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When Takayasu mimics pulmonary hypertension — severe pulmonary artery stenosis — what to do?

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When Takayasu mimics pulmonary hypertension — severe pulmonary artery stenosis — what to do?

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Short title: Severe pulmonary arteries stenosis caused by Takayasu disease
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27-year-old female, without history of cardiac diseases, with increasing fatigue and dyspnoea, up to 4th class of New York Heart Association (NYHA) during the previous two weeks was admitted to the cardiology department with suspicion of pulmonary arterial hypertension (PAH).

On admission, the patient presented resting dyspnoea, tachycardia 120 bpm and blood pressure 100/60 mm Hg. Capillary blood oxygen saturation was 80%. A loud systolic murmur was present over an entire chest. Physical examination did not reveal peripheral oedema nor features of venous thromboembolism in the lower extremities.

Laboratory test revealed amino-terminal pro-brain natriuretic peptide of 10745 pg/ml (n = 0–125 pg/ml), increased inflammatory parameters (C-reactive protein 25 mg/l; white blood cells 13 $\times 10^3$/ul), serum concentration of IgG4 of 60 mg/dl. Echocardiography showed overload of the right ventricle, tricuspid annulus dilatation with torrential regurgitation. Calculated systolic pulmonary artery pressure was 110 mm Hg. Angio-computed tomography (angio-CT) scan

revealed inflammatory infiltration of the pulmonary trunk, involving the pulmonary arteries and causing critical stenoses (Figure 1A–B). No embolic material was found in the scan.

Based on clinical and angiographic criteria proposed by the Japanese Research Committee on Vasculitis Syndromes [1], the initial diagnosis of Takayasu arteritis (TA) was stated. These criteria included: 1) angiographic evidence of narrowing or occlusion of the aorta or large arteries in CT or magnetic resonance imaging; 2) early age of onset; 3) presence of markers of inflammation; and 4) exclusion of atherosclerosis, other inflammatory diseases or congenital vascular abnormalities.

The patient received 15 mg per day of prednisone and two intravenous infusions of 400 mg cyclophosphamide, achieving resolution of resting dyspnoea. During the next 6 months, the patient received a total dose of 4.8 g cyclophosphamide and achieved total remission of clinical symptoms. Control angio-CT scan showed substantial regression of stenosis (Figure 1C–D). Control parameters are shown in Figure 1E. Further hospitalizations were planned for performing imaging examinations and evaluating the effects of the treatment.

TA is a large-vessel vasculitis affecting the aorta and its primary branches: subclavian artery (33.7%), the renal artery (25.3%) and the common carotid artery (21.7%), the pulmonary artery comprised only 0.8%. Inflammatory processes in the acute phase cause thickening of the arterial wall. The chronic phase causes vascular fibrosis, stenosis and occlusion, resulting in congestive heart failure and pulmonary hypertension [1].

We present a rare case of TA involving pulmonary arteries that clinically mimicked signs of severe PAH. Clinical presentation: age, sex, symptoms, amino-terminal pro-brain natriuretic peptide (NT-proBNP) level and echocardiographic signs suggested PAH. Detailed echocardiographic assessment and the CT-scan allowed to revealed that the PAH — like features of right ventricle overload were due to severe pulmonary arteries stenosis. In differential diagnosis, it was essential to exclude other inflammatory arteries stenosis, mainly giant cell arteritis (GCA). While TA is seen in young females, GCA affects older patients. In GCA inflammation processes often involve external carotid artery, which is not seen in TA. Arterial wall thickening can be due to retroperitoneal fibrosis and therefore mimics TA, but TA lacks peritoneal diffusion [2]. IgG4 related disease was also taken into consideration. But with serum concentration of IgG4 of 60 mg/dl, it was unlikely (96% negative predictive value) [3].

The first line of treatment for TA is glucocorticoids. Half of the patients require second-line agents: cyclophosphamide, methotrexate or biologic drugs [4]. Endovascular or surgical interventions in arteries stenosis may be necessary once irreversible stenosis starts to develop.

Because the rates of complications are the highest in patients with acute inflammatory lesions, interventional therapy should be avoided during the acute phase of disease [5].

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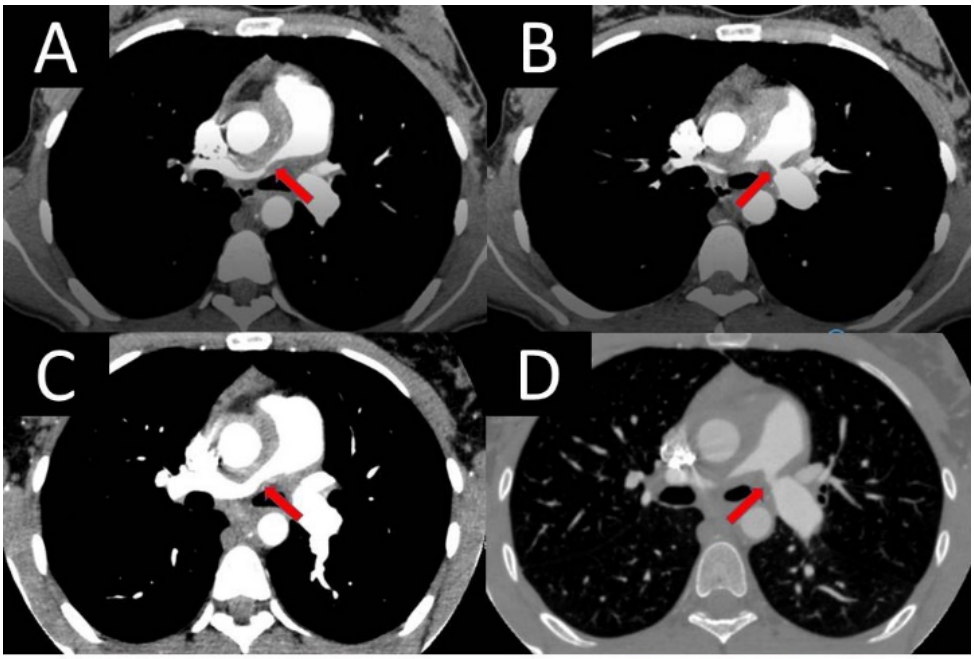


E

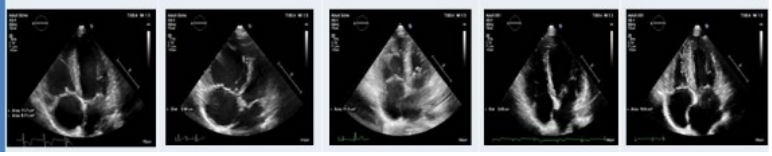
	Initial hospitalization	Control after 1 month	Control after 2 months	Control after 4 months	Control after 6 months
NYHA class	IV	III/IV	II	I	I
NT-proBNP [pg/ml]	10745=>4946	11087=>3138	1326	607	496
RVOT [mm]	48	46=>40	32	26	30
RVID [mm]	44	47=>44	39	34	33
LVEDD [mm]	38	37=>40	36	41	44
LVESD [mm]	18	17=>23	21	23	26
RVSP [mmHg]	110	131=>94	80	52	30
AcT [ms]	77	78=>87	95	110	150
Pericardial effusion [mm]	6	4	0	0	0
6-MWT distance [m]	0	110	320	440	500
Total dose of cyclophosphamide [mg]	800	1800	2800	3800	4800
Echocardiography					

Figure 1. Computed tomography with contrast. **A.** The initial angio-CT scan showed critical stenosis in pulmonary arteries, causing 99% narrowing of the lumen of RPA. **B.** 95% of the LPA. **C.** Control angio-CT scan after six months showed substantial regression of stenosis in both RPA; **D.** and LPA. **E.** Clinical, laboratory and echocardiography parameters according to the total dose of cyclophosphamide in subsequent hospitalizations.

Abbreviations: 6-MWT, 6-minute walking test; AcT, acceleration time; LVEDD, left ventricular end-diastolic diameter; LVESD, left ventricular end-systolic diameter; NT-proBNP, amino-terminal pro-brain natriuretic peptide; NYHA, New York Heart Association class; RVIT, right ventricular inflow tract; RVOT, right ventricular outflow tract; RVSP, right ventricular systolic pressure