

# A Rapidly Growing Pseudoaneurysm Secondary to Aortic Arch Periaortitis

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**Abstract** We herein report a case of rapidly growing pseudoaneurysm developed shortly after a diagnosis of aortic arch periaortitis. Pseudoaneurysm is a blood leak constrained by compliance of perivascular tissues, in persistent communication with the feeding artery. Chronic periaortitis is characterized by inflammatory involvement of the outer layer of the aorta and surrounding tissues. The major pathogenetic process leading to periaortitis is inflammation, caused by advanced atherosclerosis. Autoimmunity has been proposed as a contributing factor. The present case illustrates an ongoing process of aortic arch periaortitis toward aneurysm formation. Aortic arch pseudoaneurysm is highly susceptible to rupture and sudden death. Our patient achieved a complete remission by a month after receiving an endovascular stent repair and combined treatment with steroid and antibiotics. The disease process was documented by serial CT and radionuclide scanning.

**Keywords:** aortic arch, atherosclerosis, computed tomography, Ga-67 scan, periaortitis, pseudoaneurysm

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## 1. Introduction

Pseudoaneurysm is a transmural blood leak held by adjoining tissues and fibrous reaction, and communicates with the feeding artery [1]. Pseudoaneurysm of the thoracic aorta often occurs as a sequel of trauma, rarely from non-traumatic pathologies such as penetrating atherosclerotic ulcers and periaortitis [2]. The major pathogenetic process leading to periaortitis is inflammation, caused by advanced atherosclerosis [3]. Autoimmunity to certain components of atherosclerotic plaque has been proposed as the stimulus to initiate the inflammatory process [4]. Since a pseudoaneurysm does not contain arterial wall components, the impact of high pressure and blood flow on the aneurysmal sac may result in rupture and sudden death. Delay in diagnosis of the aneurysm and misuse of antimicrobial agents may lead to serious complications [5]. Endovascular stent grafting can be used as a temporary measure to quickly achieve hemodynamic stability and as a bridging measure to possibly allow further definitive surgical treatment [6].

## 2. Case Report

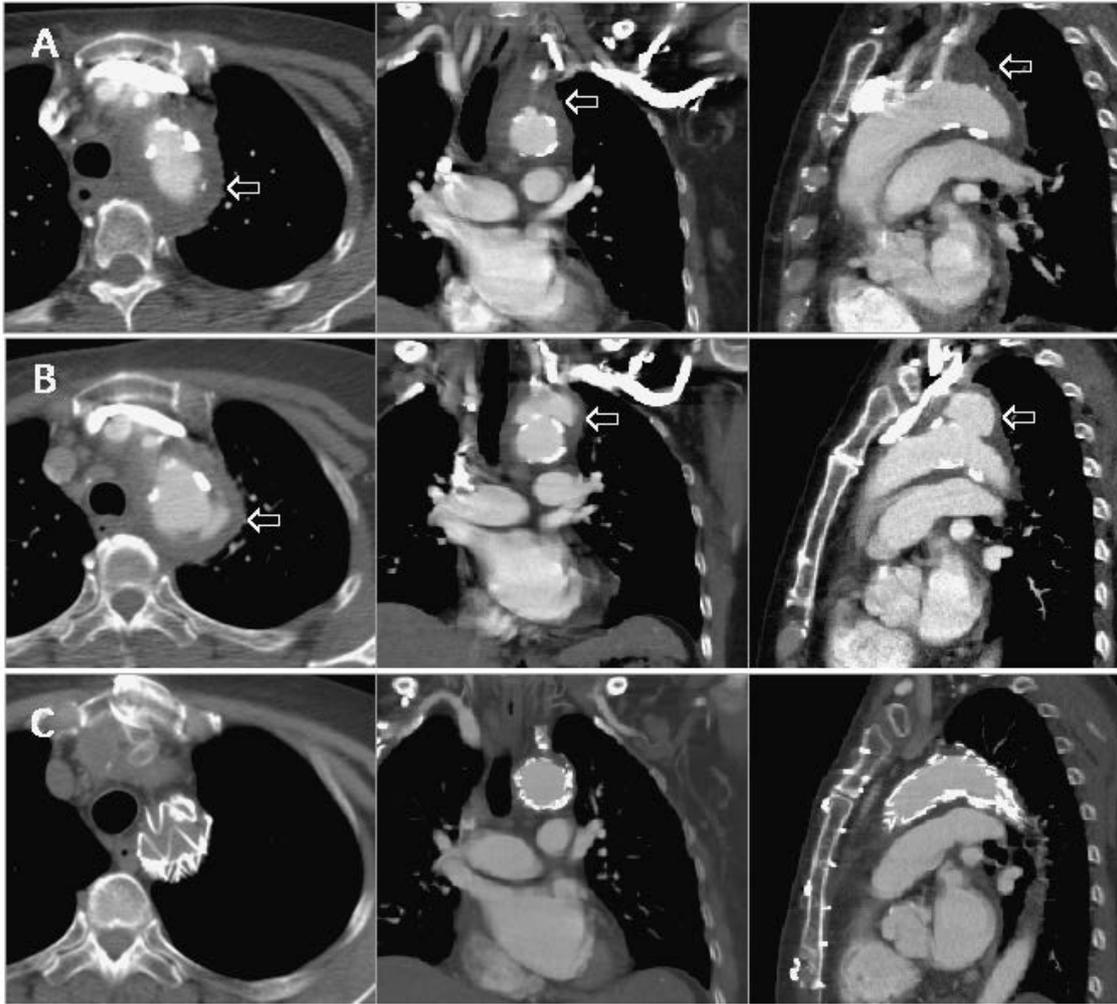
A 71-year-old hypertensive woman was referred to our hospital due to general malaise and chest pain for one week that localized to the left upper sternal border. She had been treated at another hospital, but chest pain remained her chief complaint throughout. She had been

hypertensive for 20 years and had no history of smoking, trauma or diabetes.

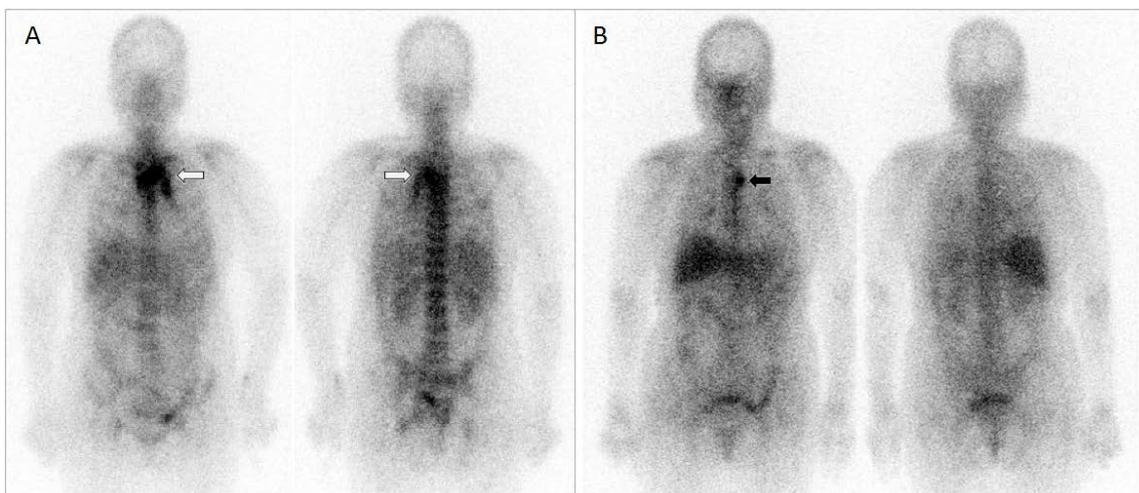
On admission, the patient was afebrile at 37°C and BP 131/78 mmHg. Cardiopulmonary, arterial pulsation and abdominal examinations were unremarkable. WBC 12,500 cells/ $\mu$ L, Hgb 11.1 g/dL, erythrocyte sedimentation rate 101 mm/hr (ESR range  $\leq$ 25 mm/hr), C-reactive protein 18.94 mg/dl (CRP range  $\leq$ 0.5 mg/dl), and negative troponin I. Infectious serology for legionella, chlamydia and syphilis was negative. Three cultures of blood failed to yield any bacterial growth. Her immune parameters came up positive for IgG 2269 mg/dl (range: 751-1560 mg/dl), ANA 1: 80 and SSA >10 U/ml. IgG4 was 65.9 mg/dl (range: 3-200 mg/dl). Echocardiography was basically normal. Contrast-enhanced CT of the chest showed diffuse wall thickening of the aortic arch and soft-tissue infiltration in superior mediastinum (Figure 1A). In the context of suspicion of mediastinitis and aortitis, intravenous ceftriaxone (2 g/day) and teicoplanin (400 mg/day) were started.

However, on day 5, the pain got worse instead of better, fever at 38.5°C, and ongoing elevation of infectious parameters, ESR 121 mm/h and CRP 20.33 mg/dl; two repeated blood cultures were sterile. As a part of infection workup, Ga-67 citrate scan illustrated pathological uptake around the aortic arch, suggestive of periaortitis (Figure 2A). Ga-67 SPECT and fused with CT images further supported the notion that aortic arch was the source of the inflammation (Figure 3A). The fever continued unabated for the next ten days despite antibiotic therapy. Follow-up chest CT revealed a newly found outpouching at the top of aortic arch (Figure 1B). A mushroom-like aneurysm of the

aortic arch was favored, and immediate repair of the aneurysm for the risk of impending rupture was required.



**Figure 1.** Serial CT scans documented an ongoing process of periaortitis. (A, upper panels) Upon presentation, contrast-enhanced CT showed a homogeneous wall thickening of the aortic arch (*arrow*) and increased soft tissue density in the superior mediastinum. The findings were consistent with chronic periaortitis or infectious aortitis. (B, middle panels) Day 14, follow-up CT displayed a contrast-filled, mushroom-like protrusion arising from the aortic arch, about 38×22 mm, suggesting a newly developed pseudoaneurysm (*arrow*). (C, lower panels) 12 months after operation, CT exhibiting significant resolution of perivascular infiltration and regression of aneurysmal pouch, the hyperdense stent graft in situ without evidence of endoleak



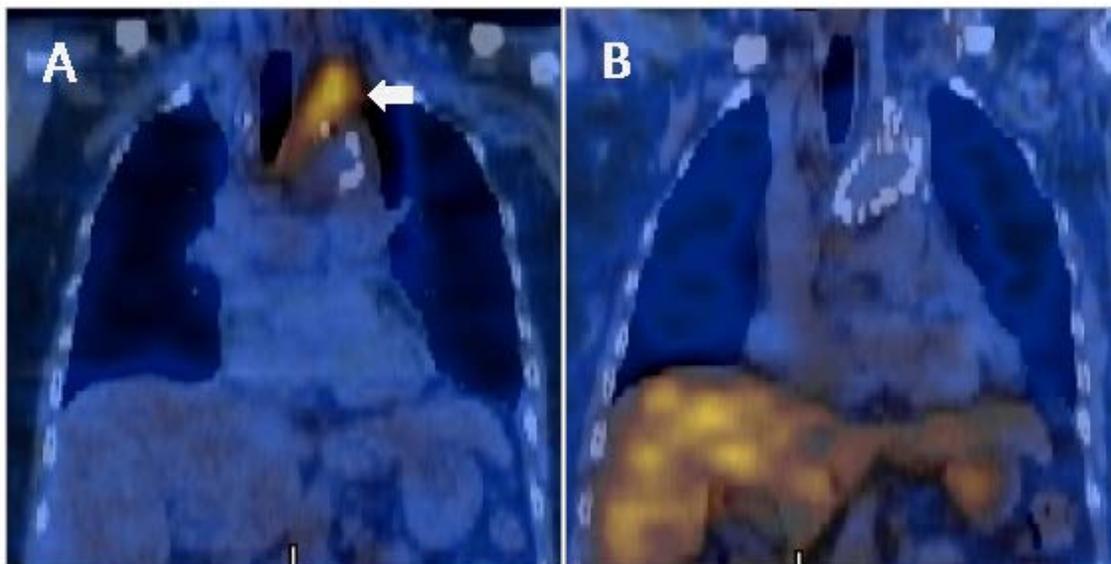
**Figure 2.** Ga-67 citrate planar imaging. (A) In initial work-up, intense activity was noted at the aortic arch (*white arrows*). (B) Follow up scan 4 weeks after surgery and steroid therapy, the previously described lesion at the aortic arch faded out. Linear uptake along the sternum corresponded to the surgical wound (*black arrow*)

The patient underwent a debranching and endovascular stent repair of the aortic arch. Histology showed myxomatous degeneration and atherosclerotic changes of

the aortic wall, and reactive hyperplasia of mediastinal lymphnodes. Microscopic and cultures of operative specimens revealed no infection. At this point, the

diagnosis of chronic periaortitis with pseudoaneurysm formation was made. The patient was considered a candidate for a trial of steroid therapy. Intravenous methylprednisolone 30 mg b.i.d was given for one week, and a steroid tapering continued over 4 more weeks. Because of her temperature and inflammatory markers remained raised at operation, and of the concern of the risk of prosthetic infections, intravenous antibiotic therapy

was continued for four weeks. Her symptoms eventually settled after the regimens. Complete remission was achieved and documented on Ga-67 scan (Figure 2B & Figure 3B). At the 1-year follow-up visit, CT scan exhibited consequent regression of perivascular infiltration and sac shrinkage (Figure 1C), considered to be evidence of clinical success.



**Figure 3.** Ga-67 SPECT/CT fused images. (A) Avid uptake of Ga-67 citrate on the wall of the aortic arch (arrow) on initial work-up. (B) Resolution of the previous disease process at the aortic arch 4 weeks after endovascular stent grafting

### 3. Discussion

A pseudoaneurysm represents an arterial leak that is confined by the outer-most adventitia or perivascular tissues, in persistent communication with the feeding artery [1]. In contrast, a true aneurysm is an outpouching due to dilation of all 3 intact layers of the arterial wall, i.e. adventitia, media and intima [1,7]. Blunt trauma and iatrogenic injury are the frequent causes of pseudoaneurysms of the thoracic aorta [7,8]. They also occur from non-traumatic pathologies such as penetrating atherosclerotic ulcers, infectious diseases, inflammatory disorders, and congenital anomalies [2,9,10]. Pseudoaneurysm caused by atherosclerotic disease is usually found in the descending aorta and aortic arch. Lesion has rarely been noted in the ascending aorta, where atherosclerotic plaque is less common [11,12]. Atherosclerotic plaques may ulcerate and erode with leaking within the aortic wall, which can spread into the adventitia, forming a pseudoaneurysm, or may also cause rupture [13]. The impact of continued arterial flow and high pressure on the aneurysmal pouch plays a major role of expansion and rupture.

Chronic periaortitis (CP) is characterized by inflammatory involvement of the outer layer of the aorta and surrounding tissues [14]. The major pathogenetic process leading to CP is inflammation, caused by advanced atherosclerosis [3]. The inflammatory intensity ranges from mild in atherosclerosis to more extensive in periaortitis [15]. Although CP has a classical predilection for abdominal aorta, an involvement of thoracic aortas in one-third of patients was described in recent cohort studies [16,17]. The atherosclerotic lesion contains large numbers

of immune cells, particularly macrophages and T cells [4]. Autoimmunity to certain components of atherosclerotic plaque has been proposed as the antigenic stimulus to initiate the inflammatory process [3,4]. More specifically, periaortitis is thought to be an autoimmune response to an insoluble lipid called ceroid that has leaked through an arterial wall being injured by the presence of atherosclerotic plaques [18,19]. It is likely that inflammatory response or autoimmune reaction affect the resistance of the aortic wall, predisposing to blood leaking and pseudoaneurysm development [20].

Dyspnea and chest pain are the most frequent symptoms of thoracic periaortitis. However, about 20% are asymptomatic at the time of detection [21]. Diagnosis of periaortitis is primarily based on imaging findings [22]. Chest x-ray might reveal a widened mediastinum but is frequently unremarkable, especially in those with small pseudoaneurysms [10]. Aortic wall thickening, periaortic infiltration and fibrosis, and aneurysm formation may be seen on CT or magnetic resonance angiography [23,24]. Ga-67 scan or F-18 FDG PET, capable of detecting radiographically occult foci of infection, may demonstrate increased radioactivity depending on the inflammatory activity. Importantly, initial avid radioactivity has been appreciated to normalize with appropriate steroid or antimicrobial therapy for a variety of aortitis [25]. Serial imaging is useful in inspecting for aneurysm development and in monitoring of established aneurysms to accommodate timely intervention for those at high risk of rupture [25]. Some late outcomes unique to endovascular aneurysm repair, such as endoleak, migration, device failure, stent-graft patency, and recurrent infection, may not be anticipated. For these reasons, ongoing surveillance appears to be of continued importance [26].

Treatment of thoracic periaortitis depends on the underlying cause. An exaggerated inflammatory response to advanced atherosclerosis has been thought to be the main pathogenetic process [3]. Autoimmunity has been proposed as a contributing factor. In patients with idiopathic periaortitis, glucocorticoids are traditionally considered the mainstay of treatment [15,22]. In a rabbit model of atherosclerosis induced by Chlamydia pneumoniae, early treatment with antibiotic (macrolide) is highly effective in abrogating the antibody response and in preventing atherosclerotic lesions (87%) and periaortitis (100%) [27]. Data from larger clinical trials are needed to determine its optimal role. With a high risk of expansion and rupture, surgical intervention is recommended for all patients with thoracic aortic pseudoaneurysm irrespective of symptom status [28]. The conventional open surgery was gradually replaced by endovascular repair, due to the complexity of the operation, surgical insult and high associated mortality rate [21].

Infected (mycotic) aneurysms need to be carefully differentiated from this syndrome in view of the similar imaging features [29]. Delay in diagnosis and misuse of corticosteroid may lead to uncontrolled growth of microorganisms [29]. Routine perioperative antibiotics might not completely suppress bacterial activity, whereas a longer period of preoperative antibiotics may have a better chance of eradicating the infection, but there is no consensus on the optimal duration of antibiotic therapy [6]. Most commonly, parenteral antibiotics are given for 2 to 8 weeks after surgery, but whether lifelong oral antibiotics are necessary is debated [6]. As explored in a meta-analysis by Kan et al, age older than 65 years, complicated aneurysm, and fever at the time of surgery were risk factors associated with persistent infection. They also noted that no microbes could be isolated from blood and tissue cultures in 25% to 40% of infected (mycotic) aortic aneurysms [6]. As illustrated in our case report, a rapidly growing pseudoaneurysm implied that the perivascular tissues might have been damaged and that the patient was in a critical condition. Recurrent fever and raised inflammatory markers might indicate that patients have an active infection or an occult infection that was not well controlled by antibiotics [6]. It is reasonable to commence a prolonged antibiotic treatment combined with adjunct procedures to achieve the therapeutic goals.

#### 4. Conclusion

Aortic arch pseudoaneurysm should be taken into account in the differential diagnosis of acute chest pain. In spite of the availability of noninvasive imaging modalities, a heightened awareness remains indispensable for an early diagnosis. Prompt and appropriate intervention is crucial, because rupture of the aneurysm can be fatal. Localization of disease activity and structural abnormality is useful for guiding diagnosis and treatment planning [25]. Multimodality imaging is useful for initial screening, routine follow up, and post therapeutic assessment.

#### Conflict of Interest

We declare that we have no conflict of interest.

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