

Inflammatory Aneurysm of Ascending Aorta: a case report

Amanda Stella (✉ as247226@pcom.edu)

Albert Einstein Medical Center <https://orcid.org/0000-0001-9049-892X>

Alexandra Tuluca

Albert Einstein Medical Center

Anastasia Arce

Albert Einstein Medical Center

Louis Samuels

Albert Einstein Medical Center

Research Article

Keywords: Inflammatory aneurysm, ascending aorta, aortitis

Posted Date: June 23rd, 2022

DOI: <https://doi.org/10.21203/rs.3.rs-1353635/v1>

License: © ⓘ This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

Abstract

Background: Inflammatory aortic aneurysms are rare and typically located in the infra-renal abdominal aorta. It is even more uncommon for this disease process to occur in the ascending aorta.

Case presentation: We report the successful case of a 42-year-old female with an inflammatory aneurysm of the ascending aorta and unusual left anterior descending coronary artery (LAD) inflammation.

Conclusions: The underlying etiology of severe inflammatory ascending aortic aneurysms in a previously healthy individual remains unknown. High surgical mortality and poor pre-operative diagnostic tools prompts further investigation to improve outcomes.

Background

Inflammatory aortic aneurysms are rare and typically located in the infra-renal abdominal aorta. It contributes to less than 5% of abdominal aortic aneurysms.¹ In 1972, Walker et al was the first to expound on this discrete entity which has significant gross pathologic features including wall thickening with extensive fibrous adhesions.² There are distinct histopathologic differences as well including marked destruction of media with heterogenous lymphoplasmacytic infiltrate and rare giant cells. Although they behave like their counterparts in the abdomen, inflammatory aortic aneurysms of the ascending aorta are even more uncommon. In 1994, Connery et al was the first to report an inflammatory aneurysm in the ascending thoracic aorta; however, this patient died shortly post-operatively.³ Also, there are no reports of this inflammatory process extending into coronary vessels. We report a successful case of an inflammatory aneurysm of the ascending aorta with comparable inflammation of the left anterior descending coronary artery (LAD) in a young female patient.

Case Presentation

A 42-year-old female with no significant past medical history presented with worsening shortness of breath and chest pain. She has a significant family history of cardiac disease consisting of coronary artery disease (CAD) and valvular disease in her mother, CAD in her father, and two sisters both with congestive heart failure. She had been admitted one month prior for sharp, midsternal chest pain that radiated to the right arm, new onset dyspnea with minimal activity and ankle edema. At that time, a myocardial infarction had been ruled out as the electrocardiogram and troponin was normal three times. A Transthoracic echocardiogram (TTE) showed the aortic root to be 3.5 cm with a normal ejection fraction greater than 55%. A Computed tomography (CT) was performed which identified an aneurysmal dilatation of the mid ascending aorta measuring 5.1 cm with extensive calcifications seen along the LAD and right coronary artery concerning for extensive atherosclerosis. The follow-up visit one month later consisted of a TTE that identified a dilated aortic root at 4.1 cm and a CT Angiography that identified a 5.7 cm fusiform aneurysm of the ascending thoracic aorta with a suspected intramural hematoma (Fig.

1). Additionally, Left Heart Catheterization identified the LAD to have a mid-saccular aneurysm which was immediately preceded by a severe 85% eccentric lesion and followed by a second 70% stenosis (Fig. 2).

In the operating room, a large inflammatory-appearing aneurysm was identified in the ascending aorta which consisted of a shiny, eggshell appearing glistening capsule. The thick wall appeared pink-yellow and had areas of white-tan fibrosis and calcifications (Fig. 3A). A similar inflammatory, thickened plaque was found overlying the mid-LAD in the area of the presumed aneurysm (Fig. 3B). The aneurysm plaque extended to just proximal to the innominate artery (IA) branchpoint. Therefore, standard aortic cannulation was not possible as there was not enough mobility due to severe tissue inflammation. Interestingly, the patient had a bovine arch anatomy with the left carotid artery (LCA) branching off a common trunk with the IA, which was carefully avoided. A partial occluding clamp was placed on the IA distal to the LCA takeoff. Central cannulation was achieved with a graft to the IA for arterial inflow. Standard cannulation followed with dual-stage venous return through the right atrial appendage. Cerebral protection was achieved by modified deep hypothermic circulatory arrest with antegrade cerebral perfusion via the innominate artery. The ascending aorta and the proximal hemiarch aneurysm were resected and replaced with a 26 mm Gelweave Graft (Terumo Aortic, Sunrise, FL). A coronary artery bypass graft was performed bypassing with LAD with the left internal mammary artery. The patient was extubated shortly after the operation, and the remainder of the postoperative course was uneventful.

Pathologic evaluation of the aneurysm revealed aortitis consisting of lymphoplasmacytic infiltrate with patchy acute inflammation, destruction of the media, and edema with a rare giant cell. An elastic stain also demonstrated the destruction of elastin fibers in the area of inflammation. Bacterial, fungal and treponemal stains were negative. Testing for SARS CoV-2 RNA was also negative.

Discussion And Conclusions

Inflammatory aortic aneurysms comprise a minor subset of aortic aneurysms. Although they are mostly seen in the infrarenal abdominal aorta, there have been cases reported where it occurs in the thoracic aorta as well. Connery et al. described the first inflammatory aneurysm of the ascending aortic in 1994.³ Since then, only twelve other case studies have been reported, most of which consisted of individuals over the age of 54 or with related inflammatory diseases.^{1, 4} Other studies have described incidental findings of aortitis upon retrospective pathology reviews of aortic surgical specimens, however, the severity of inflammation in this case is incomparable.⁵ Also, most cases consisted of elderly individuals with related comorbid diseases including giant cell arteritis and other rheumatologic diseases. A severely inflamed ascending aortic aneurysm with the inflammation extending into the LAD in a young female with no related co-morbid diseases is a medical phenomenon that has never been documented.

The cause of inflammatory aortic aneurysms is still unclear. Inflammatory diseases of the aorta are thought to be either infectious or non-infectious. Because of the negative bacterial, fungal and treponemal stains, the infectious forms are ruled out in our patient. The role of a viral infection involving SARS CoV-2 was postulated but also found to be noncontributory. The differential diagnosis for a non-

infectious aortitis could be due to an autoimmune response against atherosclerosis.⁶ Being that this inflammatory process occurred in the occluded LAD as well, it could be postulated that our patient had an autoimmune reaction against the atherosclerotic plaques. Further studies to diagnose an underlying inflammatory, rheumatologic, or autoimmune etiology are required to expound on this phenomenon. Other noninfectious inflammatory diseases have been postulated to be the underlying cause of thoracic aortitis including Takayasu arteritis, granulomatosis with polyangiitis and sarcoidosis.⁷ Some studies have reported IgG4-related sclerosing disease is involved in thoracic aortitis.⁵ However, some authors contend the elevated IgG4 levels are an incidental finding and don't contribute to the diagnosis due to the inadequate serologic correlation.⁷

Preoperative diagnosis was difficult because CT could not differentiate an inflammatory aneurysm from an intramural hemorrhage. This drawback is also demonstrated in our literature search where six of the thirteen cases were preoperatively diagnosed with an intramural hemorrhage. Furthermore, only two of the thirteen cases had accurate preoperative diagnoses of an inflammatory aortic aneurysm.^{1,4} Echocardiogram is also unable to correctly identify this disease due to the echocardiographic similarities of an aortic wall with inflammation, atherosclerosis, and a thrombosed false lumen.¹ The final diagnosis is presumed by the gross appearance during the operation and confirmed by pathologic evaluation of the tissue. Therefore, an inflammatory aortic aneurysm should be considered preoperatively when there is an abnormal dilated aortic aneurysm with a severely thick wall. At the time of surgery, an inflammatory aneurysm is suspected when there is inflammatory thickening of the aortic wall and excessive adherence to surrounding structures. The histopathologic evaluation of inflammatory aortic aneurysms confirms the diagnosis and typically shows inflammatory lymphoplasmacytic infiltration with destruction of the media.

In summary, we described the case of an inflammatory aneurysm of the ascending aorta with comparable inflammation of the LAD in a young female patient who underwent successful surgical treatment. The underlying etiology of severe inflammatory ascending aortic aneurysms with lymphoplasmacytic infiltration and media destruction without contributory diseases remains uncertain and still open for debate. There is a high surgical mortality (16%) of the inflammatory subtype compared to elective repair of ascending aortic aneurysms.¹ More advanced preoperative imaging is needed to unmask this diagnosis as it is questioned whether this fragile inflammatory aneurysm requires surgery at earlier stages compared to its non-inflammatory counterparts.

Abbreviations

CAD = coronary artery disease

CT = computed tomography

IA = innominate artery

LAD = left anterior descending coronary artery

LCA = left carotid artery

TTE = transthoracic echocardiogram

Declarations

Ethics approval and consent to participate: Not applicable

Consent for publication: Informed consent for publication of this case report and any accompanying images was obtained from the patient.

Availability of data and materials: Not applicable

Competing interests: Not applicable

Funding: Not applicable

Authors' contributions: AS analyzed the case and participated in the design and drafting of the manuscript. AT performed the surgical procedure and participated in the design and drafting of the manuscript. AA assisted in the surgical procedure and participated in the design and drafting of the manuscript. LS analyzed the case and participated in the design and drafting of the manuscript. All authors read and approved the final manuscript.

Acknowledgements: Not applicable

References

1. Pagni S, Ganzel BL, Williams M, Slater AD, Trivedi JR. Inflammatory aneurysms of the ascending aorta: Diagnosis and surgical treatment. *J Card Surg.* 2012;27:70-74.
2. Walker DI, Bloor K, Williams G, Gillie I. Inflammatory aneurysms of the abdominal aorta. *Brit. J. Surg.* 1972;59(8):609-614.
3. Connery CP, Descalzi ME, Kirshner R. Inflammatory aneurysm of the ascending aorta; An unreported entity. *J Cardiovasc Surg.* 1993;35:33-34.
4. Yoshida M, Mukohara N, Honda T, Ozaki N, Shida T. Inflammatory aneurysm of the ascending aorta: report of a case. *Surg today.* 2007;37(9):794-797.
5. Agaimy A, Weyand M, Strecker T. Inflammatory thoracic aortic aneurysm (lymphoplasmacytic thoracic aortitis): a 13-year-experience at a German Heart Center with emphasis on possible role of IgG4. *Int J Clin Exp Pathol.* 2013;6(9):1713-1722.
6. Girardi LN, Coselli JS. Inflammatory aneurysm of the ascending aorta and aortic arch. *Ann Thorac Surg.* 1997;64:251-253.

Figures

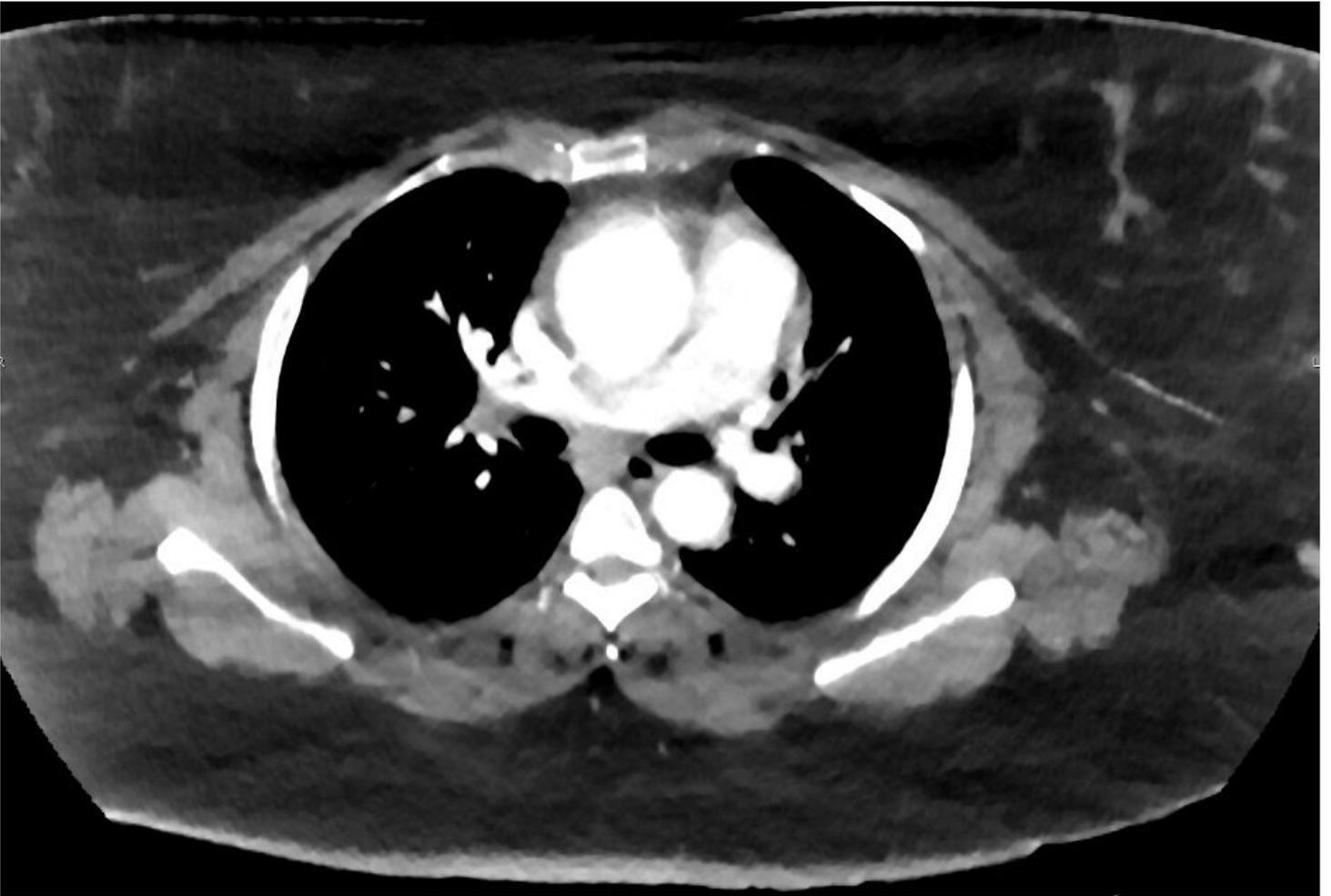


Figure 1

Computed tomogram of an ascending aortic aneurysm with a suspected intramural hematoma.

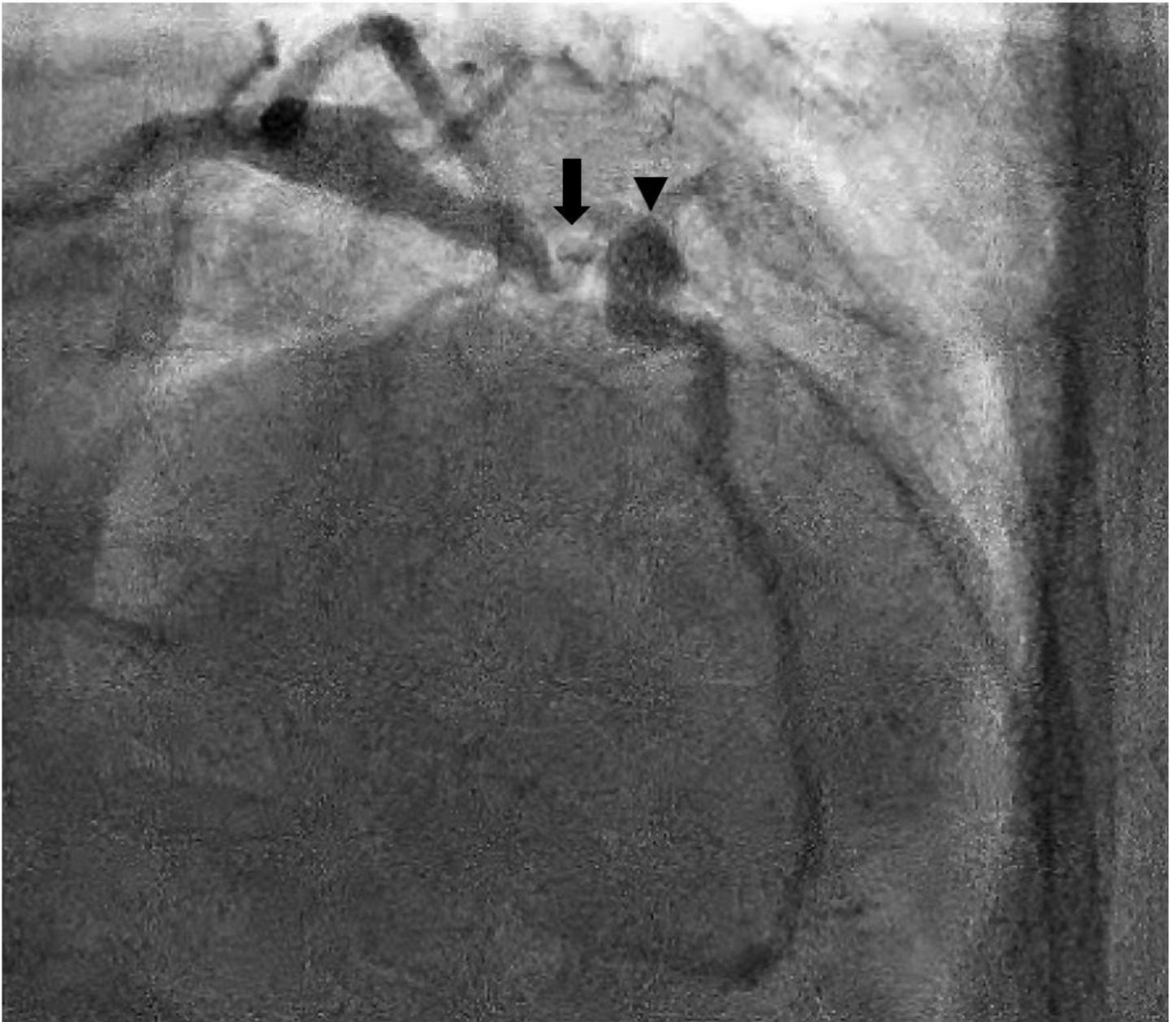


Figure 2

Left Heart Catheterization identifying the left anterior descending coronary artery with 85% stenosis (arrow) followed by a mid-saccular aneurysm (arrowhead)

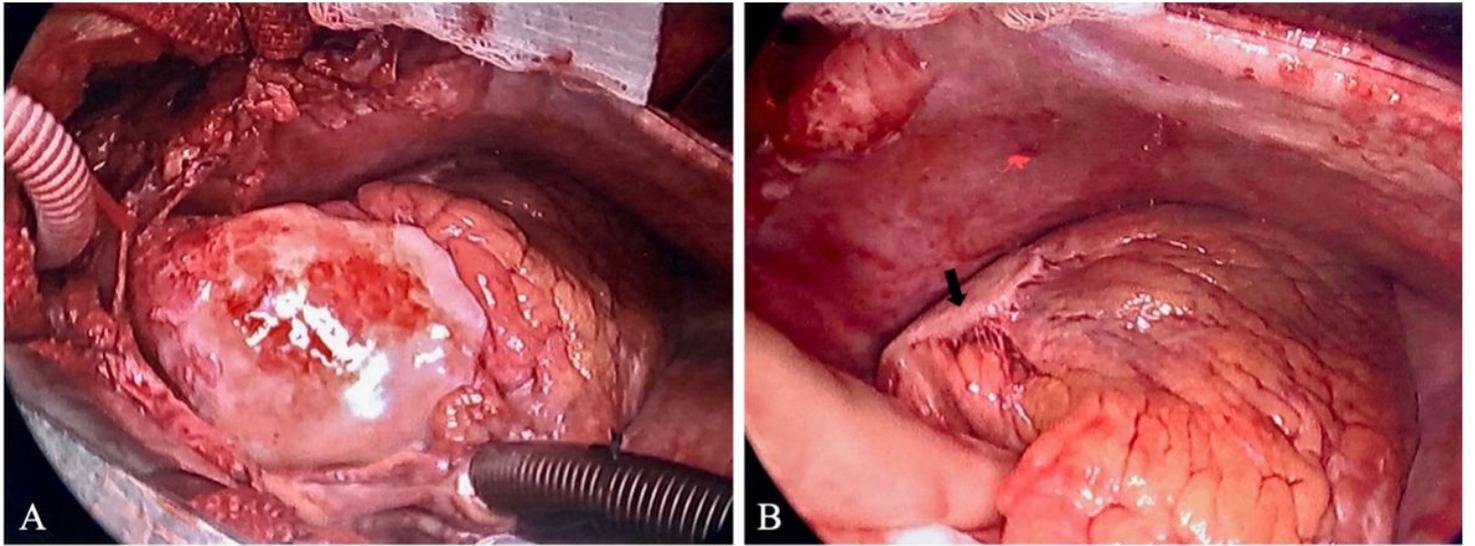


Figure 3

(A) Intraoperative view of the inflammatory ascending aortic aneurysm. (B) Inflammatory plaque overlying the LAD (arrow).