

Incidentally detected aortitis during coronary bypass surgery: A case report

Koroner baypas ameliyatı sırasında tesadüfen tespit edilen aortit: Bir olgu sunumu

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ABSTRACT

Aortitis is the inflammation of the aortic wall. In our case, in which we planned elective coronary bypass surgery, a firm and dilated ascending aorta with a pearlescent color was encountered intraoperatively. Histopathological examination revealed extensive lymphoplasmacytic infiltration and storiform fibrosis. Aortitis may be a component of a multisystemic or autoimmune disease. The time of diagnosis may coincide with the asymptomatic period of the systemic disease. This case was presented as it was incidentally detected during coronary bypass surgery and was histopathologically diagnosed as immunoglobulin G4-related aortitis, although it could not be diagnosed in clinical and laboratory evaluations.

Keywords: Aortic surgery, aortitis, immunoglobulin G4-related disease.

Aortitis is inflammation of the aortic wall. The incidence is about 2.1 to 12%.^[1] Infectious and noninfectious conditions play a role in the etiology. The most commonly known causes are giant cell (temporal) and Takayasu arteritis.^[2] However, there may be noninfectious autoimmune causes, such as Behçet's disease, Reiter's syndrome, and Cogan syndrome. In addition, an involvement that causes aortitis can be observed in staphylococcal, mycobacterial, and syphilis infections.^[1,2]

Immunoglobulin (Ig) G4-associated aortitis and isolated thoracic aortitis, which are characterized

ÖZ

Aortit, aort duvarının enflamasyonudur. Elektif koroner baypas ameliyatı planlanan olguda ameliyat sırasında inci renginde, dilate ve sert kıvamda çıkan aort ile karşılaşıldı. Histopatolojik incelemede yaygın lenfoplazmositik infiltrasyon ve storiform fibroz saptandı. Aortit, multisistemik veya otoimmün bir hastalığın bir bileşeni olabilir. Kimi zaman tanı bu sistemik hastalığın asemptomatik safhasına denk gelebilir. Bu olgu koroner baypas cerrahisi sırasında tesadüfen saptanması ve klinik ve laboratuvar incelemelerde saptanamamasına rağmen histopatolojik olarak immunoglobulin-G4 ilişkili aortit tanısı konması nedeniyle sunuldu.

Anahtar sözcükler: Aort cerrahisi, aortit, immunoglobulin-G4 ile ilişkili hastalıklar.

by IgG4-dominated lymphoplasmacytic infiltration, have also been frequently diagnosed recently.^[3] It's a member of IgG4-related diseases with multisystemic involvement, such as IgG4-aortitis, autoimmune pancreatitis, sclerosing cholangitis, and retroperitoneal fibrosis. Histopathological examination is the gold standard in the diagnosis with components such as lymphoplasmacytic infiltration, lymphoid follicle formation, obliterative phlebitis, and storiform fibrosis.^[3] Increased serum IgG4 levels, positron emission tomography imaging, and 18-fluorodeoxyglucose involvement in scintigraphic

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examination play a prominent role in diagnosis and follow-up.^[3,4]

Idiopathic isolated aortitis generally has a subclinical nature and is often incidentally diagnosed in aneurysm surgery by histopathological examination.^[2] Microscopically, lymphoplasmacytic infiltration is in the media and adventitia layers with varying densities and is dominated by IgG.^[3] Idiopathic isolated aortitis incidence is about 4 to 47% and is divided into two groups: isolated thoracic aortitis and chronic periaortitis, which mostly involves the abdominal aorta.^[3]

The first-line treatment is systemic steroid use, followed by chemotherapeutic agents and monoclonal antibodies. Surgical planning is made in accordance with cardiac pathology during the disease process.^[1-4]

In this case report, we aimed to share our experience in the diagnosis and treatment of a rare pathology in which the surgical procedure was altered intraoperatively.

CASE REPORT

A 53-year-old male patient was admitted to the cardiology clinic with symptoms of stable angina. Three-vessel coronary artery disease was detected as a result of angiographic evaluation, and an elective coronary artery bypass operation was planned. No comorbid systemic disease was detected in preoperative preparations. A chest roentgenogram revealed no abnormality in the aortic silhouette. In the echocardiographic evaluation, the ejection fraction was 55%, the valve structures were functional, and the diameters of the aortic root, ascending aorta (33 mm), and heart chambers were within normal limits. No additional imbalance was detected in laboratory tests.

In the operation, the pericardium was opened after median sternotomy. It was observed that the ascending aorta was firm, fibrotic, dilated, and of a pearlescent color (Figure 1a). The right femoral artery and right atrium were cannulated for cardiopulmonary bypass. The cross-clamp was placed in the clean area proximal to the aortic arch, and then an aortotomy incision was

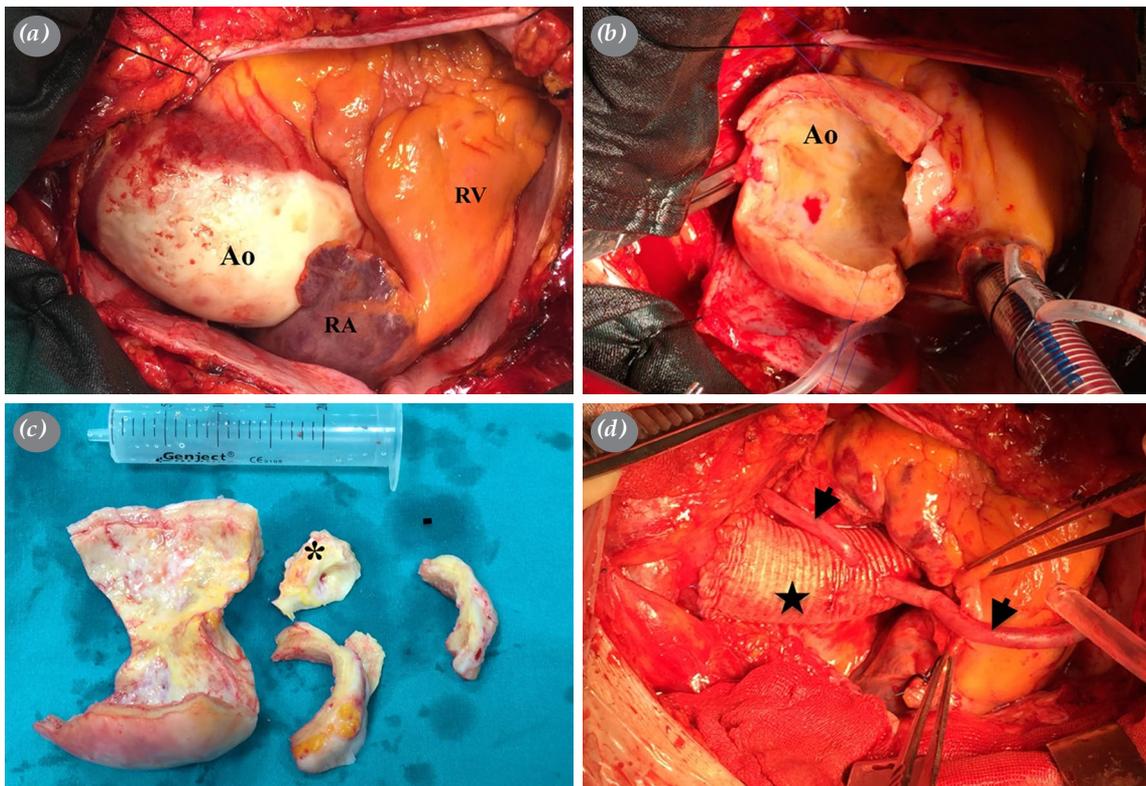


Figure 1. (a) Intraoperative view of the heart. (b) Intraluminal view of the aorta. (c) Macroscopic view of the excised ascending aorta and coronary ostia (asterisk). (d) Postoperative view of the heart, composite valve graft conduit (star), and saphenous vein graphs (arrow heads).

Ao: Aorta; RA: Right atrium; RV: Right ventricle.

performed. We observed that after aortotomy, the aortic wall thickening progressed towards the aortic root, including the coronary ostia, and there was also a thickening of the valves. It was observed that the aortic wall was approximately 15 mm thick (Figures 1b and c). Three-vessel coronary bypass surgery and the Bentall procedure was performed with a No. 23 mechanical composite valve graft conduit (St. Jude Medical Inc., Saint Paul, MN, USA) (Figure 1d). There were no complications in the postoperative period.

Postoperative computed tomography revealed thickening of the arch and descending aortic wall. It was observed that the coronary grafts were open. Echocardiographic examination revealed mild mitral insufficiency with 55% of the ejection fraction with a functional prosthetic aortic valve.

In histopathological examination, an intense lymphoplasmacytic infiltration accompanied by

storiform fibrosis and accompanying lymphoid follicles were observed (Figure 2a). In addition, endothelial swellings in the vasa vasorum and lymphoplasmacytic infiltration in the vascular walls were detected (Figures 2b and c). The storiform fibrosis was highly remarkable in staining with Masson trichrome (Figure 2d). Immunohistochemical examination with IgG showed diffuse staining in plasma cells (Figure 3a). Immunoglobulin G4 staining in this area was evaluated as less than 50 cells at $\times 400$ magnification (Figure 3b).

No surgical pathology or clinical complaints were found in the five-year follow-up of the patient. Syphilis and other infectious causes were ruled out with negative blood and microbiology samples. Serum IgG and IgG4 levels were normal, and there was no other organ involvement.

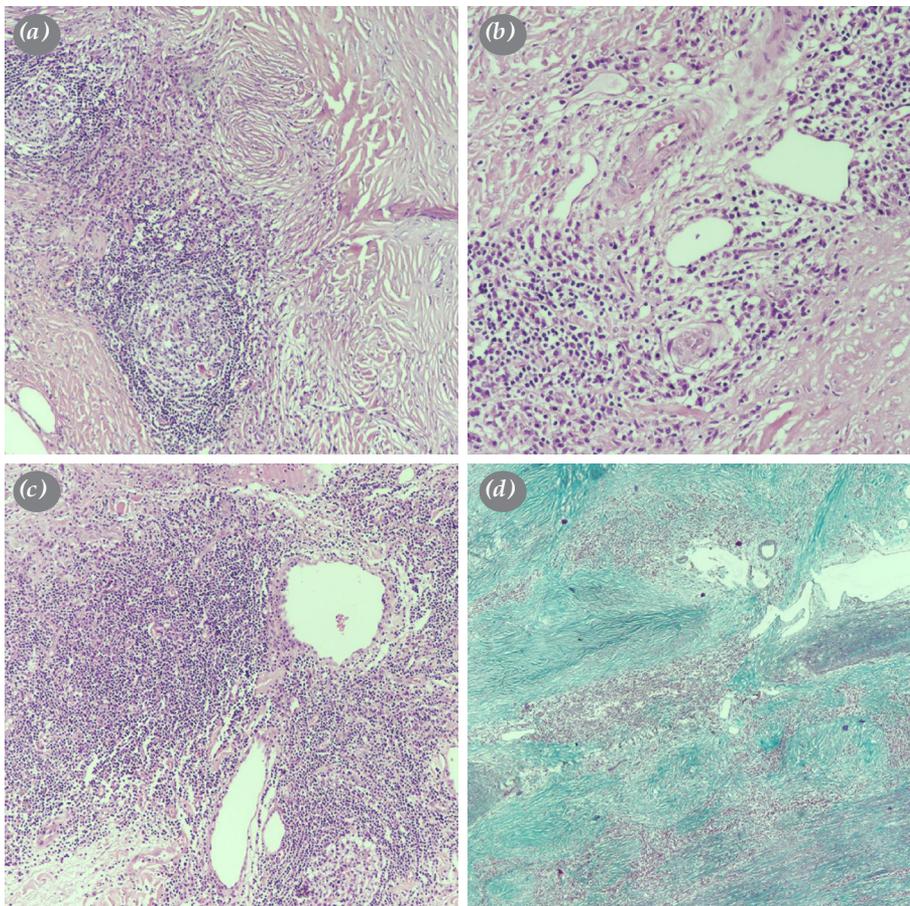


Figure 2. (a) Hematoxylin and eosin (H&E)-stained section demonstrating lymphoid follicle formation and storiform fibrosis. (b) Hematoxylin and eosin-stained section at $\times 200$ magnification. (c) Endothelial swellings and lymphoplasmacytic infiltration in the vascular wall (H&E, $\times 100$). (d) Section showing storiform fibrosis (Masson trichrome, $\times 40$).

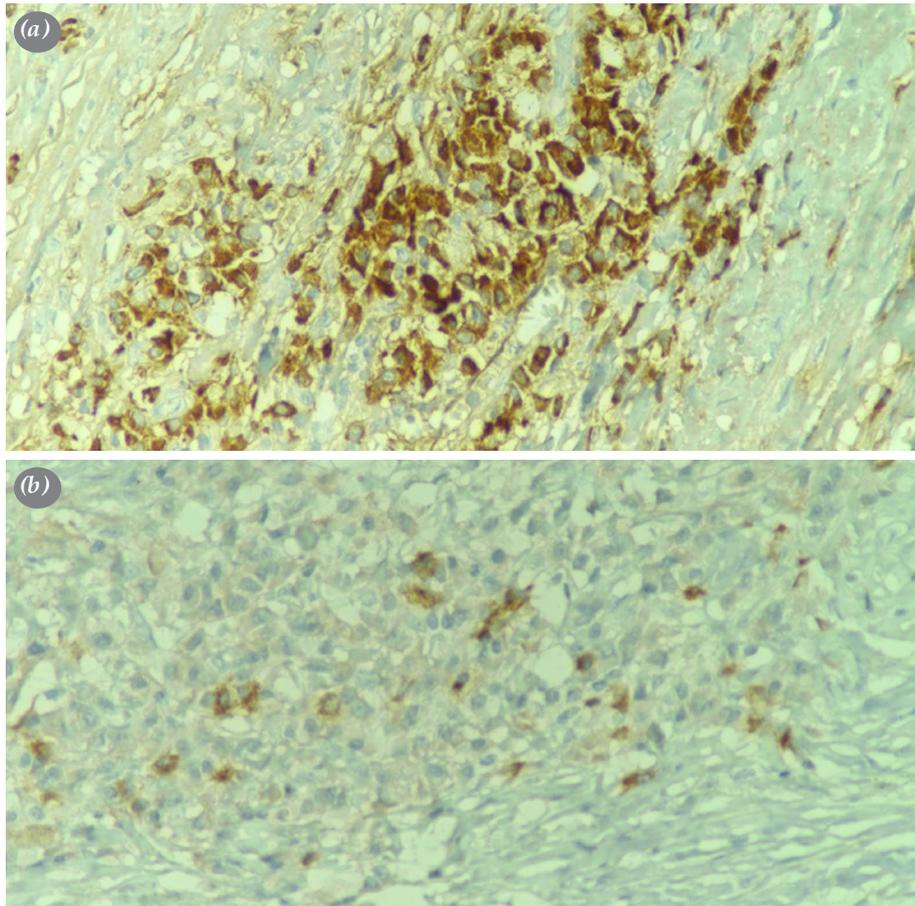


Figure 3. (a) Diffuse IgG staining in plasma cells (IgG immunostaining, $\times 400$). (b) Immunoglobulin G4 staining with less than 50 cells in high power field (IgG immunostaining, $\times 400$).

DISCUSSION

In most of the surgical procedures performed for aortic aneurysms, although there is no underlying systemic disease, there is incidental evidence of aortitis. Idiopathic isolated aortitis generally has a subclinical nature.^[2] We incidentally detected aortitis in our case, in which we planned elective isolated coronary artery surgery.

In our case, there was no aortic valve pathology detected by preoperative imaging methods. However, we observed that the aortic wall thickened after aortotomy progressed towards the aortic root, including the coronary ostia, and there was also a thickening of the valves. We did not know whether the disease was present in the aortic root and aortic valve or whether it would progress. Cardiopulmonary bypass was continued, and the Bentall procedure was applied, considering that waiting for the intraoperative histopathological evaluation would be to the detriment

of the patient. Macroscopically, there was no thickening of the arch and descending aorta, and it looked normal. In addition, since we did not detect dissection, ulcer, or hematoma on the inner surface of the resected aortic wall, we did not intervene in the aortic arch and its distal. No sample was taken from the intact area for pathological examination. Therefore, we cannot comment on whether there is inflammation at a microscopic level even though it is macroscopically normal.

Immunoglobulin G4-related diseases have an indolent character, may take a long time to manifest symptoms, and may present with a myriad of different clinical manifestations. Primary and secondary IgG4-related vasculopathies may present with aortic aneurysm and dissection.^[4] Immunoglobulin G4-associated aortitis is a rare form of aortitis and usually occurs after 50 years of age.^[5] It can cause sialadenitis, pancreatitis, sclerosing cholangitis, and

retroperitoneal fibrosis with systemic inflammatory response. An increase in the level of serum IgG4, histopathological confirmation of fibrosis, an IgG4/IgG ratio of more than 50%, and more than 50 IgG4 positive plasma cells at $\times 400$ magnification are required for diagnosis.^[5]

As it is a newly recognized disease, the diagnosis can be missed in the first pathological evaluation, and the diagnosis is made in the re-evaluation. Occasionally, nonspecific increases in new lesions may explain the progression of IgG4 with the process. Delay in diagnosis may lead to underestimation of the disease. There is some evidence that the presence of IgG4 is a premalignant or paraneoplastic condition. Therefore, cancer and infections should be considered in differential diagnosis.^[6]

Isolated thoracic aortitis etiopathogenesis is unclear. Laco *et al.*^[3] did not detect any IgG4-related disease in their series of 11 cases. Histopathologically, lymphoplasmacytic infiltration and fibrosis were found in varying intensity. Obliterative or obstructive phlebitis was not detected in the vasa vasorum. The IgG4/IgG ratio of the cases ranged between 0.07 and 0.98. Increased fibrosis observed in advanced or chronic disease stages may cause IgG4-positive plasma cells to appear less frequently. The end-stage immunoglobulin G4-related aortitis may not fit histopathological diagnosis criteria as the inflammation profile mostly turns to fibrosis. Consequently, it is thought that idiopathic thoracic aortitis may be a subtype or variant of IgG4-related diseases.^[3] In our case, fibrosis was widespread, IgG4-positive staining ratio may be diminished for this reason.

In our case, syphilis and other infectious causes were ruled out with negative blood and microbiology samples. With normal serum IgG and IgG4 levels and the absence of other organ involvement, the diagnosis of IgG4-related disease remained in the background. However, histopathological examination revealed dense lymphoplasmacytic infiltration with lymphoid follicles that involved the vessel walls, endothelial swelling in the vasa vasorum, and storiform fibrosis. Consequently, the patient was diagnosed with IgG4-aortitis according to the international consensus guidelines for the pathological diagnosis of IgG4-related aortitis.^[5]

Löffler *et al.*^[6] stated in their study that IgG4 positivity may not always be present in aortitis, dissection, and aneurysm, and sampling from adventitia should be performed to evaluate infiltration. They predicted that aortic elasticity might decrease due to this external-to-internal infiltration mechanism.

Therefore, we may speculate that patients with this pathology are prone to aortic aneurysm, dissection, or rupture, and replacement of the aorta can be recommended to prevent fatal complications.

In conclusion, it should be kept in mind that a patient presenting with isolated aortitis may be a variant of a multisystemic disease in the follow-up and treatment. The severity and stage of the disease vary depending on many factors, and it is not exactly known what triggers or slows the disease down. Surgical resection of diseased segments of the aorta can be beneficial for the differential diagnosis and prevention of severe complications.

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