

Case Report

Takayasu's Arteritis Mimicking Aortic Intramural Hematoma in a Female Patient with Chest Pain

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Key words:
Takayasu's arteritis, chest pain, transesophageal echocardiography, aortic diseases.

We report the case of a young female patient who presented with chest pain, having a background of systemic complaints and a history of essential thrombocytosis. Transesophageal echocardiography (TEE) was suggestive of an intramural hematoma of the distal ascending aorta and transverse arch, confirming computed tomography findings. However, surgery revealed only diffuse aortic wall thickening without hematoma or dissection, despite an epiaortic ultrasonogram corroborating the TEE findings. Histopathology demonstrated inflammation involving all layers of the aortic wall, with intimal fibrosis, disrupted elastic fibers in the media, and marked fibrous thickening of the adventitia with multiple necrobiotic foci; the latter were apparently the cause of the markedly hypodense region mimicking intramural hematoma on imaging. The findings were consistent with Takayasu's arteritis, a large vessel vasculitis of unknown etiology that predominantly affects females.

Manuscript received:
January 22, 2008;
Accepted:
June 4, 2008.

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Pain of aortic origin does not necessarily signify an aortic emergency, even in the presence of imaging findings suggestive of an acute aortic process. In this report, we describe the case of a young female patient who underwent emergency aortic surgery because of acute chest pain and imaging findings consistent with an intramural hematoma of the ascending aorta. However, histopathology demonstrated prominent inflammation of the aortic wall consistent with Takayasu's arteritis.

Case presentation

A 36-year-old Caucasian female was referred to our hospital for new-onset, intermittent chest pain and a discrete, hypodense region in the wall of the ascending aorta on an outside chest computed tomography (CT); this finding raised the question of acute aortic syndrome. The patient described multiple distinct episodes of severe

central chest pain radiating to the back over the last week; she also reported several episodes of profuse sweating and near syncope over the previous months. Her medical history featured essential thrombocytosis diagnosed 10 years before, currently treated with aspirin and hydroxyurea. The physical examination was unremarkable. She was in normal sinus rhythm without signs of ischemia or other ECG abnormalities. A routine laboratory workup was unremarkable apart from a platelet count of 876,000 / μ l. Transesophageal echocardiography (TEE) was performed in order to clarify the CT findings. The aorta measured normal in size at all levels. However, both long- and short-axis views of the aorta demonstrated localized thickening of the aorta with a thrombus-like appearance characterized by echolucent areas. These features were suggestive of a crescentic intramural hematoma of the distal ascending aorta and the transverse arch (Figure 1, A & B).

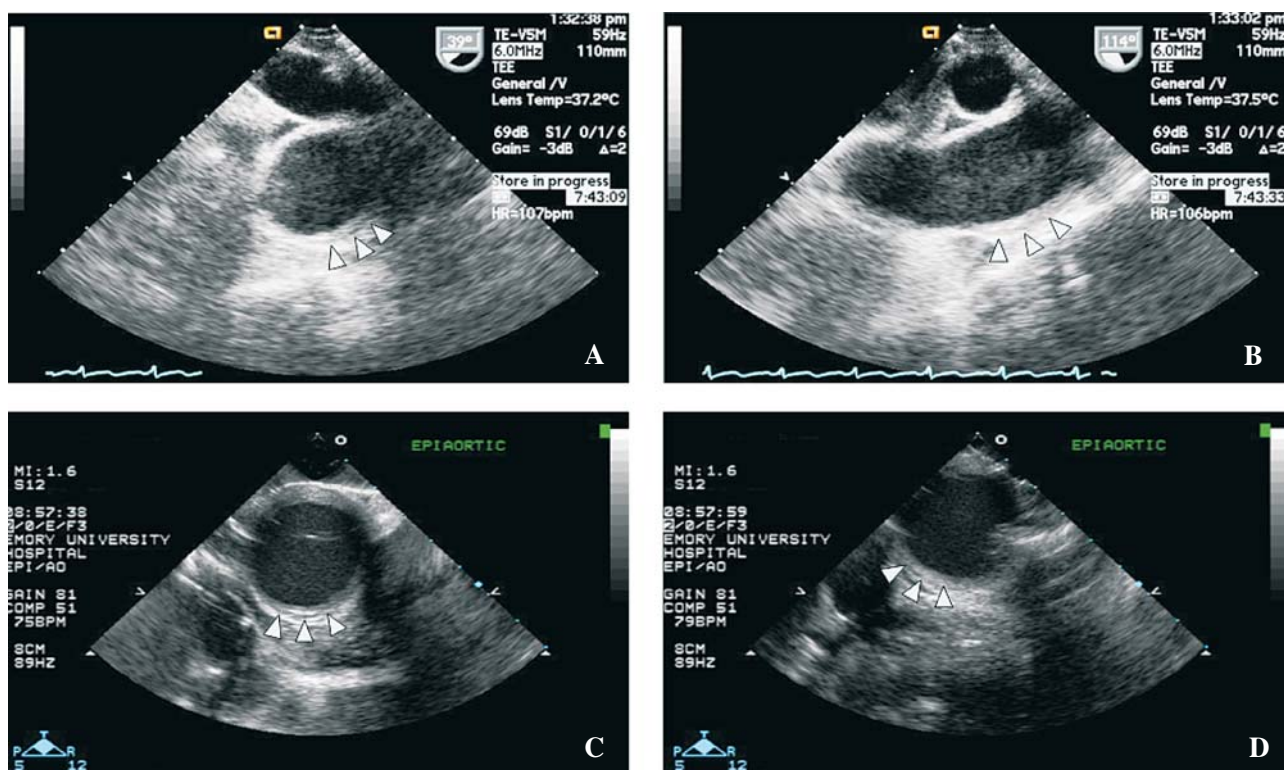


Figure 1. A & B: Transesophageal echocardiography images in the short (A) and long (B) axis at the level of the distal ascending aorta reveal areas of crescentic thickening with a thrombus-like appearance and distinct areas of echolucency (arrowheads) suggestive of intramural hematoma. C & D: Intraoperative epi-aortic scanning confirms the findings of transesophageal echocardiography.

Considering the acute prognosis of type A intramural hematomas,¹ surgery was deemed necessary at this point. In the operating room, while the aorta did not appear ecchymotic on visual inspection, an intraoperative epi-aortic ultrasonogram continued to show what appeared to be an intramural hematoma between the adventitia and the intima at the distal ascending aorta and the transverse arch (Figure 1, C & D). Surprisingly, subsequent division of the aorta revealed no hematoma but severe, diffuse thickening of the aortic wall, which was consistent with aortitis. Because of their abnormal appearance and concerns about future enlargement due to the vasculitic process, both the ascending aorta and the transverse arch were excised and replaced with synthetic grafts.

The histopathologic examination of the aortic wall specimens revealed an inflammatory process involving all three layers of the aortic wall, with fibrosis and collagenization of the intima, disrupted elastic fibers in the media, and marked fibrous thickening of the adventitia with multiple necrobiotic foci. The latter were apparently the cause of the markedly hypodense region mimicking intramural hematoma on

imaging (Figures 2, 3). These findings were consistent with Takayasu's arteritis.² The subsequent workup was also suggestive of an autoimmune process, featuring positive cytoplasmic antibodies and an elevated erythrocyte sedimentation rate (ESR: 81 mm/hr). Magnetic resonance angiography of the neck and head vessels did not reveal further abnormalities. The patient did well and was discharged on steroids.

Discussion

Takayasu's arteritis is an idiopathic systemic inflammatory disease involving mainly the large elastic arteries. This entity affects predominantly young women (female to male ratio ~8:1), with an incidence of 2-3 cases per million per year in North American populations.³ The process involves all layers of the arterial wall; although the pathogenesis is still unknown the majority of data point to an autoimmune etiology.² The disease has a biphasic course. The early systemic or 'pre-pulseless' phase is characterized by active inflammation and nonspecific signs and symptoms, including fever, malaise, weight loss, anorexia, night sweats, and occa-

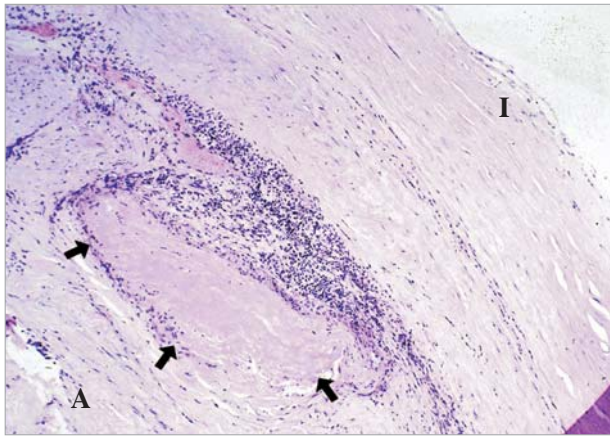


Figure 2. Necrotic area (arrows) surrounded by reactive infiltrating cells. H & E stain. I – intima; A – adventitia.

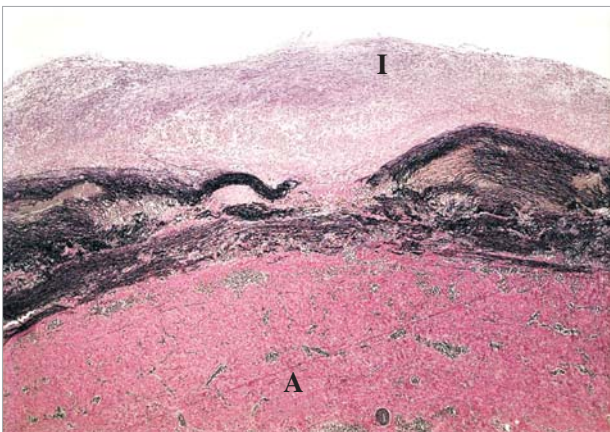


Figure 3. Full thickness of aortic wall reveals severe thickening of the intima (I), marked fragmentation and distortion of the smooth muscle cells and elastic lamellae of the media, as well as fibrosis and thickening of the adventitia (A). Within the latter, there are nests of inflammatory cells surrounding penetrating vessels. Elastic stain (Verhoeff).

sionally chest pain.⁴ Manifestations also include hypertension, bruits, headaches, postural dizziness, and syncope.⁴ The disease gradually progresses at variable rates to a late occlusive or ‘pulseless’ phase, which is characterized by intimal hyperplasia and fibrosis leading to obliterative luminal changes. The manifestations are largely dependent upon the site and grade of arterial lesion.

How is the clinician supposed to discern an inflammatory aortic process in an acute setting? Although ruling out or confirming the presence of life-threatening causes of chest pain is conceivably the main concern in similar situations, there are telltale signs that should be taken into account when evaluat-

ing the individual patient. For example, the presence of thrombocytosis has been recognized as part of several chronic inflammatory diseases, including Takayasu’s arteritis,⁵ although the mechanism by which thrombocytosis is induced is still under investigation. Obtaining an ESR preoperatively might have also raised the question of a systemic process, especially when considering the presence of systemic complaints that are common in these settings. On the other hand, a key feature of Takayasu’s arteritis, namely peripheral vascular signs (diminished pulses, bruits), was absent in the present case.

Importantly, involvement of the coronary circulation is not rare in Takayasu’s arteritis. Although the probability of underlying atheromatous coronary artery disease was considered negligible in the current case and not worth delaying surgical intervention, non-atheromatous coronary involvement was not excluded preoperatively. Moreover, pain originating from the aorta has been described in patients with impaired aortic elastic properties, without aortic dissection or other active intramural process.⁶ These possibilities should be considered in patients who complain of chest pain in the presence of atypical and/or systemic findings.

From an imaging standpoint, both CT and TEE are pivotal in the timely diagnosis of acute aortic syndromes. Indeed, TEE has the ability to discriminate among different variants of acute aortic syndromes, including intramural hematoma and penetrating atherosclerotic ulcer, both of which carry an adverse prognosis if left untreated.¹ However, the current case highlights a drawback of both these imaging modalities: the limitations in tissue characterization. Circumferential thickening of the thoracic aorta is a common TEE finding in Takayasu’s arteritis, affecting a significant proportion of segments.⁷ Moreover, adventitial and medial fibrosis along with focal destruction of the external elastic membrane and replacement by fibrous tissue are the rule in the chronic phase of the disease.² These ‘necrotic’ regions can be misdiagnosed as intramural aortic hematoma. Similar findings have been described in CT studies.⁸ On the other hand, newer modalities are promising in this direction. F¹⁸-fluorodeoxyglucose (FDG) positron emission tomography co-registered with enhanced CT has been reported to ‘trace’ the inflammatory process in patients with Takayasu’s arteritis,⁹ and delayed hyperenhancement MRI may also be useful in detecting disease activity in the arterial wall.¹⁰

In conclusion, certain imaging features of Takaya-

su's arteritis can resemble those of an aortic syndrome and lead to diagnostic dilemmas in the setting of acute chest pain. The presence of systemic complaints along with other clues pointing to an inflammatory process should alert the clinician towards this possibility, especially in young female patients who have no proatherosclerotic background or known aortic pathology.

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