

# Imaging Features and Diagnosis of Infected Aortic Aneurysms and Inflammatory Diseases Involving the Aorta

Takao Kato\* and Moriaki Inoko

Cardiovascular Center, Tazuke Kofukai Medical Research Institute, Kitano Hospital, Japan

## Abstract

In this review article, we focused on the imaging features and diagnosis of infected aortic aneurysms and inflammatory diseases involving the aorta. The imaging features of other inflammatory diseases involving the aorta resemble those of infected aneurysms. In particular, chronic periaortitis is a manifestation of immunoglobulin G4 (IgG4)-related disease, which is a newly recognized syndrome of unknown etiology that has attracted much attention. IgG4-related disease is characterized by a fibroinflammatory condition with a dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells. Considering their similar imaging features, infected aneurysms should be carefully differentiated from chronic periaortitis. The treatment options for these 2 potential diagnoses are completely different and consist of either corticosteroids or antibiotics. Medical history, severity (high or low grade) of fever, physical examination, clinical course (rapidly or slowly progressive), repeated blood cultures, multiple imaging modalities, and elevated IgG4 levels in addition to pathological findings would be useful in obtaining a differential diagnosis in patients suspected of having either of the aforementioned conditions.

**Keywords:** Infected aneurysm; Chronic periaortitis; IgG4-related disease

## Introduction

Infected aortic aneurysm was first reported by Osler [1] in 1885, and it has received increasing attention since then, owing to its rapidly progressive clinical course and fatal outcome [2]. Although the true prevalence of infected aneurysm is unknown, the disease accounts for 0.7-2.6% of cases of aortic aneurysm [3]. The number of reports of infected aneurysms (mycotic aneurysms) in Pubmed search are increasing [4,5], which is a finding that is consistent with our personal experience. The reasons for such increases include: 1) the increased numbers of aged patients, who are at greater risk of atherosclerosis; 2) improved Computed Tomography (CT) imaging and Magnetic Resonance Imaging (MRI) techniques; and 3) greater awareness of the disease. In the present study, we focus on the diagnosis, especially imaging features, of infected aneurysms and diseases related to inflammation of the aorta that require differentiation from infected aneurysms, such as chronic periaortitis (one of the presentations of IgG4-related disease) and collagen vascular diseases.

## Infected Aneurysms

The damaged endothelium is more susceptible to bacterial infections in the presence of bacteremia [6]. Risk factors for infected aneurysm include: 1) endothelial damage caused by atherosclerosis including a pre-existing aneurysm [7]; 2) an antecedent infection including bacteremia; and 3) arterial injury including iatrogenic mechanisms such as percutaneous coronary intervention [8]. When the intima is diseased or if a pre-existing aneurysm is present, bacteria can pass through intima into the deep layers of the aorta and establish an infection. Infected aneurysm develops from a transient or persistent bacteremia or septic emboli. Infective endocarditis (IE) remains the main cause of infected aneurysms [6] because its risk factors are very similar to those of infected aneurysms. Moreover, bacteremia and septic emboli from the heart are often common features of IE.

Along with high fever, the other symptoms of an infected aneurysm vary according to the diseased lesion. For example, abdominal pain and diarrhea are observed if the abdominal aorta is involved, a painful pulsatile mass is noted if the superficial artery is involved, and chest

pain is noted if the thoracic aorta is involved. An infected aneurysm involving the deep arteries or the aorta may cause only fever. It might be labeled as fever of unknown origin and be diagnosed as an infected aneurysm only after CT imaging findings are obtained. The suprarenal abdominal aorta is the most commonly involved site [9]. If the patients had bacteremia or a persistent high fever of unknown etiology, contrast-enhanced CT (CE-CT) imaging would be the choice for identifying the diseased lesion and an asymptomatic infected aneurysm would be among the differential diagnoses. The mortality rate of infected aneurysm is higher than that of non-infected aneurysm [2], especially without appropriate treatments [10,11]. A rapidly progressive clinical course and changes in imaging findings are also useful in the diagnosis, with the time course varying from days [12] to months [11,13].

The next step in diagnosing an infected aneurysm is based on identifying the infection-causing organisms and imaging features. Given that imaging alone cannot clearly differentiate an infection from an inflammation, determining the etiology of high fever is essential. Blood cultures should be performed in all patients in whom infected aneurysm is suspected. An estimated 50-85% of patients may yield positive results [14,15]. *Staphylococcus* spp. and *Salmonella* spp. are 2 major bacteria that cause infected aneurysms [14,16], followed by *Streptococcus pneumoniae* and *Campylobacter* [17,18]. However, even if the patient exhibits a negative blood culture result, infected aneurysm still cannot be ruled out. In such cases, polymerase chain reaction (PCR) amplification targeting the bacterial rRNA gene or direct sequencing may facilitate the identification of the bacteria [19], as in case of culture-

\*Corresponding author: Takao Kato, Cardiovascular Center, Tazuke Kofukai Medical Research Institute, Kitano Hospital, 2-2-20 Ogimachi, Kita-ku, Osaka 530-8480, Japan, Tel: +81-6-6312-8831; Fax: +81-6-6312-8867; E-mail: [takao-kato@kitano-hp.or.jp](mailto:takao-kato@kitano-hp.or.jp)

Received June 27, 2013; Accepted July 28, 2013; Published July 30, 2013

Citation: Kato T, Inoko M (2013) Imaging Features and Diagnosis of Infected Aortic Aneurysms and Inflammatory Diseases Involving the Aorta. Angiol 1: 110. doi: 10.4172/2329-9495.1000110

Copyright: © 2013 Kato T, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

negative endocarditis [20]. CE-CT imaging can be used to identify the location of a diseased aorta and is often useful for identifying infected aneurysms. CT angiographic findings of infected aneurysms include saccular aneurysmal formation with irregular configuration, disrupted aortic wall calcification, soft tissue inflammation or a mass around a vessel, and periaortic fluid or air collection [9,21-23]. Atherosclerotic findings can help clinicians differentiate infected aneurysms from other inflammatory aortic diseases, and periaortic fluid or air collection may indicate bacterial involvements [9,21-23]. An aorta with bacterial infection may appear to be normal in size and to have a thickened aortic wall, with mild contrast enhancement in the initial evaluation [11,12]. Serial CE-CT scans can visualize the extension or dilatation of the diseased area and thus contribute to the effectiveness of therapy [11-13]. However, CT imaging cannot clearly differentiate an infection from an inflammation, because the inflammatory aortic disease coincidentally occurs in an atherosclerotic lesion. Few reports have described MRI findings of infected aortic aneurysms [9,24,25]; however, T2-weighted images or mixed T1/T2-weighted STIR images can be used to visualize the edematous lesion. Diffusion images can be used to detect fluid collection, whereas gadolinium enhancement can indicate increases in the amount of inflammatory connective tissue.

Because mortality rates without surgery were 85% with infected thoracic aneurysm and 96% with infected aortic aneurysm [26,27], surgical replacement or debridement is the treatment of choice combined with antibiotic therapy [16]. The main aims of the surgical procedures include the removal of the infected tissue and revascularization in cases where distal perfusion is limited. Figure 1 shows a gradually enlarging infective aneurysm that was treated with medication alone despite the control of bacteremia [11]. Of note, the treatment choice for diseases with inflammation of the aorta, which require differentiation from infected aneurysms, such as chronic periaortitis [28] and collagen vascular diseases, is completely different from that for infected aneurysm. The incorrect administration of immunosuppressing agents to patients with infected aneurysms is believed to be hazardous [29].

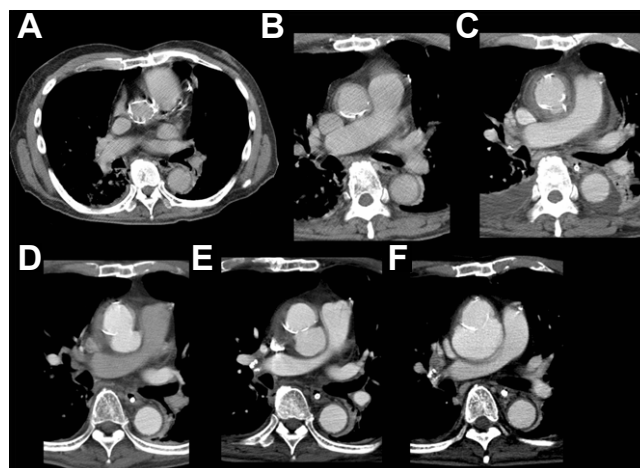
In summary, the important aspects in the diagnosis of infected aneurysm include high suspicion of the disease, medical history, severity (high-grade) of fever, physical examination, rapidly progressive clinical course, positive blood culture results, and enhanced CT or MR imaging findings. Considering the rapid clinical course of the disease, serial imaging may be useful for making a diagnosis.

### Chronic Periaortitis

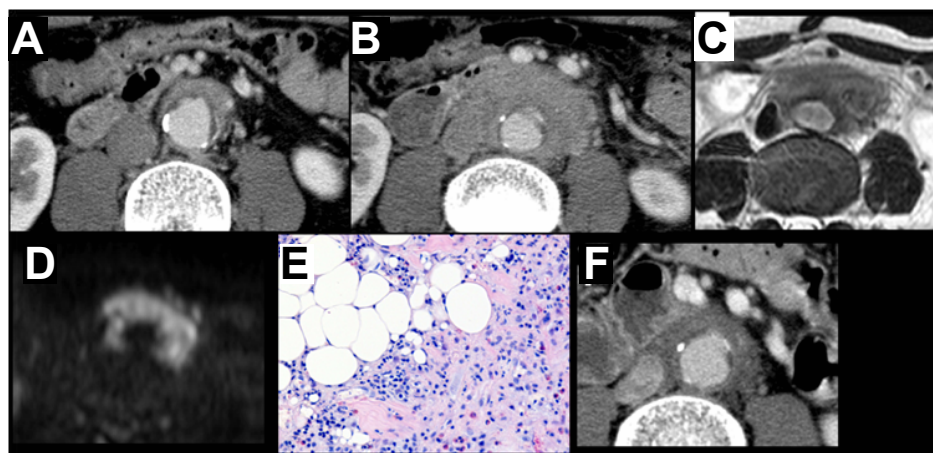
Some of the etiologies of chronic periaortitis are believed to be IgG4-related disease [30], inflammatory aortic aneurysm [30], idiopathic retroperitoneal fibrosis [31], a manifestation of systemic autoimmune diseases [32], and a condition secondary to drug use or malignancy [33]. Various symptoms are observed according to the lesions involved, although patients with this condition appear well without the presence of fever at the time of diagnosis.

IgG4-related disease is a newly recognized syndrome of unknown etiology that has attracted much attention [28]. Approximately 70% of patients with IgG4-related disease demonstrate elevated serum IgG4 concentrations [28]. Allergic features such as atopy, asthma, and modest peripheral eosinophilia, along with tumorous swelling in many organs and elevated serum IgG4 levels above the upper limit of normal (>135 mg/dL), are the characteristics of IgG4-related diseases. Pathological examinations can also confirm the diagnosis of IgG4-related diseases by the presence of tumefactive lesions and dense lymphoplasmacytic infiltrates that are rich in IgG4-positive plasma cells [28]. This diagnosis can be made if these findings are noted in the absence of detectable bacteria, granulomatous lesions, or malignancies.

The imaging features of patients with chronic periaortitis are characterized by homogeneous wall thickening on CE-CT imaging [28,33], with an increase in connective tissue (the so-called “mantle sign”) that is indicative of sclerosing inflammation, and occasionally, with adhesions to the surrounding organs. In inflammatory aortic aneurysm, calcification in the aortic wall may differ according to cases. Because edema and inflammatory connective tissue are common features of inflammation, the features of MRI findings in patients with



**Figure 1:** A case of medical treatment of an infected aneurysm. A 75-year-old man with familial hypercholesterolemia, cerebral infarction, and coronary bypass grafting presented with high fever and chest pain, with five positive blood cultures for methicillin-susceptible *Staphylococcus aureus* and an elevated CRP of 24 mg/dL, but without vegetation on echocardiography or other infected focus. Panels A and B show a severely calcified ascending aorta with thickened wall. Repeated contrast-enhanced computed tomography on the 5th day revealed an aneurysmal change with protrusion (Panel C), indicating the presence of an infected aneurysm. Due to the prohibitive risk of surgery, medical treatment was chosen by the patient and his family, but the infected aneurysm gradually enlarged despite the control of bacteremia with antibiotics (Panel D, 13th day; Panel E, 24th day; Panel F, 58th day) and he died 2 months after onset. Kato T, Kimura M, Inoko M, Nohara R (2012) Infected aneurysm in an atherosclerotic lesion. *Intern Med.* 51:983. Permission obtained from the Japanese Society of Internal Medicine.



**Figure 2:** A) case of chronic periaortitis with retroperitoneal fibrosis. A 42-year-old man presented with vague discomfort within the lower abdomen. Panel A shows the area at the time of presentation. Two months later, the patient's symptoms remained unchanged (C-reactive protein level, 8.5 mg/dL), and no fever was noted. Repeated computed tomography was performed (Panel B). (C) Low-intensity enhancement was observed on a T2-weighted, gadolinium-enhanced magnetic resonance image. (D) High intensity was observed on a diffusion-weighted image. (E) A tissue biopsy revealed the infiltration of inflammatory cells and the absence of bacteria. (F) A contrast-enhanced computed tomography image obtained 3 weeks after the initiation of glucocorticoid therapy.

chronic periaortitis resemble those of patients with infected aneurysms. However, serial follow-up of suspicious lesions to document the response to therapy can be of great value (Figure 2). Negative results on repeated blood cultures and the presence of elevated IgG4 levels in addition to pathological findings can help in the differential diagnosis. Pathological examinations can confirm the presence of the IgG4-related diseases mentioned above [28]. Moreover, glucocorticoids are typically the first line of therapy in such cases (Figure 2).

In summary, chronic periaortitis should be differentiated from infected aneurysm not only by the imaging features but also by the clinical symptoms, findings, and negative results on blood cultures in addition to pathological findings. Careful attention is needed to treat both conditions effectively [34].

### Other Inflammatory Diseases Involving the Aorta

Large-vessel vasculitis such as Takayasu's arteritis and Giant Cell Arteritis (GCA), rheumatic and HLA-B27-associated spondyloarthropathies, Behçet's syndrome, and infections such as syphilis and tuberculosis may be the causes of aortic inflammation. Therefore, in cases where negative results are obtained on blood culture and where the infected focus remains unclear, it is essential that the abovementioned conditions should be differentiated from infected aneurysms or chronic periaortitis. A detailed history and careful physical examination are important for the diagnosis and assessment of the extent of the vascular lesion. Systemic symptoms such as fatigue, weight loss, and low-grade fever are common in these disorders, along with local symptoms such as arthralgia, skin lesions (erythema nodosum), and abdominal pain.

Takayasu's arteritis, also called pulseless disease, involves the ascending aorta, aortic arch, and carotid and subclavian arteries, causing dilation and obstruction at the healing stage as well as recurrence. Takayasu's arteritis primarily affects women who are 17–26 years of age at the time of onset [35]. CT angiography can identify the diseased lesion and a thickened arterial wall in the acute phase and aneurysmal or stenotic lesions in the chronic phase [36]. Ultrasonography and MRI angiography are also useful for the detection of this condition. In the acute phase, the high signal of T2-weighted and/or STIR MRI and the

increased uptake of <sup>18</sup>F-fluorodeoxyglucose indicate the presence of active inflammation [37]. Glucocorticoid administration remains the primary therapy for Takayasu's arteritis.

GCA consists of chronic vasculitis of the large and medium vessels and mainly affects women [35]. The following classification criteria were developed: 1) age > 50 years at onset; 2) localized de novo headache; 3) tenderness or decreased pulse of the temporal artery; 4) erythrocyte sedimentation rate >50 mm/h; and 5) biopsy-proven necrotizing arthritis with multinucleated giant cells [38,39].

### Conclusions

Considering their similar imaging features, infected aneurysms and chronic periaortitis should be carefully differentiated. We believe that history, severity (high- or low- grade) of fever, physical examination, clinical course (rapidly or slowly progressive), repeated blood cultures, and multiple imaging modalities can be useful in obtaining a differential diagnosis, as well as in determining the presence of elevated IgG4 levels and pathological findings.

### References

- Osler W (1885) The Gulstonian Lectures, on Malignant Endocarditis. *Br Med J* 1: 467-470.
- Oderich GS, Panneton JM, Bower TC, Cherry KJ Jr, Rowland CM, et al. (2001) Infected aortic aneurysms: aggressive presentation, complicated early outcome, but durable results. *J Vasc Surg* 34: 900-908.
- Hsu RB, Chen RJ, Wang SS, Chu SH (2004) Infected aortic aneurysms: clinical outcome and risk factor analysis. *J Vasc Surg* 40: 30-35.
- <http://www.ncbi.nlm.nih.gov/pubmed/?term=infected+mycotic+aneurysm>
- Kato T (2013) Infected aneurysm and inflammatory aorta: Diagnosis and management In: Aronow WS, eds. In Tech, 2013. DOI: 10.5772/54415.
- Johansen K, Devin J (1983) Mycotic aortic aneurysms. A reappraisal. *Arch Surg* 118: 583-588.
- Ernst CB, Campbell HC Jr, Daugherty ME, Sachatello CR, Griffen WO Jr (1977) Incidence and significance of intra-operative bacterial cultures during abdominal aortic aneurysmectomy. *Ann Surg* 185: 626-633.
- Samore MH, Wessollosky MA, Lewis SM, Shubrooks SJ Jr, Karchmer AW (1997) Frequency, risk factors, and outcome for bacteremia after percutaneous transluminal coronary angioplasty. *Am J Cardiol* 79: 873-877.

9. Macedo TA, Stanson AW, Oderich GS, Johnson CM, Panneton JM, et al. (2004) Infected aortic aneurysms: imaging findings. *Radiology* 231: 250-257.
10. Müller BT, Wegener OR, Grabitz K, Pillny M, Thomas L, et al. (2001) Mycotic aneurysms of the thoracic and abdominal aorta and iliac arteries: experience with anatomic and extra-anatomic repair in 33 cases. *J Vasc Surg* 33: 106-113.
11. Kato T, Kimura M, Inoko M, Nohara R (2012) Infected aneurysm in an atherosclerotic lesion. *Intern Med* 51: 983.
12. Imori A, Kanzaki Y, Ito S, Kotani T, Hirano-Kuwata S, et al. (2010) Rapidly progressing aneurysm of infected thoracic aorta with pseudoaneurysm formation. *Intern Med* 49: 2461-2465.
13. Murakami I, Kou T, Oda Y, Yazumi S (2011) Rapidly progressing mycotic aortic aneurysm. *Intern Med* 50: 161.
14. Johnson JR, Ledgerwood AM, Lucas CE (1983) Mycotic aneurysm. New concepts in therapy. *Arch Surg* 118: 577-582.
15. Maeda H, Umezawa H, Goshima M, Hattori T, Nakamura T, et al. (2011) Primary infected abdominal aortic aneurysm: surgical procedures, early mortality rates, and a survey of the prevalence of infectious organisms over a 30-year period. *Surg Today* 41: 346-351.
16. Moneta GL, Taylor LM Jr, Yeager RA, Edwards JM, Nicoloff AD, et al. (1998) Surgical treatment of infected aortic aneurysm. *Am J Surg* 175: 396-399.
17. Brouwer RE, van Bockel JH, van Dissel JT (1998) *Streptococcus pneumoniae*, an emerging pathogen in mycotic aneurysms? *Neth J Med* 52: 16-21.
18. Cinà CS, Arena GO, Fiture AO, Clase CM, Doobay B (2001) Ruptured mycotic thoracoabdominal aortic aneurysms: a report of three cases and a systematic review. *J Vasc Surg* 33: 861-867.
19. Koo M, Manalili S, Bankowski MJ, Sampath R, Hofstadler SA, et al. (2010) A "silent culture-negative" abdominal aortic mycotic aneurysm: Rapid detection of *Bartonella* species using PCR and high-throughput mass spectrometry. *Hawaii Med J* 69: 68-69.
20. Goldenberger D, Künzli A, Vogt P, Zbinden R, Altwegg M (1997) Molecular diagnosis of bacterial endocarditis by broad-range PCR amplification and direct sequencing. *J Clin Microbiol* 35: 2733-2739.
21. Gomes MN, Choyke PL (1992) Infected aortic aneurysms: CT diagnosis. *J Cardiovasc Surg (Torino)* 33: 684-689.
22. Lee WK, Mossop PJ, Little AF, Fitt GJ, Vrazas JI, et al. (2008) Infected (mycotic) aneurysms: spectrum of imaging appearances and management. *Radiographics* 28: 1853-1868.
23. Oz MC, Brenner BJ, Buda JA, Todd G, Brenner RW, et al. (1989) A ten-year experience with bacterial aortitis. *J Vasc Surg* 10: 439-449.
24. Walsh DW, Ho VB, Haggerty MF (1997) Mycotic aneurysm of the aorta: MRI and MRA features. *J Magn Reson Imaging* 7: 312-315.
25. Moriarty JA, Edelman RR, Tumei SS (1992) CT and MRI of mycotic aneurysms of the abdominal aorta. *J Comput Assist Tomogr* 16: 941-943.
26. Weis-Müller BT, Rascanu C, Sagban A, Grabitz K, Godehardt E, et al. (2011) Single-center experience with open surgical treatment of 36 infected aneurysms of the thoracic, thoracoabdominal, and abdominal aorta. *Ann Vasc Surg* 25: 1020-1025.
27. Taylor CF, Lennox AF (2007) Treatment options for primary infected aorta. *Ann Vasc Surg* 21: 225-227.
28. Stone JH, Zen Y, Deshpande V (2012) IgG4-related disease. *N Engl J Med* 366: 539-551.
29. Smith FC, Rees E, Elliott TS, Shearman CP (1994) A hazard of immunosuppression: *Aspergillus niger* infection of abdominal aortic aneurysm. *Eur J Vasc Surg* 8: 369-371.
30. Kasashima S, Zen Y, Kawashima A, Endo M, Matsumoto Y, et al. (2009) A new clinicopathological entity of IgG4-related inflammatory abdominal aortic aneurysm. *J Vasc Surg* 49: 1264-1271.
31. Vaglio A, Buzio C (2005) Chronic periaortitis: a spectrum of diseases. *Curr Opin Rheumatol* 17: 34-40.
32. Vaglio A, Greco P, Corradi D, Palmisano A, Martorana D, et al. (2006) Autoimmune aspects of chronic periaortitis. *Autoimmun Rev* 5: 458-464.
33. Inoue D, Zen Y, Abo H, Gabata T, Demachi H, et al. (2011) Immunoglobulin G4-related periaortitis and periarteritis: CT findings in 17 patients. *Radiology* 261: 625-633.
34. Ishizaka N, Sohmiya K, Miyamura M, Umeda T, Tsuji M, et al. (2012) Infected aortic aneurysm and inflammatory aortic aneurysm--in search of an optimal differential diagnosis. *J Cardiol* 59: 123-131.
35. Hunder GG, Arend WP, Bloch DA, Calabrese LH, Fauci AS, et al. (1990) The American College of Rheumatology 1990 criteria for the classification of vasculitis. Introduction. *Arthritis Rheum* 33: 1065-1067.
36. Yamada I, Nakagawa T, Himeno Y, Numano F, Shibuya H (1998) Takayasu arteritis: evaluation of the thoracic aorta with CT angiography. *Radiology* 209: 103-109.
37. Kissin EY, Merkel PA (2004) Diagnostic imaging in Takayasu arteritis. *Curr Opin Rheumatol* 16: 31-37.
38. Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, et al. (1990) The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. *Arthritis Rheum* 33: 1122-1128.
39. Oz MC, Brenner BJ, Buda JA, Todd G, Brenner RW, et al. (1989) A ten-year experience with bacterial aortitis. *J Vasc Surg* 10: 439-449.