ANIL AGGRAWAL'S
Internet Journal of Forensic Medicine and Toxicology
The world's first online journal devoted entirely to forensic medicine and toxicology · Published 1 January and 1 July

VOL. 16, NO. 1 · JANUARY – JUNE 2015
Downloaded from www.anilaggrawal.com · ISSN 0972-8074 (Online) · https://www.anilaggrawal.com/ij/vol_016_no_001/papers/paper002.html

An Autopsy Case of Hemopericardium Caused by Ascending Aortic Rupture Associated with Idiopathic Aortitis.

Hiromitsu Kurata[a, b, c], Akiko Ishigami[a, b], Itsuo Tokunaga[a], Yasushi Nagasaki[b], Akiyoshi Nishimura[a, b, *]

[a]Department of Forensic Medicine, Institute of Health Biosciences, The University of Tokushima Graduate School, 3-18-15 Kuramotocho Tokushima 770-8503, Japan
[b]Hyogo Medical Examiner's Office, 2-1-31 Aratacho Hyogoku Kobe 652-0032, Japan
[c]Department of Neurosurgery, Mima Hospital, 497 Jogeshima Kamojimacho Yoshinogawa Tokushima 776-0013, Japan

Contact: A. Nishimura · ncc1701abcde@basic.med.tokushima-u.ac.jp

CITE THIS ARTICLE

Kurata H, Ishigami A, Tokunaga I, Nagasaki Y, Nishimura A. An Autopsy Case of Hemopericardium Caused by Ascending Aortic Rupture Associated with Idiopathic Aortitis.. *Anil Aggrawal's Internet Journal of Forensic Medicine and Toxicology* [serial online], 2015; Vol. 16, No. 1 (January – June 2015): [about 12 p]. Available from: https://www.anilaggrawal.com/ij/vol_016_no_001/papers/paper002.html

ABSTRACT

We report the case of a 92-year-old Japanese woman who suddenly died of rupture of the ascending aorta following hemopericardium due to idiopathic aortitis. Rupture of the ascending aorta is usually caused by trauma, an aortic aneurysm, aortic dissection, infection, or a neoplasm, but no such etiologies were found in this case. The pathogenesis of the rupture of the aorta in the present case might have involved the destruction of elastic and collagen fiber due to lymphoplasmacytic infiltration through idiopathic aortitis and increased vulnerability of aortic wall following rupture. Aortitis is usually the cause of aortic aneurysm or dissection, however, aortic perforation is very rare. The frequency of sudden death due to this condition might be much higher than has been reported because cases in which no autopsy was performed would have been classified as unknown acute cardiac death. In cases involving sudden death, we have to keep in mind that the aorta can rupture in the absence of trauma or other typical etiologies.

Keywords: Forensic pathology, Idiopathic aortitis, Hemopericardium, Spontaneous rupture, Ascending aorta

Introduction

Hemopericardium due to rupturing of the ascending aorta is usually caused by trauma, an aortic aneurysm, aortic dissection, infection of the aortic wall, or tumoral invasion of the aortic wall. We describe a very rare case of hemopericardium caused by rupture through idiopathic aortitis of the ascending aorta and discuss its pathogenesis.

Case report

A 92-year-old Japanese woman was found dead on her bed by her son. She had a history of hypertension, atrial fibrillation, and angina pectoris and had been taking aspirin and warfarin. She had no other clinical symptoms and had not required nursing care during her daily life.

Autopsy findings

The subject was 145.0cm in height and weighed 57.2kg. Moderate rigor mortis was observed in all joints, and severe livor mortis was also present although it was easily displaced by digital pressure. Her rectal temperature was 31.0°C (ambient temperature: 22°C). The bilateral bulbar conjunctivas were hyperemic, and many petechiae were observed. The bilateral corneas were mildly turbid. No injuries were observed.

Her heart weighed 342g (normal weight [1]: 327.0+/-49.3g) and contained 200ml of dark red fluid blood. In addition, 190ml blood containing soft clots had accumulated in the pericardial space (Fig. 1A). No perforations or necrotic changes were found in the cardiac wall. The ascending aorta contained a perforation at 2cm distal from the aortic valve together with a subadventitial hematoma that measured 1.3cm in length (Fig. 1B). The aorta did not display any dissection or aneurysms, but did exhibit macroscopic atherosclerotic changes without ulceration or plaque at perforating site. All of the coronary arteries showed moderate atherosclerotic changes but remained patent. No myocardial degeneration or scar formation was found. Her other organs were congested, but did not display any remarkable findings.



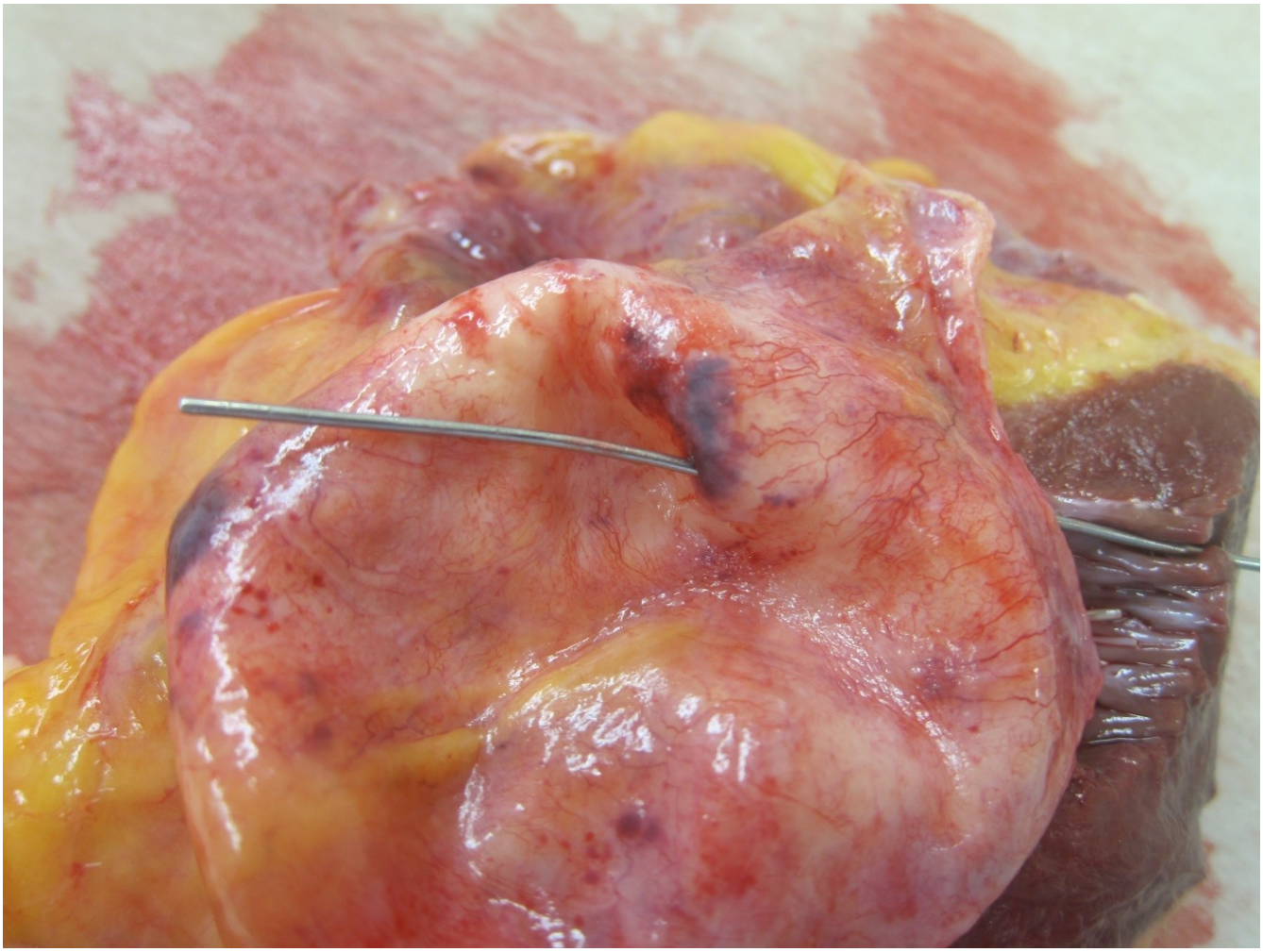
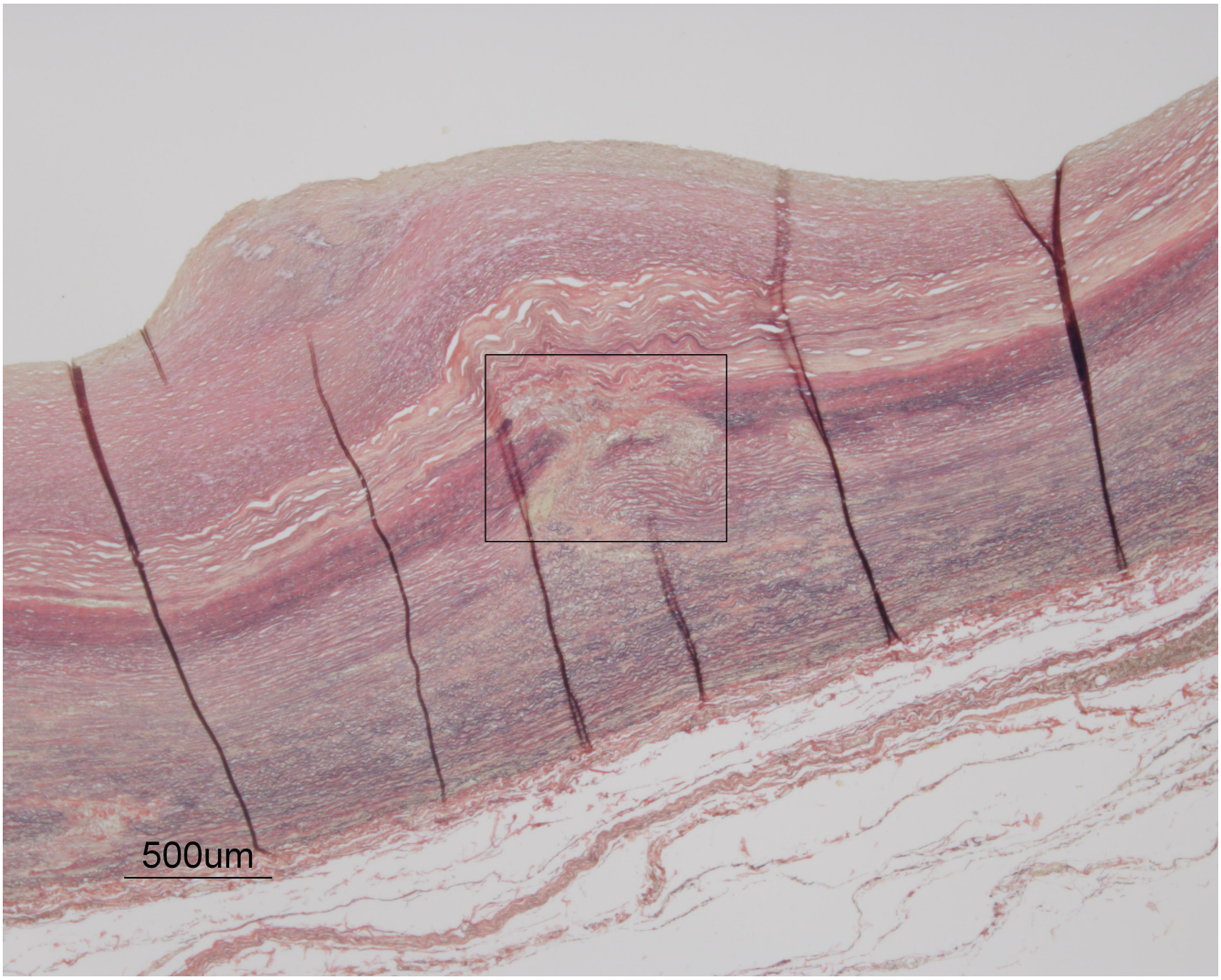


Fig. 1 Hemopericardium containing fluid blood mixed with soft clots is visible within the pericardial cavity (A). Perforation accompanied by intramural hematoma formation visualized on the aortic wall 2 cm distal to the aortic valve (B).

Histological findings

Histological examination of sections stained with Elastica van Gieson (EVG) showed rupture of the elastic fibers and collagen fibers constituting the internal elastic lamina of the aortic wall (Fig. 2A). Examination of HE-stained sections revealed sporadic infiltration by lymphocytes and plasmocytes at the sites of rupture of the elastic fibers, partially accompanied by bleeding, although polynucleated giant cells were absent (Fig. 2B). On the other hand, examination of EVG-stained sections did not reveal lymphocyte or plasmocyte infiltration at the site of rupture of the elastic fibers of the internal elastic lamina, although partial scarring was noted. The tunica intima was thickened, showing some atherosclerotic changes (e.g., hyalinization). There was no evidence of vasculitis in any other organs.


500um

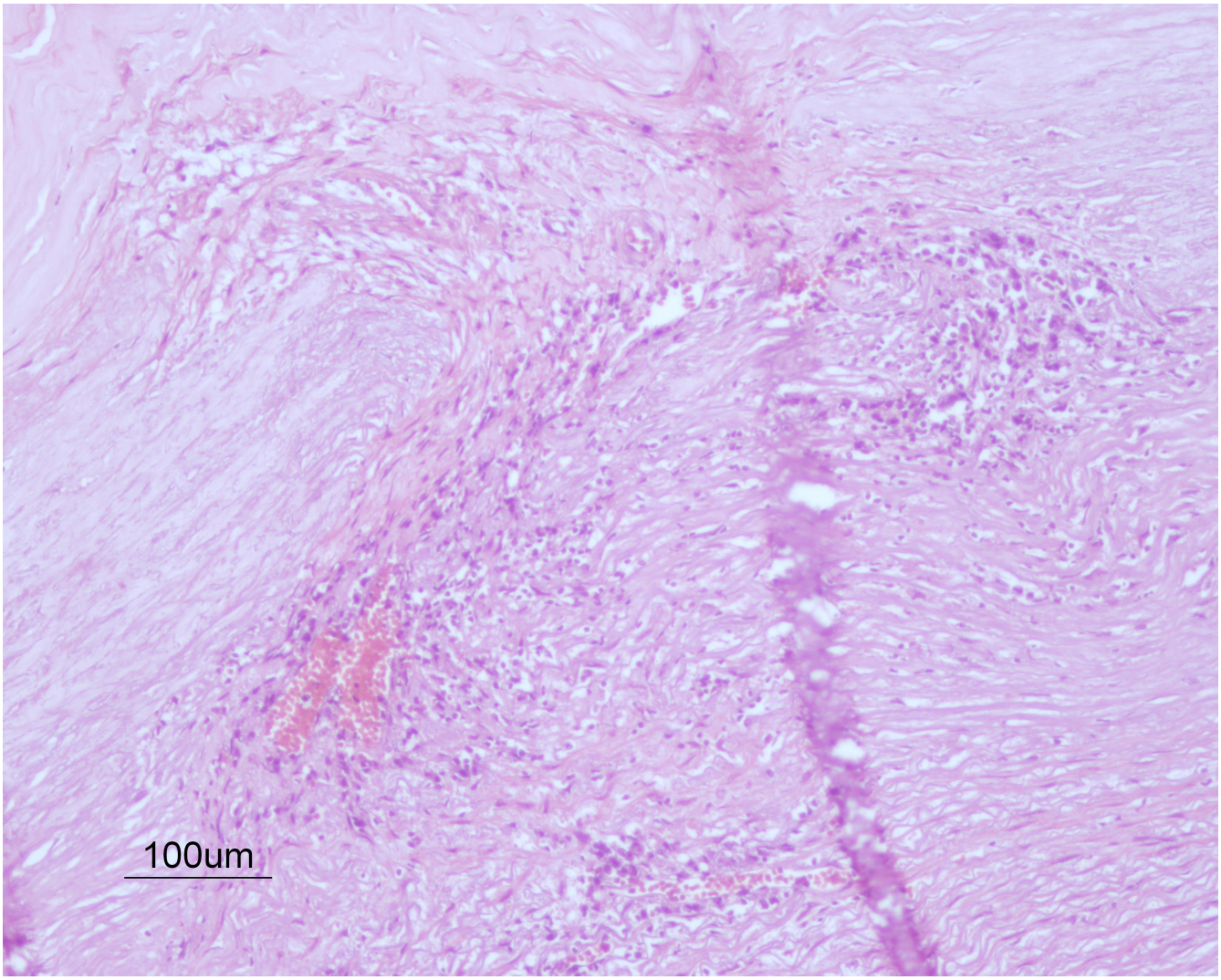


Fig. 2 Rupture of the internal elastic lamina of the aorta revealed by Elastica van Gieson staining, with destruction of the elastic fibers being a predominant change (A). Hematoxylin-Eosin staining of square site of Fig. 2(A) showing lymphocyte and plasma cell infiltration and bleeding around the destroyed elsatic fibers (B).

Discussion

Aortitis is pathologically defined as inflammation of the aortic wall [2]. However, there is no uniform clinical definition or classification of vasculitis, including aortitis, and various syndromes have been referred to under the heading of vasculitis, depending on the site affected, the underlying disease, etc. According to the CHCC2012 adopted at Chapel Hill in 2012 to establish a uniform definition for vasculitis, vasculitis involving the aorta includes that associated with systemic diseases such as giant cell arteritis (GCA), Takayasu's arteritis, rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), and Behcet's disease [3].

In the present case, there was no history of systemic diseases such as RA, and it was possible to rule out systemic disease as the cause of the aortitis. GCA more commonly occurs in elderly people; however, it is characterized histologically by the presence of polynucleated giant calls and granulomas, differing from the histological findings in the present case. Takayasu's arteritis appears more frequently in younger persons, and differs from the present case in that it is

characterized by the involvement also of branching blood vessels other than the aorta. Thus, the vasculitis seen in the present case did not satisfy any of the definitions adopted by CHCC2012.

Recently, the new disease entity “idiopathic aortitis” has been proposed, characterized by evidence of non-infectious aortitis in the absence of underlying vascular diseases such as GCA or Takayasu’s arteritis, or indeed any multi-system diseases such as RA, SLE, or Behcet’s disease [2, 4]. Idiopathic aortitis confined to the thoracic aorta is often not associated with any clinical manifestations of inflammation and is detected accidentally based on the detection of lymphoplasmacytic infiltration of the aortic wall in histopathological specimens of the aorta obtained at the time of surgical resection for thoracic aortic aneurysm, etc.[2]. In the present case, a diagnosis of idiopathic aortitis was made on the basis of the histopathological findings in the absence of any underlying diseases and any evidence of vasculitis involving other blood vessels.

In a previous study carried out to determine the incidence of aortitis, the complication of thoracic aortic aneurysms by aortitis was reported. Of the 70638 individuals in Malmö City (Sweden), with a population of approximately 240,000, who died during the 27-year period from 1958 to 1985, autopsy was performed in 85% and the causes of death were analyzed [5]. Among these, the complication of thoracic aortic disease by aortitis was seen in 9 of the 216 patients with fatal thoracic aortic dissection, 8 of the 63 patients with ruptured thoracic aortic aneurysm, and 22 of the 205 patients with asymptomatic thoracic aortic aneurysm. Thus, evidence of aortitis was found in 39 (8.1%) of the 484 cases of thoracic aortic dissection or aneurysm.

In addition, another study of aortitis in cases operated for thoracic aortic aneurysm has been reported [6]. According to this report, aortitis was detected in 39 (4.9%) of 788 cases of thoracic aortic aneurysm; the condition was non-infectious in 38 of the 39 cases (4.8%). The underlying disease was GCA in 30 cases, inflammatory aneurysm in 3 cases, aortitis accompanied by non-specific lymphocyte/plasma cell infiltration in 2 cases, Takayasu’s arteritis in 1 case, SLE in 1 case, and Behcet’s disease in 1 case. Of these cases, cases with aortitis characterized by the histological findings of non-specific lymphoplasmacytic infiltration were considered to fall under the definition of idiopathic aortitis, seen at an estimated frequency of 0.3% of all cases with thoracic aortic aneurysm and 5.1% of all cases with aortitis.

Although aortitis is known to be associated frequently with aortic aneurysm or dissection, aortic rupture in the absence of aneurysm in such cases is considered to be rare, with only one such case of rupture associated with GCA having been reported [2, 7]. Hemopericardium due to perforation of the ascending aorta usually develops as a complication of rupture of an aortic aneurysm or aortic dissection, infection or neoplasm, other than the cases attributable to trauma; rare causes include penetrating atherosclerotic ulceration (PAU), a disease entity proposed in 1986 by Stanson et al, and spontaneous rupture. In cases of PAU, ulceration in an atheroma of the tunica intima can occur, causing perforation of the internal elastic lamina, and if it spreads to the tunica media, loading of the tunica media by the arterial pressure would eventually lead to the formation of an intramural hematoma and dissection. Occasionally, perforation of the internal elastic lamina does not spread to the tunica media, but directly spreads to the tunica externa, causing perforation [8, 9]. Some cases are reported as cases of spontaneous rupture; however, the vascular findings in such cases include signs of atherosclerosis and cystic necrosis of the tunica media, indicating the necessity of discussing their possible association with PAU [10, 11]. In the present case, macroscopic signs of atherosclerosis were noted and perforation due to PAU could not initially be ruled out; however, no ulceration was found at perforation site of aorta, and the histopathological finding of lymphocyte

and plasmocyte infiltration of the aortic wall allowed precise differential diagnosis as idiopathic aortitis. In other words, the definite cause of aortic rupture cannot be identified solely on the basis of the macroscopic findings at autopsy, without histopathological examination.

Patients developing hemopericardium due to thoracic aortic perforation are more likely to undergo autopsy from the standpoint of forensic medicine, because they often die suddenly without any precursor signs. The percentage of cases autopsied among all cases with this condition is particularly low in Japan and death from this disease is counted as sudden cardiac death. Even if hemopericardium can be confirmed by post-mortem CT scanning, there are diverse causes for this condition, including trauma, infection, aortic aneurysm, aortic dissection, etc., this condition, although rarely, can arise from aortitis, like in the present case. So, active autopsy and histopathological examination seem to be desirable to identify the cause of death in cases like this one.

References

1. Investigation committee of the Medico-Legal Society of Japan. Reports on medico-legal data from massive investigation performed by the Medico-Legal Society of Japan – weight and size of internal organs of normal Japanese today (in Japanese). Jpn J Legal Med 1992. p.335-99
2. Gornik HL, Creager MA. Aortitis. Circulation. 2008;117:3039-3051 PMID: 18541754
3. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, Flores-Suarez LF, Gross WL, Guillevin L, Hagen EC, Hoffman GS, Jayne DR, Kallenberg CGM, Lamprecht P, Langford CA, Luqmani RA, Mahr AD, Matteson EL, Merkel PA, Ozen S, Pusey CD, Rasmussen N, Rees AJ, Scott DGI, Specks U, Stone JH, Takahashi K, Watts RA. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum. 2013; 65(1):1-11 PMID: 23045170
4. Schmidt J, Sunesen K, Kornum JB, Duhaut P, Thomsen RW. Predictors for pathologically confirmed aortitis after resection of the ascending aorta: A 12-year Danish nationwide population-based cross-sectional study. Arthritis Research & Therapy 2011, 13:R87 PMID: 21676237
5. Svensjo S, Bengtsson H, Bergqvist D. Thoracic and thoracoabdominal aortic aneurysm and dissection: an investigation based on autopsy. British Journal of Surgery 1996;83:68-71 PMID: 8653370
6. Pacini D, Leone O, Turci S, Camurri N, Giunchi F, Martinelli GN, DiBartolomeo R. Incidence, etiology, histologic findings, and course of thoracic inflammatory aortopathies. Ann Thorac Surg 2008;86:1518–23 PMID: 19049742
7. Eklund E, Englund E, Valdemarsson S. Rupture of a non-aneurysmatic aortic trunk in a patient with giant cell arteritis. Ann Rheum Dis 1998;57:422-423 PMID: 9797576
8. Stanson AW, Kazmier FJ, Hollier LH, Edwards WD, Pairolero PC, Sheedy PF. Penetrating atherosclerotic ulcers of the thoracic aorta: natural history and clinicopathologic correlations. Ann Vasc Surg 1986;1:15-23 PMID: 3504683
9. van Driel EM, Klein P, de Jonge E, Arbous MS. Pericardial tamponade caused by a perforating atherosclerotic ulcer of the ascending aorta. Neth J Crit Care 2010;14(6):395-8
10. Akashi H, Tayama K, Otsuka H, Tobinaga S, Aoyagi S. Spontaneous rupture of the ascending aorta: case report and review. Circ J 2003;67:461-3 PMID: 12736488
11. Kashiyama N, Kubota Y, Nishikawa D, Izutani H. A case of spontaneous rupture of the ascending aorta. Jpn J Cardiovasc Surg 2010;39:45-8