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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Late Takayasu Arteritis Diagnosis in a Female Patient With Prior Coronary Artery Bypass Grafting

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ABSTRACT

Takayasu arteritis is a rare cause of cardiovascular morbidity in the Western world. As consequence, vasculitis may be misdiagnosed and treated as atherosclerotic cardiovascular disease. We present a case of late Takayasu arteritis diagnosis, in a female patient with peripheral artery disease and previous coronary artery bypass grafting. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:19–23) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 55-year-old Caucasian female patient of Albanian origin presented to the emergency department complaining of retrosternal chest pain, deteriorating during the past 15 days. The pain lasted more than 30 min, mainly at rest, was not related with effort, and was associated with malaise and fatigue.

LEARNING OBJECTIVES

- In female patients with severe coronary artery disease, concomitant stenosis of large vessels and absence of risk factors of atherosclerotic disease, Takayasu arteritis should be included in the differential diagnosis.
- Transesophageal echocardiogram, computed tomography, and magnetic resonance imaging are useful in characterizing aortic wall lesions.

On physical examination she was afebrile, with a normal heart and respiratory rate and arterial blood oxygen saturation. Brachial blood pressure measurement was higher in the left than in the right arm (149/65 mm Hg vs. 125/55 mm Hg) and a diminished pulse was found in the palpitation of the left radial artery. The auscultation revealed normal heart and respiratory sounds and a bruit in the right carotid artery.

Electrocardiogram showed sinus rhythm, without ST-segment or T-wave abnormalities. High-sensitivity troponin I, D-dimers, C-reactive protein, and white blood cells were within normal range. Normocytic normochromic anemia (hematocrit 34%) was found.

As an acute coronary syndrome seemed unlikely (absence of electrocardiogram changes and troponin elevation), a transthoracic echocardiogram was performed to rule out an aortic syndrome or pulmonary embolism, because of the prolonged and persisting chest pain. This demonstrated a normal left ventricle

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Informed consent was obtained for this case.

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ABBREVIATIONS AND ACRONYMS

MRI = magnetic resonance imaging

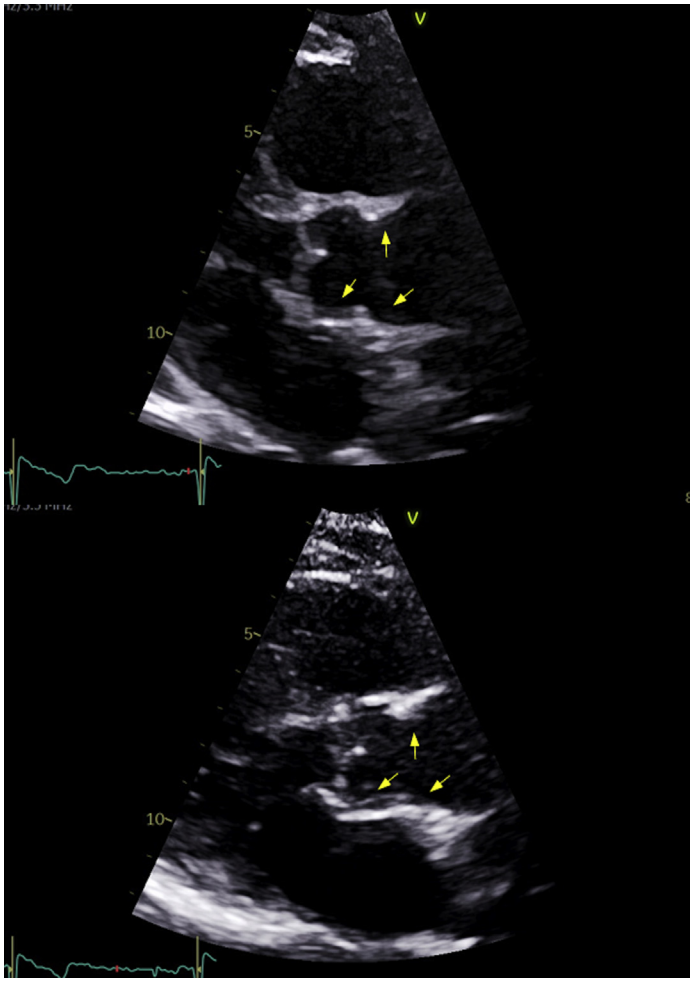
TOE = transesophageal echocardiogram

size, without any wall motion abnormalities (estimated ejection fraction: 55%) and a normal trileaflet aortic valve. However, a circular echogenic thickening in aortic root at the level of sinotubular junction (Figure 1) without significant aortic regurgitation was revealed.

MEDICAL HISTORY

In spite of not stating a history of traditional cardiovascular risk factors, coronary artery bypass grafting was conducted 5 years earlier, with left internal mammary artery grafted to left anterior descending artery and 2 saphenous vein grafts grafted to the right coronary artery and Ramus Intermedius, respectively, due to unstable angina and left main disease. Left carotid artery stenosis and left subclavian artery occlusion were diagnosed concomitantly.

FIGURE 1 Transthoracic Echocardiogram: Proximal Aorta Wall Thickening



Transthoracic echocardiogram- parasternal long axis views focused on the aortic root and proximal ascending aorta, demonstrating echogenic, longitudinal thickening of 4 mm in the aortic root at the level of the sinotubular junction and proximal ascending aorta **(yellow arrows)**.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included an aortic intramural hematoma (based on the findings of transthoracic echocardiogram), an acute coronary syndrome, and pulmonary embolism.

INVESTIGATIONS

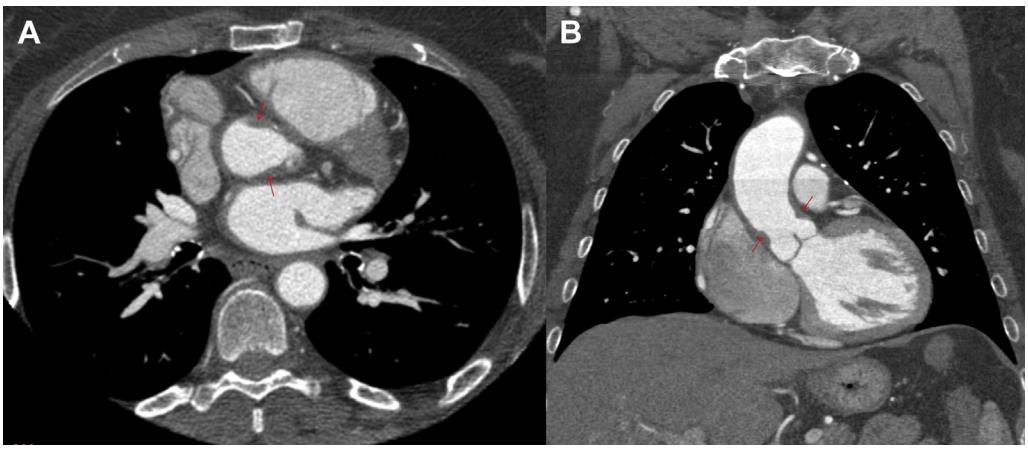
A computed tomography angiography of the pulmonary artery, aorta, and coronary arteries was performed. The test excluded pulmonary embolism and revealed normal patency of the saphenous vein grafts to the right coronary artery, a 70% stenosis of the left internal mammary artery at insertion to the left anterior descending artery, a subtotal obstruction of the left and multiple stenoses of both subclavian arteries. Brachial stenoses distally to the left subclavian occlusion were also found, explaining the differences in blood pressure and radial pulse between the arms. An annular thickening of the proximal ascending aorta also was observed (Figure 2).

The patient was admitted to the cardiology department for further investigation. To evaluate the findings on the aorta, a transesophageal echocardiogram (TOE) was performed, which demonstrated an annular echogenic ring of 4-mm maximal thickness in the aortic wall above the level of sinotubular junction, extending toward the aortic arch. This aortic lesion was thought more likely to represent an inflammatory/fibrotic lesion rather than an intramural hematoma, as it was highly echogenic and homogeneous, with well-defined borders, with no flow by color Doppler and had a thickness <5 mm (Figure 3).

Meticulous clinical examination also revealed arthritis of the proximal and distal interphalangeal joints. At this point, the patient was evaluated by a rheumatologist and an extensive laboratory workup for autoimmune and infectious diseases was run: antinuclear antibodies, rheumatoid factor, anticyclic citrullinated peptide, antineutrophil cytoplasmic antibodies, serologic test for syphilis, hepatitis, and human immunodeficiency virus were negative. Interferon gamma release assay blood test was positive, indicative of previous tuberculosis. Erythrocyte sedimentation rate was mildly elevated (43 mm/h).

Taking into consideration the clinical and imaging findings, which enhanced the probability of large

FIGURE 2 **CTA of Coronary Arteries and Aorta**



CTA illustrates wall thickening of proximal ascending aorta **(red arrows)**. Ascending aortic dissection is ruled out. Proper image acquisition allows computed tomography pulmonary angiography to be performed without the need for second dose of intravenous contrast administration and concomitantly pulmonary embolism was ruled out. CTA = computed tomography angiography.

vessel vasculitis, with lower likelihood of angitis of infectious origin, magnetic resonance imaging (MRI) of the aorta and the great vessels was scheduled. The MRI revealed a concentric thickening of the aortic wall at the level of the aortic root and in lesser extent thickening of the ascending aorta and distally, with marked delayed enhancement, probably due to vasculitis. Other findings included extensive focal stenoses of carotid and subclavian arteries (**Figure 4**). These findings advocated over Takayasu vasculitis.

A computed tomography angiography was finally performed for classification of disease and demonstrated stenosis of the right renal artery, classifying the patient in type V.

MANAGEMENT

The patient was treated with methylprednisolone 0.5 mg/kg, which resulted in remission of the symptoms. The pharmacologic regimen also included aspirin, metoprolol, atorvastatin/ezetimibe, omeprazole, and levothyroxine, along with isoniazid for prophylaxis from tuberculosis and osteoporosis prevention treatment.

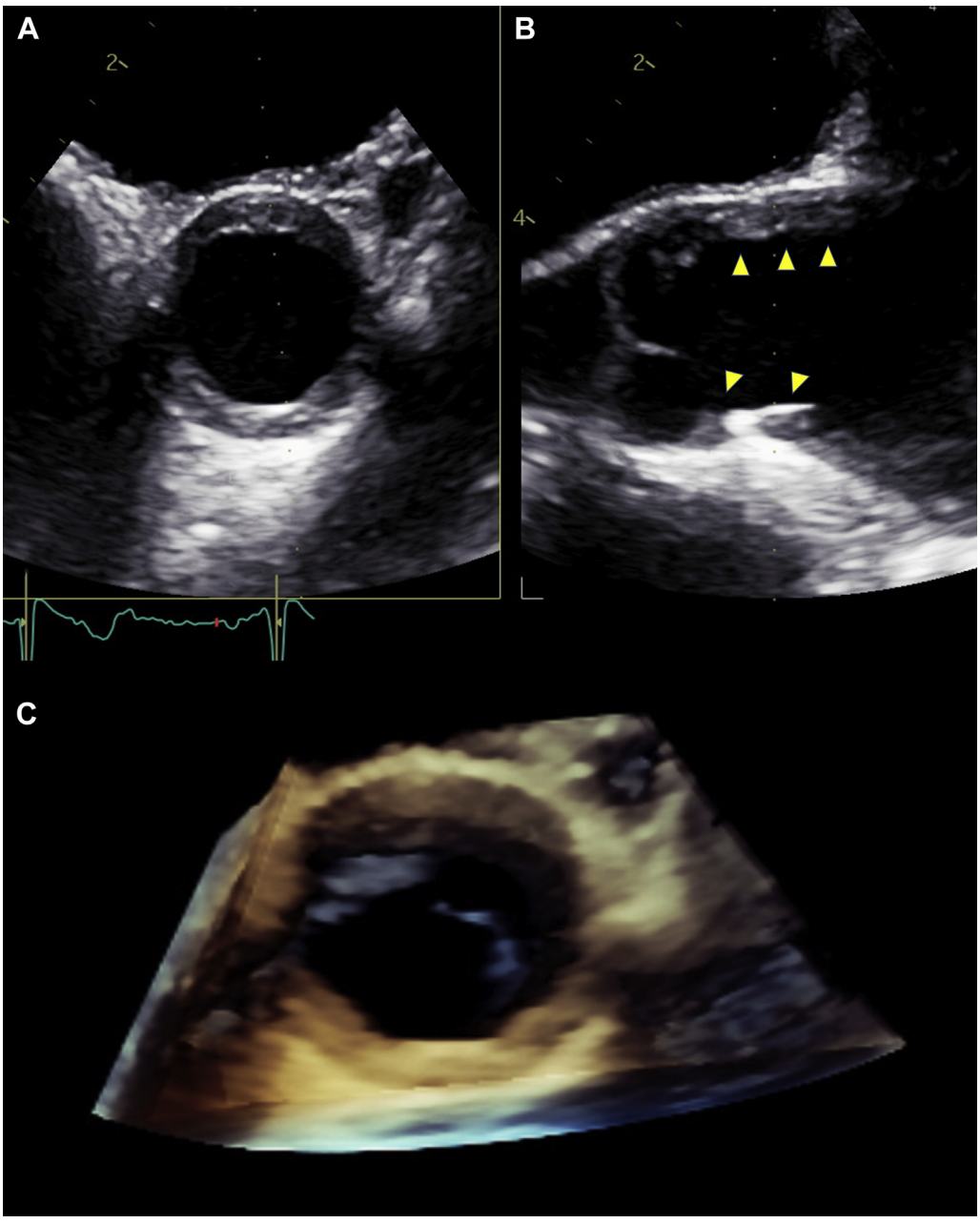
DISCUSSION

Takayasu arteritis is a granulomatous vasculitis of the large- and medium-size vessels, affecting mainly women and manifesting before the sixth decade of life. The incidence is greater in Asia than in Europe (0.4 to 1.5 per million). The etiology of Takayasu arteritis is still unknown; however, an autoimmune process is implicated, possibly triggered by common pathogens (viruses, bacteria, or mycobacterium tuberculosis) via a molecular mimicry mechanism (1,2). The clinical presentation includes a variety of constitutional symptoms and vascular manifestations related to the affected vessels. Measurement of the arterial blood pressure in both arms is a simple and helpful examination (difference of >10 mm Hg is observed in 45% to 69% of the patients) (2). Takayasu disease is usually diagnosed in those younger than 50 years of age. Thus, regarding our patient's age, the differential diagnosis should include other causes of large vessel vasculitis, namely giant cell arteritis and Behcet disease.

Commonly, giant cell arteritis affects medium and small arteries, resulting to headache and vision impairment in association with high erythrocyte sedimentation rate and C-reactive protein, whereas Behcet disease is associated with iridocyclitis, oral and genital ulcers, and is complicated by aortitis and vascular thrombosis. None of these findings were evident in our patient (3,4).

TOE is a simple and available method to assess lesions in the thoracic aorta. Imaging of the large vessels with MRI and computed tomography is useful to confirm the diagnosis. The sensitivity of MRI is similar to angiography in detecting stenotic lesions in the aorta and its main branches. Moreover, MRI depicts the disease's activity, demonstrating wall edema, even in subclinical cases (3,5).

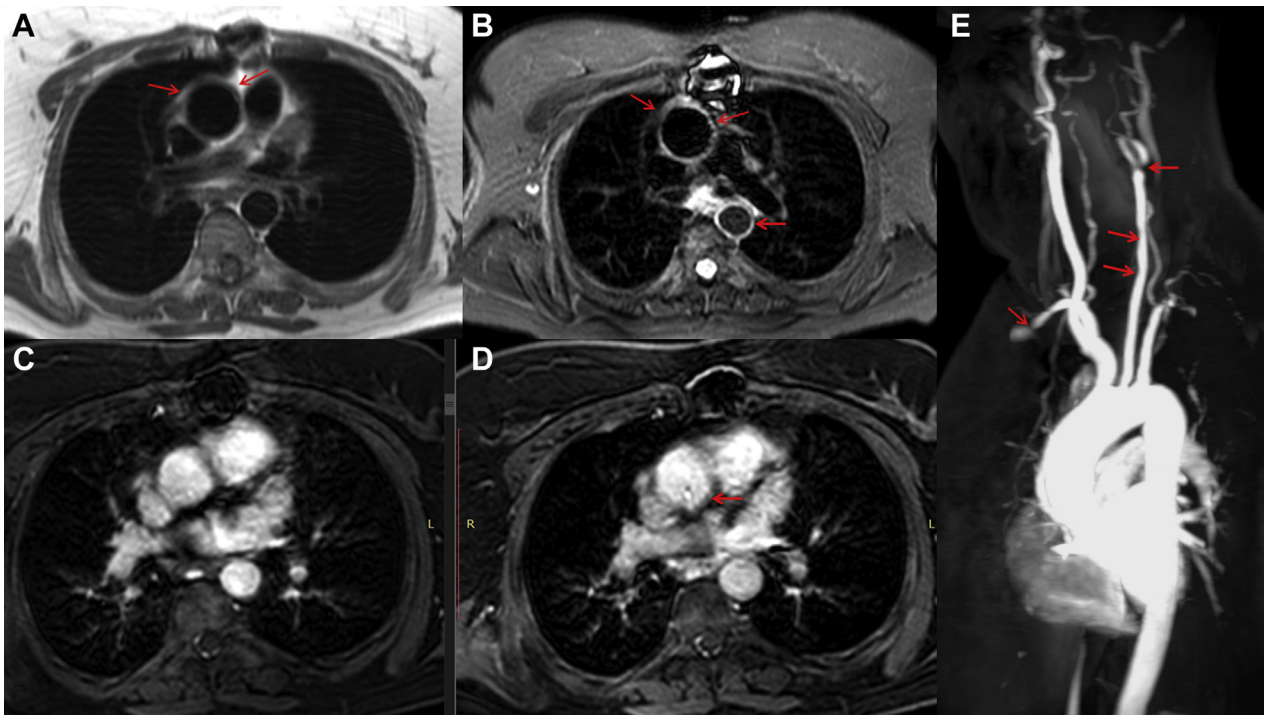
FIGURE 3 TOE: Proximal Aorta Wall Thickening



TOE biplane imaging of proximal ascending aorta in 45° (short axis) **(A)** and 135° (long axis) **(B)** vertical views. A longitudinal and annular thickening is illustrated, which is highly echogenic and homogeneous, with smooth, well-defined borders **(yellow arrows)**. **(C)** Three-dimensional illustration of proximal ascending aorta. TOE = transesophageal echocardiogram.

Concerning the arterial stenosis treatment, coronary artery bypass grafting is preferred over percutaneous coronary intervention due to high restenosis rates of stenting, although active inflammation is related to increased incidence of graft stenosis. The accurate and timely diagnosis and treatment is therefore of paramount significance to select the appropriate revascularization method and achieve the optimal postsurgery outcome (5).

FIGURE 4 **MRI: Angiography**



(A) Concentric and homogeneous thickening of the proximal wall of ascending aorta **(red arrows)**. **(B)** T2 sequences after fat suppression, with increased signal intensity of the arterial wall **(red arrows)**. **(C)** T1 sequences just after intravenous gadolinium administration. **(D)** Ten min after gadolinium administration, late gadolinium enhancement of the aortic wall is observed, a finding consistent with vasculitis **(red arrow)**. **(E)** Focal stenoses of the left internal and external carotid arteries and of the right subclavian artery are demonstrated **(red arrows)**. MRI = magnetic resonance imaging.

FOLLOW-UP

In a follow-up examination a year after diagnosis, MRI and TOE were performed. Findings were similar to the previous examinations. Moreover, the patient stated symptom improvement and no rehospitalization for any reason.

CONCLUSIONS

The practicing clinician should include Takayasu arteritis in the differential diagnosis in female patients with early findings of diffuse vascular disease. Measurement of the arterial blood pressure in both arms consists of a practical screening method. TOE, computed tomography, and MRI angiography are appropriate methods for thoracic aorta assessment and help differentiate between acute aortic syndromes and large vessel vasculitis.

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REFERENCES

1. Espinoza JL, Ai S, Matsumura I. New insights on the pathogenesis of Takayasu arteritis: revisiting the microbial theory. Pathogens 2018;7:73.

2. Seyahi E. Takayasu arteritis: an update. Curr Opin Rheumatol 2017;29:51–6.

3. Ikonomidis I, Makavos G, Katsimbri P, Boumpas DT, Parissis J, Iliodromitis E. Imaging risk in multisystem inflammatory disease. J Am Coll Cardiol Img 2019;12:2517–37.

4. Caspary L. Vasculitides of large vessels. Vasa 2011;40:89–98.

5. Erbel R, Aboyans V, Boileau C, et al. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases. Eur Heart J 2014;35:2873–926.

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