

Takayasu's arteritis with familial Mediterranean fever

Sirs,

Familial Mediterranean fever (FMF) is an autoinflammatory disease with autosomal recessive inheritance. Coexistence of FMF and Takayasu's arteritis (TAK) occurs rarely and there are only two cases reported in the literature so far. We herewith report the case of a male patient who is the third case of coexistence of FMF and TAK.

A 22-year-old male patient presented with the complaints of weight loss, claudication in arms, left radial artery pulselessness for the last few months. He had been diagnosed as FMF at the age of 17 years, because of recurrent abdominal pain, and fever attacks. Colchicine treatment had been started 1.5 mg daily. His father and uncle also have had diagnosis of FMF. In physical examination, pulse could not be felt in both radial arteries. Laboratory tests were as follows: erythrocyte sedimentation rate (ESR): 80 mm/h, C-reactive protein (CRP):100 mg/L. On neck-thorax and abdominal CT angiography, occlusion was observed in the right subclavian and left subclavian arteries, and diffuse wall thickness was seen to increase in thoracic and abdominal aorta (Figs. 1, 2). He was diagnosed with TAK and administered 1 gr pulse steroid for 3 days and for maintenance treatment 1 mg/kg/day prednisolone was started. The dose of corticosteroid was decreased for 20% monthly. Cyclophosphamide (Cyp) was administered intravenously at the dose of 1 gr/month. At the sixth month Cyp administration, control evaluation with this treatment, ESR was found to be: 4 mm/h and CRP:6 mg/L. TAK is still in remission and the patient is being followed with medical treatment including azathioprine 150 mg, colchicine 1.5 mg daily.

As supported by many case reports and case series, vasculitis mainly involving small and medium sized vessels may occur in a minority of FMF patients. In FMF patients, the prevalence of polyarteritis nodosa and Henoch-Schönlein purpura have increased (1-3). In this case series, the association between FMF and TAK have not been observed. To our knowledge, in the literature only two cases of coexistence between FMF and TAK have been reported so far (4, 5). Hence, the fact that two previous cases and our cases are male is interesting. In one of these cases, as the patient was resistant to cyclophosphamide, he was administered anti-TNF- α treatment and the other case received maintenance treatment with azathioprine following Cyp (4, 5). Azathioprine was administered for maintenance in the present case following Cyp. In both cases, the diagnosis of TAK was made years after the diagnosis of FMF and MEFV mutations M694V/V726A and R314R, E474E, Q476Q were found het-



Fig. 1. CT angiography, occlusion was observed in right subclavian artery and left subclavian and diffuse wall thickness was seen to increase in thoracic aorta.

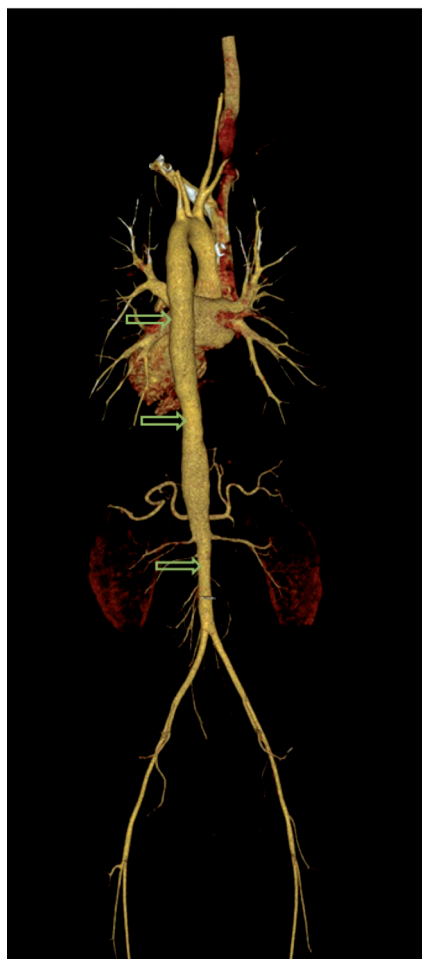


Fig. 2. CT angiography diffuse wall thickness was seen to increase in thoracic and abdominal aorta.

erozygote. In the present case, we found FMF gene compound heterozygous mutation V726A and M694V. Although the cause of FMF-associated vasculitis is not completely known, it is thought that both environmental and genetic factors may play a role. MEFV mutation leads to IL-1 β release by stimulating immune system. IL-1 β is the predominant cytokine in FMF and is thought to cause vasculitis by lead-

ing to endothelial cell dysfunction. In FMF patients, acute phase markers such as ESR, CRP, fibrinogen and serum amyloid A are frequently increased during episodes (6). Even if inflammation markers return to normal levels in attack free periods, subclinical inflammation may continue in some cases (7, 8). High levels of inflammatory markers may lead to a delay in the diagnosis of vasculitis and sometimes to an increase in the costs of diagnosis and treatment, due to unnecessary investigations. Therefore, in FMF cases presenting with high acute phase reactant levels, if there are accompanying ischaemic symptoms, vasculitis should be considered in differential diagnosis. As the number of cases demonstrating the coexistence of FMF and TAK are increasing, it will be possible to obtain more detailed information on this issue.

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