography could be accepted (3-4).

The early diagnosis of TA may be difficult since early symptoms such as fatigue, malaise, joint pain and fever are nonspecific. When our patient was hospitalized she had no claudication, decreased pulsation, remarkable difference in systolic pressure or arterial bruits, but only a cardiac murmur. Because of that, we performed an echocardiogram that revealed the dilatation of the aortic root and subsequent imaging studies which led to the diagnosis.

Our patient did not meet the ACR 1990 criteria for TA, probably because of the early stage of the disease. With our case we want to point out the accuracy of the new imaging methods to identify very early disease manifestation.

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