

CASE REPORT

Exceptional cause of chest pain: idiopathic ascending aortitis

Göğüs ağrısının istisnai nedeni: İdiyopatik çıkan aortit

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Summary– Idiopathic ascending aortitis is an insidious disease most often encountered during an evaluation for other cardiovascular and connective tissue diseases. To the best of our knowledge, admission to an emergency department with chest pain due to idiopathic ascending aortitis has not been reported before. Herein, the case of a 46-year-old man with chest and back pain symptoms that were discovered to be secondary solely to idiopathic ascending aortitis is described.

Özet– İdiyopatik çıkan aortit genellikle kalp damar ve bağ dokusu hastalıklarının araştırılması sırasında saptanan sinisi bir hastalıktır. Bildiğimiz kadarıyla, idiyopatik çıkan aortite bağlı olarak göğüs ağrısı ile acil servise başvuru daha öncesinde bildirilmemiştir. Bu yazıda, göğüs ve sırt ağrısı semptomları sadece idiyopatik çıkan aortite sekonder olduğu farkedilen 46 yaşındaki erkek hasta sunuldu.

Acute chest pain is one of the leading causes of emergency department admissions. Unlike acute aortic syndromes, admission with chest pain due solely to aortitis is very rare. Aortitis is most often secondary to a specific cause; the remainder of cases constitute idiopathic aortitis, which is almost always asymptomatic. Presently described is a male patient with idiopathic aortitis, who was referred to the hospital due to chest pain that had started a month earlier and had intensified in the previous several days.

CASE REPORT

A 46-year-old man was admitted to the emergency department with ripping, knife-like chest and back pain that had started a month prior and had intensified in recent days. Apart from smoking, the patient did not have any other remarkable medical history. He did not describe arthralgia or weight loss. On physical examination, he was restless, tachypneic, and afebrile. His blood pressure was 135/55 mmHg on the left arm

and 138/60 mmHg on the right arm; pulse oximeter measured an oxygen saturation level of 97%. His body temperature was 36.6°C. Auscultation of the patient

revealed a diminished S1 and a loud S2; no additional sound was heard. Nothing unusual was detected on pulmonary auscultation. The temporal arteries were non-tender and exhibited palpable pulses. He did not describe new onset, localized headache. Brachial, radial, and dorsalis pedal pulses were intact. There were no carotid, subclavian, abdominal, or femoral artery bruits. An electrocardiogram revealed sinus tachycardia of 102 beats/minute without any ST segment changes in the isoelectric line.

Despite the normal pulmonary auscultation, a chest X-ray was obtained and helped to exclude pneumo-

Abbreviations:

CRP	C-reactive protein
CT	Computed tomography
ESR	Erythrocyte sedimentation rate
IgG	Immunoglobulin G
MRI	Magnetic resonance imaging
TEE	Transesophageal echocardiography
WBC	White blood cell

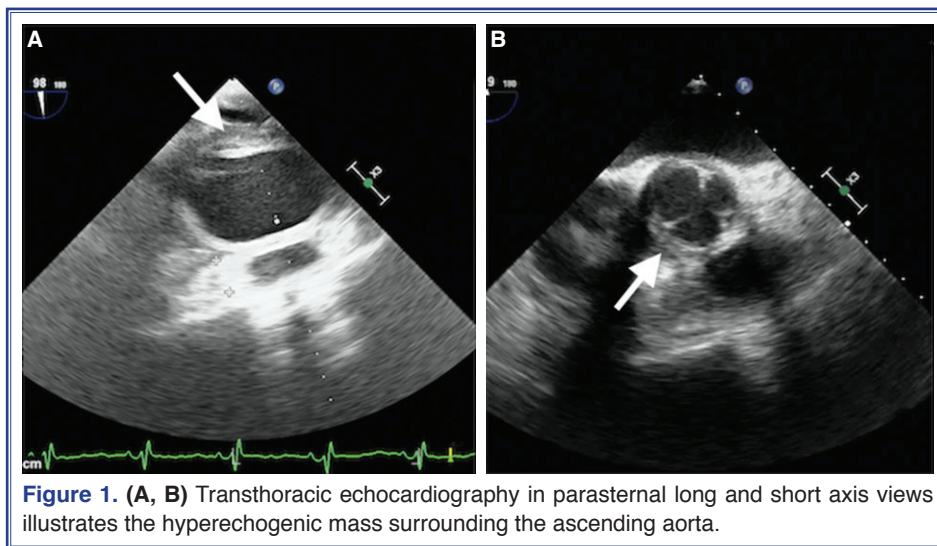
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thorax and pneumonia. The laboratory results included: troponin I level of 0.1 ng/mL (reference: 0–0.04 ng/mL), D-dimer of 952 ng/mL (reference: 0–500 ng/mL), C-reactive protein (CRP) of 7.2 mg/L (reference: 0–4.9mg/L), erythrocyte sedimentation rate (ESR) of 32 mm/hour (reference: 0–15 mm/hour), and a white blood cell (WBC) count of 13,500/mm³ (reference: 3,500–10,500/mm³). The patient was referred to the cardiology department with a primary diagnosis of acute coronary syndrome. A bedside transthoracic echocardiography (TTE) was performed, providing an ejection fraction of 65% and ascending aorta measurement of 38 mm. The right chambers were of standard dimensions. A hyperechogenic mass encircling the ascending aorta was detected on parasternal long and short axis views (Figure 1a, b). The maximal thickness of the ascending aorta wall was measured as 14 mm. Contrast-enhanced thorax computed

tomography (CT) was performed in order to exclude aortic syndromes. No significant pathological lymph node was detected in the mediastinum. Diffuse isohypodensity surrounding the thoracic ascending aorta with a maximal thickness of 14 mm was detected in the thorax CT (Figure 2a-c). The main branches of the aorta appeared to be normal on CT angiography. The pulmonary arteries and main branches were also intact. Troponin I values obtained 3 hours apart were measured as 0.09 ng/mL and 0.1 ng/mL, respectively. The patient was hospitalized in the internal medicine service for further evaluation after relieving his pain slightly with nonsteroidal anti-inflammatory drugs. A coronary angiography was performed in order to rule out coronary artery stenosis and embolization, and revealed normal coronary arteries. Transesophageal echocardiography (TEE) was performed and demonstrated aortitis with 14-mm wall thickness (Figure 3a-



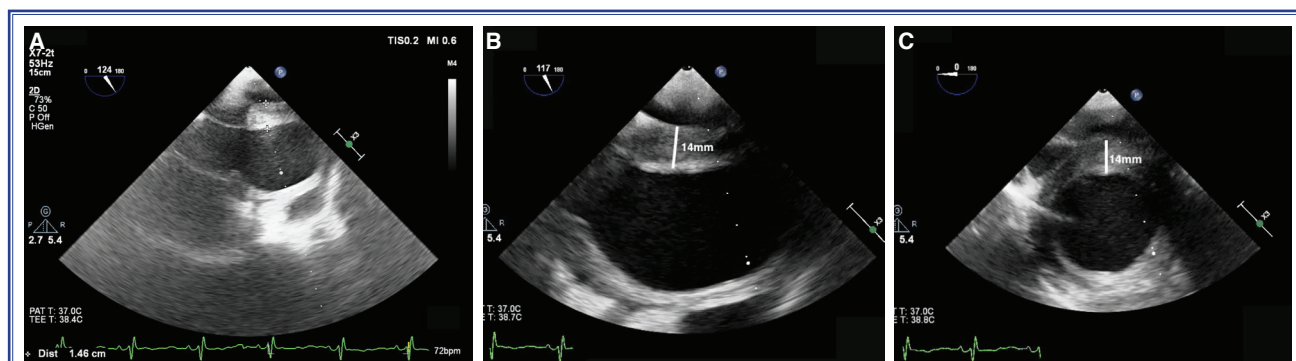


Figure 3. (A-C) Transesophageal echocardiographic images of the aortitis with 14-mm wall thickness.

c). Contrast-enhanced thoracic magnetic resonance imaging (MRI) was performed in order to clarify the density. Homogeneously hypointense tissue surrounding the ascending aorta was observed on MRI (Figure 4a, b). Infectious and noninfectious causes of ascending aortitis were further investigated. Three sputum cultures were negative for active tuberculosis. For latent tuberculosis infection, QuantiFERON-TB Gold In-Tube test (Quiagen, Hilden, Germany) was used as a kind of interferon-gamma release assay, and found to be negative. Findings of a rapid plasma reagin test and a fluorescent treponemal antibody absorption assay were also both negative. He did not have any of the typical clinical features or radiological criteria of Takayasu arteritis or giant cell arteritis.^[1] There was no need to perform a temporal artery biopsy as none of the other diagnostic criteria were fulfilled.^[2] Serum immunoglobulin G (IgG) and IgG subgroup levels were within the normal limits. The absence of clinical features and negative autoimmune markers excluded the possibility of connective tissue disorders. WBC count and CRP did not increase during follow-up, and

there were repeated negative blood cultures. These circumstances indicated idiopathic ascending aortitis.

The administration of 0.5 mg/kg daily intravenous prednisone dramatically relieved the disturbing back pain. The patient was discharged with oral prednisone treatment and was asymptomatic for 3 months. At the 3-month follow-up visit, a contrast enhanced CT was performed and similar findings were detected. ESR was 12 mm/hour. Azathioprine was recommended in addition to prednisone treatment because of the CT findings; however, patient refused to be on regular medication. The oral prednisone treatment was gradually terminated and the patient began monthly follow-up visits with the cardiology and rheumatology departments.

DISCUSSION

The most common causes of aortitis are noninfectious inflammatory vasculitis, giant cell arteritis, and Takayasu arteritis.^[3] The prevalence of aortitis among patients undergoing resection of the ascending aorta

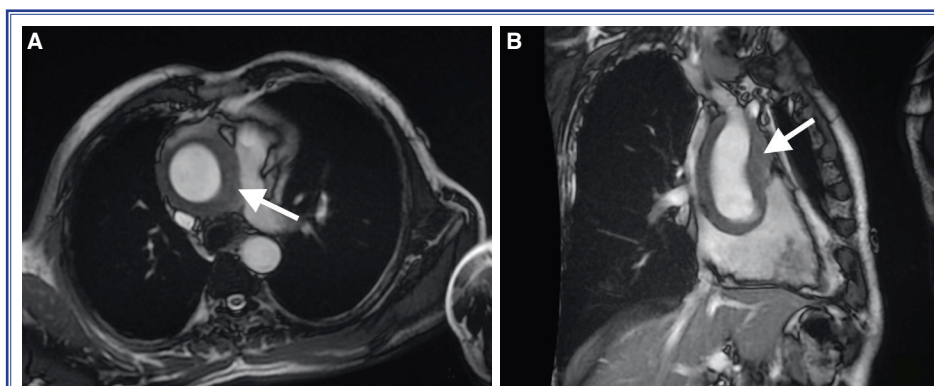


Figure 4. (A, B) Thoraco-abdominal contrast enhanced magnetic resonance image illustrating the homogeneously hypointense tissue surrounding the ascending aorta.

has been reported to be 4.3% to 8.8%.^[4,5] The prevalence of aortitis is thought to perhaps be greater than has been reported in studies of patients undergoing aortic surgery. Aortitis may be underdiagnosed for several reasons: The disease course can be asymptomatic for a long time, and patients are often diagnosed when complications occur, typically in the form of aortic aneurysms requiring surgery.^[6] Therefore, our case is important because the patient was admitted to the emergency department with augmented chest pain without complication. Idiopathic aortitis can be the underlying cause of chest and back pain treated in emergency departments. An unexplained rise in the troponin I level accompanied by chest pain guided the diagnosis.

TTE is a simple diagnostic modality that can easily be performed in arriving at the definitive diagnosis of chest pain. It plays a major role in determining acute aortic syndromes.^[7] TTE enhances the accuracy of TTE in the differential diagnosis of aorta diseases. TTE together with TEE properly designates aortic valve endocarditis and the ascending aorta wall, as documented in this case report. The ascending aorta wall was measured at 14mm by both TTE and TEE. The most thickened idiopathic ascending aorta wall was 15 mm, which also concomitantly had an ascending aortic aneurysm.^[8] In our case, early diagnosis was made based on chest pain and increased troponin level, which directed us to further evaluation.

When evaluating the aorta with CT, an unenhanced acquisition is important for the diagnosis of intramural hematoma. The combination of an unenhanced and contrast-enhanced acquisition offers very high sensitivity for the diagnosis of intramural hematoma.^[9] In our case, we did not detect intramural hematoma. In cases of type A intramural hematoma, urgent surgery is indicated. Another important disease to be considered in the differential diagnosis is retrograde dissections localized in the descending thoracic aorta and the abdominal aorta. These emergent diagnoses are important with respect to the treatment choice. In our case, the abnormality was limited to the ascending aorta, which was surrounded by a thickened wall.

The diagnosis of idiopathic aortitis is difficult because it is based on excluding the other probable diagnoses. The mortality rate is high in infectious aortitis patients, and if left untreated, the disease is fatal.^[10] Thus, if suspected, treatment with broad spectrum

antibiotics should be initiated as soon as possible.^[11] Our patient's main complaint was chest and back pain, which could not be explained by uncomplicated aortitis. The clinical status was also unlike that of bacteremia or infectious aortitis. Further laboratory investigations are also important to draw a clear line between infectious and noninfectious aortitis since the treatment strategy is completely different. Cases of multiple mycotic aneurysms, which we did not detect in our patient, have also frequently been observed in infectious aortitis.^[11]

The treatment strategy in idiopathic aortitis is based on immunosuppressive therapy with corticosteroids.^[12] An adequate response to treatment verifies the strategy. In addition to monitoring clinical symptoms, ESR is a good follow-up parameter.^[12]

Conclusion

Idiopathic ascending aortitis can be an underlying cause of chest pain. The definitive diagnosis is based on excluding other possible causes using diagnostic modalities, such as a chest X-ray, TTE, TEE, and CT. Immunosuppressive drugs, such as corticosteroids, play an important role in the treatment of idiopathic ascending aortitis.

Conflict-of-interest: None declared.

REFERENCES

1. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. *Arthritis Rheum* 1990;33:1129–34. [\[CrossRef\]](#)
2. Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. *Arthritis Rheum* 1990;33:1122–8. [\[CrossRef\]](#)
3. Evans JM, O'Fallon WM, Hunder GG. Increased incidence of aortic aneurysm and dissection in giant cell (temporal) arteritis. A population-based study. *Ann Intern Med* 1995;122:502–7. [\[CrossRef\]](#)
4. Rojo-Leyva F, Ratliff NB, Cosgrove DM 3rd, Hoffman GS. Study of 52 patients with idiopathic aortitis from a cohort of 1,204 surgical cases. *Arthritis Rheum* 2000;43:901–8.
5. Miller DV, Isotalo PA, Weyand CM, Edwards WD, Aubry MC, Tazelaar HD. Surgical pathology of noninfectious ascending aortitis: a study of 45 cases with emphasis on an isolated variant. *Am J Surg Pathol* 2006;30:1150–8. [\[CrossRef\]](#)
6. Schmidt J, Sunesen K, Kornum JB, Duhaut P, Thomsen RW. Predictors for pathologically confirmed aortitis after resec-

- tion of the ascending aorta: a 12-year Danish nationwide population-based cross-sectional study. *Arthritis Res Ther* 2011;13:R87. [\[CrossRef\]](#)
7. Sobczyk D, Nycz K. Feasibility and accuracy of bedside transthoracic echocardiography in diagnosis of acute proximal aortic dissection. *Cardiovasc Ultrasound* 2015;13:15.
 8. Yoon J, Gruboy IP, Zaid A, Salil SG, Connolly M, Aronow WS, et al. Isolated idiopathic aortitis with an unusually thickened aortic wall: case report. *Ann Transl Med* 2016;4:428.
 9. O’Gara PT, DeSanctis RW. Acute aortic dissection and its variants. Toward a common diagnostic and therapeutic approach. *Circulation* 1995;92:1376–8. [\[CrossRef\]](#)
 10. Gornik HL, Creager MA. Aortitis. *Circulation* 2008;117:3039–51. [\[CrossRef\]](#)
 11. Lopes RJ, Almeida J, Dias PJ, Pinho P, Maciel MJ. Infectious thoracic aortitis: a literature review. *Clin Cardiol* 2009;32:488–90. [\[CrossRef\]](#)
 12. Mukhtyar C, Guillevin L, Cid MC, Dasgupta B, de Groot K, Gross W, et al; European Vasculitis Study Group. EULAR recommendations for the management of large vessel vasculitis. *Ann Rheum Dis* 2009;68:318–23. [\[CrossRef\]](#)
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