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1. Introduction

Due to the increase of aged patients with atherosclerosis, more attention should be paid to the endothelial damage of great vessels. The damaged endothelium is more susceptible to bacterial infections. The reports or papers of infected aneurysms (mycotic aneurysms) are increasing in Pubmed search (Figure 1), which is consistent with personal experience in daily practice. The reasons for the increase were due to several causes: 1) the increase of aged patients with more risk of atherosclerosis than before, 2) an improvement of CT imaging and MR imaging, 3) more awareness of the disease. We focused the diagnosis and managements of infected aneurysms, and the diseases needed to differentiate from infected aneurysms, such as collagen vascular diseases and periaortitis.

1.1. Diagnosis of infected aneurysms

Risk factors for infected aneurysm include: 1) Endothelial damage caused by atherosclerosis including pre-existing aneurysm [1], 2) Antecedent infection including bacteremia, 3) Arterial injury including iatrogenic mechanisms, such as percutaneous coronary intervention [2]. When the intima is diseased, bacteria can pass through it into deep layers of the aorta and can establish infection. Infective endocarditis (IE) remains to be main cause of infected aneurysm [3], because the risk factors of IE are very similar to those of infected aneurysms, and bacteremia and septic emboli from heart are often common features of IE.

Staphylococcus species (spp) and Salmonella spp are two major bacteria causing infected aneurysm, 28 to 71 percent and 15 to 24 percent of causes, respectively [4,5]. Streptococcus pneumoniae may be the third major, re-emerging, cause of infected aneurysms [6]. The pathology of the diseased site includes acute or chronic inflammation with bacterial
infection, abscesses, and necrosis. The suprarenal abdominal aorta is most commonly involved site.

Figure 1. Results of Pubmed search using mycotic aneurysm or infected aneurysm.

Symptoms of an infected aneurysm vary according to the lesion diseased. For example, abdominal pain and diarrhea is observed if abdominal aorta is involved, painful pulsatile mass if superficial artery is involved, and chest pain if thoracic aorta is involved. An infected aneurysm involving deep arteries or aorta may cause only fever. It might be followed as fever of unknown origin, and could be diagnosed as an infected aneurysm only after CT imaging is acquired. If the patients were with bacteremia or a persistent high fever and their etiology was not determined, the contrast-enhanced CT imaging would be the choice for searching the diseased lesion, and an asymptomatic infected aneurysm would be one of differential diagnoses.

The next step of diagnosing an infected aneurysm is based on blood cultures and imaging. In all suspicious patients, blood cultures should be examined. About 50-85% of patients may be positive [5,7]. If negative, however, the infected aneurysm cannot be ruled out. Contrast-enhanced CT imaging identified the infected aneurysm. Findings on CT angiography of an infected aneurysm include a disruption of aortic wall calcification, soft tissue inflammation or mass around a vessel, and periaortic fluid or air collection [8,9]. These findings can differentiate from other inflammatory aortic disease. The wall of inflammatory aorta is thickened and periaortic fibrosis sometimes observed in adhesion to surrounding organs. MR imaging is another strong tools. The T2-weighted images or mixed T1/T2-weighted STIR images are
able to visualize the edematous lesion. The diffusion images can detect fluid collection, and gadolinium enhancement indicates the increase of inflammatory connective tissue.

In summary, the important things for the diagnosis of an infected aneurysm are the high suspicion of this disease and obtaining blood cultures and enhanced CT or MR imaging.

1.2. Management of infected aneurysms

Surgical replacement or debridement is the treatment of choice combined with antibiotic therapy [4]. The main aims of surgical procedures are removal of infected tissue and re-vascularization if distal perfusion is limited. Mortality rate without surgery was 85 percent with infected thoracic aneurysm and 96 percent with infected aortic aneurysm [10,11]. Figure 2 shows a gradually enlarged infective aneurysm treated on medication alone despite the control of bacteremia [12]. Among patients who underwent surgery, mortality rates were the highest for patients with infected arch aneurysms (50 %) compared with supra-renal aortic aneurysms (43%), distal descending thoracic aneurysms (33 %), proximal descending thoracic aneurysms (16 %), or infra-renal aortic aneurysms (4 %) [10,13]. Endovascular stenting is reported to be effective in some systematic reviews with low mortality [14,15]. Because the infected focus is not removed by endovascular stenting, the procedures may be palliative, and more persistent or recurrent infections are likely to occur compared to surgical procedures. However, endovascular procedures could be a secondary choice for patients who refuse surgery, those with a very high risk for surgery, and those with a ruptured infected aneurysm.

The initial choice of antibiotic therapy should be based on the culture and susceptibility results. Until the results become available, the combination treatment with vancomycin and a ceftriaxone, a fluoroquinolone, or piperacillin-tazobactam is preferable targeting gram-negative Salmonella and enteric bacteria. The optimal duration of antibiotic therapy is uncertain because of the lack of randomized clinical trials. In general, four to six weeks of parenteral antimicrobial therapy is performed for the treatment of infected aneurysm followed by principles of vascular graft infection or infective endocarditis of prosthesis valve. A longer duration of treatment or additional oral antibiotics may be warranted in the clinical course of persist elevation of C-reactive proteins or recurrence of fever when drug-related fever is excluded.

In summary, the surgical replacement in combination with antibiotics is the treatment of choice, and endovascular procedures may be palliative. The management is followed by principles of vascular graft infection or infective endocarditis of prosthesis valve.

2. Inflammation of aorta: “Aortitis”

Large vessel vasculititis such as Takayasu’s arteritis and giant cell arteritis, rheumatic and HLA-B27–associated spondyloarthropathies, Behçet’s syndrome, and infections such as syphilis, tuberculosis may be the cause of inflammation of aorta, and we must differ-
entiate these from infected aneurysms when blood culture is negative or infected focus remains unclear. Another disease which we must differentiate is IgG4-related diseases (chronic periaortitis).

Figure 2. 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. Panel A and B showed a severely calcified ascending aorta. The repeated CE-CT on the 5th day revealed aneurysmal change with protrusion (Panel C), indicating an infected aneurysm. Due to a prohibitive risk of surgery, medical treatment was the choice of him and his family, but the infected aneurysm gradually enlarged despite the control of bacteremia with antibiotics (13th day: Panel D, 24th day: Panel E, and 58th day: Panel F), and he died 2 months after onset.

A detailed history and a careful physical examination are important for the diagnosis and assessment of the extent of vascular lesions. The mean age at onset of Takayasu’s arteritis and giant cell arteritis was between 17 and 26 years of age and with 69 years, respectively, primarily in women (about 80 %) [16]. Systemic symptoms are common such as fatigue, weight loss, and low-grade fever are common in these disorders, along with local symptoms, for example, arthralgia, skin lesion (erythema nodosum), and abdominal pains. HLA-B27–associated spondyloarthritis accompanies with ankylosing spondylitis, reactive arthritis, or inflammatory bowel disease with negative rheumatic factors. Skin and mucosa, ocular system, GI manifestations include abdominal pain, nausea, and diarrhea with or without blood, and/or musculoskeletal and neurological system are involved in Behçet’s
syndrome. 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 IgG-4 related diseases [17].

3. Takayasu’s arteritis, giant cell arteritis

Takayasu’s arteritis, also called pulseless disease, involves the ascending aorta and aortic arch, and carotid and subclavian arteries, causing dilations and obstruction at the stage of healing and recurrences. CT angiography revealed the diseased lesion, thickened arterial wall in acute phase and aneurysmal or stenotic lesion in chronic phase [18]. The ultrasonography and MR angiography are also useful. In acute phase, the high signal of T2-weighted and/or STIR MR images and the increased uptake of 18Fluorodeoxy-glucose indicate the presence of active inflammation [19]. The mainstay of therapy for Takayasu’s arteritis is glucocorticoids. Giant cell arteritis (GCA) is a chronic vasculitis of large and medium sized vessels. The following classification criteria were as follows: 1) Age older than 50 years at onset, 2) Localized headache de novo, 3) Tenderness or decreased pulse of the temporal artery, 4) Erythrocyte sedimentation rate (ESR) greater than 50 mm/h, 5) Biopsy-proven necrotizing arteritis with multinucleated giant cells [20].

4. IgG4-related diseases: Chronic periaortitis

IgG4-related disease is a newly recognized syndrome of unknown etiology characterized by fibroinflammatory condition, in which tumefactive lesions, a dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells. Various symptoms are observed according to the lesions involved, although patients feel well at the time of diagnosis without fever. Seventy percent of patients have elevated serum IgG4 concentrations [17]. CT imaging features of arterial lesions are characterized by homogeneous wall thickening and enhancement in the late phases after contrast infusion accompanying the increase of connective tissue indicating sclerosing inflammation [21]. This indicates chronic periaortitis, which resembles the infected aneurysm with periaortic abscess (Figure 3). Therefore, the diseases should be differentiated from infected aneurysm by not only the imaging features but also clinical symptoms or negative blood cultures. Glucocorticoids typically the first line of therapy.

5. Conclusions

Due to the increase of aged patients with atherosclerosis, more attention should be paid to the endothelial damage of great vessels and an infected aneurysm should be properly diagnosed and carefully managed.
Figure 3. A case of chronic periaortitis with retroperitoneal fibrosis. A 75-year-old man presented with vague discomfort of lower abdomen. Panel A was at presentation. Two months later the symptoms remained unchanged with 8.5mg/dL of C-reactive protein but without fever. Repeated CT was performed (Panel B). Biopsy of the tissue revealed the infiltration of inflammatory cells and no bacteria (Panel C). Panel D was three weeks after the glucocorticoid therapy.

Disclosures

None.

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References


