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Immunoglobulin G4-related large thoraco-abdominal aortic aneurysm

Yuji Sekine*, Shin Yamamoto, Takuya Fujikawa and Shiro Sasaguri

Department of Aortic Surgery, Kawasaki Saiwai Hospital, Kawasaki, Japan

* Corresponding author. 31-27, Ohmiyacho, Saiwai ward, Kawasaki, Kanagawa 212-0014, Japan. Tel: +81-4-45444611; fax: +81-4-45494858; e-mail: yuji-sekine@kif.biglobe.ne.jp (Y. Sekine).

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Abstract

We report a case of immunoglobulin G4-related large thoraco-abdominal aortic aneurysm in a 38-year old man. Preoperative contrast-enhanced computed tomography revealed that the mid-descending thoracic aorta was extremely enlarged and the maximum diameter of the aneurysm was 92 mm. The patient underwent thoraco-abdominal aortic replacement through a thoraco-abdominal incision under left heart bypass. The postoperative pathological examination diagnosed immunoglobulin G4-related aortic aneurysm.

Keywords: Thoraco-abdominal aortic aneurysm • Immunoglobulin G4-related disease • Pathological examination

INTRODUCTION

Immunoglobulin G4-related disease (IgG4RD) is a novel pathological entity characterized by elevated serum IgG4 levels, significant thickening of the adventitia and extensive IgG4-positive plasma cell infiltration [1]. Although IgG4RD can cause any vascular system disorders, it mostly involves the abdominal aorta and retroperitoneal tissues [2, 3]. Few cases of IgG4-related thoracic aortic aneurysm have been reported [4]. Here, we present an extremely rare case of IgG4-related thoraco-abdominal aortic aneurysm.

CASE REPORT

A 38-year old man with no medical history of allergic disorders, such as bronchial asthma or drug allergies and no family history of cardiac sudden death or aortic event, was referred to our institution. He was diagnosed with large thoraco-abdominal aortic aneurysm by contrast-enhanced computed tomography at a previous institution. It was performed for differential diagnosis of the cause of chest discomfort. The aneurysm extended from the proximal descending thoracic aorta to the supraceliac abdominal aorta (Fig. 1A). The maximum diameter was 92 mm, and it had an eccentric thickened aortic wall (Fig. 1B). No abnormalities were observed on physical examination. Connective tissue disorders such as Marfan or Loeys–Dietz syndrome were not suspected from his figure and facial features. He was not hypertensive and the laboratory data revealed no specific results. At the operation, we could not dissect the left lung and descending thoracic aorta due to tight adhesion. Transected aneurysmal wall showed severe calcification and an extremely thickened mural thrombus. We performed graft replacement from the proximal descending thoracic aorta to the supraceliac abdominal aorta under the left heart bypass, which was established between left superior pulmonary

vein drainage and left femoral artery perfusion. Additionally, we reconstructed a pair of the ninth intercostal arteries, using the graft interposition technique. The aortic wall at the proximal and distal anastomotic sites was very fragile, thin and not atherosclerotic. The patient could be easily weaned from the left heart bypass and haemostasis was easily achieved. Surgically resected specimens were examined histopathologically. Elastica–Masson staining revealed marked fibrosis and significant infiltration of plasma cells in the adventitia and disrupted elastic fibres and smooth muscle in the media which was degenerative (Fig. 2A). Immunostaining revealed that the number of IgG4-positive plasma cells was ≥ 50 counts in all fields, and the ratio of IgG4-/IgG-related positive plasma cells was $\geq 50\%$ (Fig. 2B and C). Two weeks after the operation, the levels of serum IgG, IgG4 and anti-nuclear antibody were 1149 mg/dl (reference range 820–1740 mg/dl), 64 mg/dl (4–108 mg/dl) and 9.9 mg/dl (0.0–19.9 mg/dl), respectively. On the basis of the pathological examination, we confirmed IgG4-related thoraco-abdominal aortic aneurysm. The patient's postoperative course was uneventful. During the follow-up period of 2 years, there was no recurrence of IgG4-related aortitis and no extra-aortic manifestations were observed.

DISCUSSION

Inflammatory aneurysm has different clinical characteristics from atherosclerotic aneurysm and can be divided into two different clinical entities. IgG4-related aortic aneurysm is clinicopathologically different from non-IgG4-related aneurysm in some clinical features, which contain elevated serum IgG4 concentration, less frequency of symptomatic pain, low incidence of rupture and more frequency of allergic disease such as bronchial asthma. Serum IgG4 levels are elevated in $\sim 75\%$ of patients with IgG4-related disease [1]. However, the mechanisms underlying elevated serum

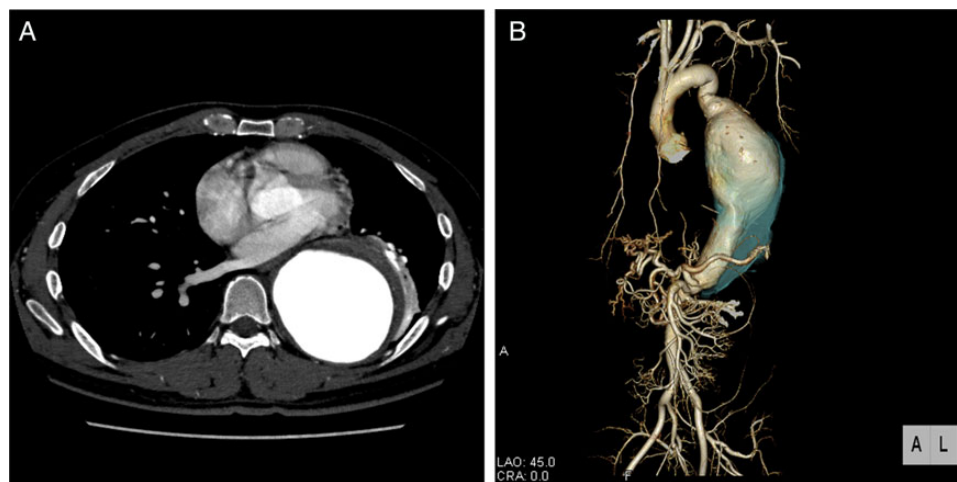


Figure 1: Preoperative enhanced computed tomography. (A) 3D computed tomography shows aortic aneurysm that extends from the proximal descending thoracic aorta to the supraceliac abdominal aorta. (B) The maximum diameter of the aneurysm is 92 mm at the level of the mid-descending thoracic aorta.

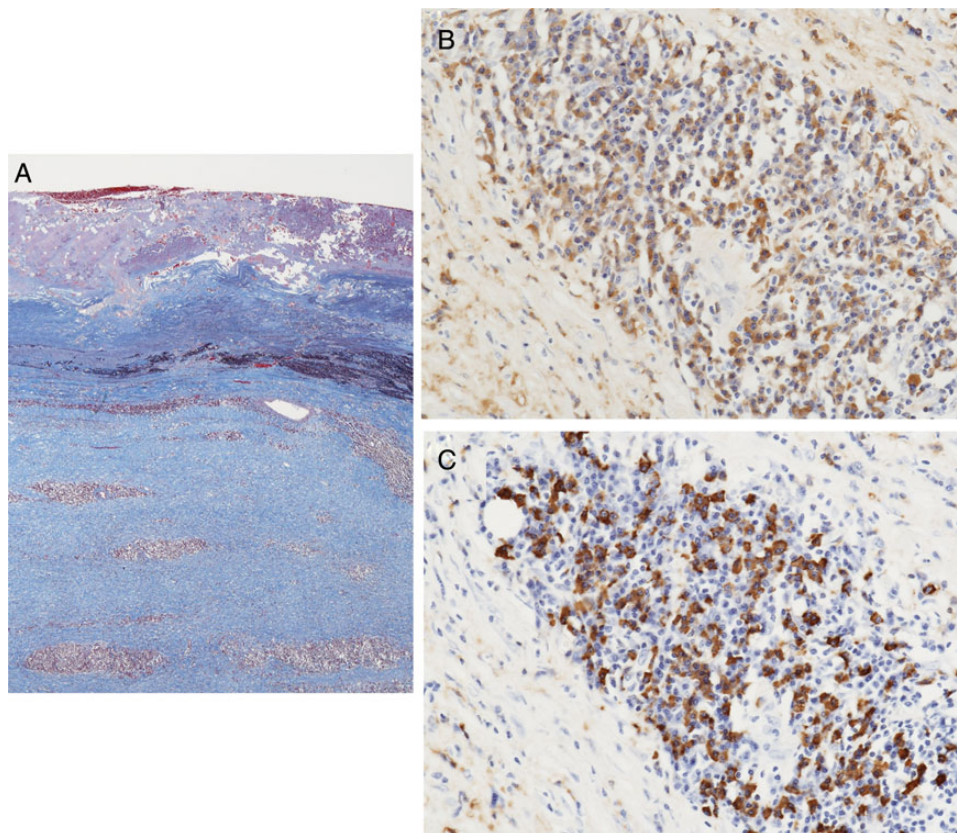


Figure 2: Histopathological examination of the transected aortic wall. (A) The aortic adventitia has the most severe inflammation and the media has disrupted elastic fibres and smooth muscle (Elastica-Masson, original magnification, $\times 20$). (B) Numerous IgG-positive plasma cells in the adventitia (IgG immunostain, original magnification, $\times 200$). (C) Numerous IgG4-positive plasma cells in the adventitia in almost the same field. The number of IgG4-positive plasma cells is more than 50 counts in this high-power field, and the ratio of IgG4/IgG-related positive plasma cells is $\geq 50\%$ (IgG4 immunostain, original magnification, $\times 200$). IgG: immunoglobulin G.

IgG4 concentrations and IgG4-positive plasma cell infiltration have not been elucidated.

Although some cases concerning IgG4-related abdominal aortic aneurysm have been reported [2, 3], only a few cases of IgG4-related thoracic aortic aneurysm have been reported [4]. Previously reported cases of IgG4-related thoracic aortic aneurysm were in the aortic arch or distal aortic arch and the aneurysmal figures were saccular [4]. In addition, IgG4-related aortic aneurysm is

more likely to be diagnosed in older male patients [1, 4]. Although the first-line strategy for extravascular lesions in IgG4RD is corticosteroid therapy, the efficacy of this strategy remains uncertain in IgG4-related aneurysm [1]. In this case, we selected surgical repair because the risk of aneurysmal rupture was very high and the patient was young. If we can diagnose this entity in early period pre-operatively, therapeutic option of steroid therapy can be selected more effectively [5]. In summary, to the best of our knowledge,

this is the first reported case of IgG4-related thoraco-abdominal aortic aneurysm. This clinical entity must be considered when making a differential diagnosis of undiagnosed aneurysmal pathology.

Conflict of interest: none declared.

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