Aort the pearl, incidentally detected aortitis during coronary bypass surgery: A case report

Abdul Kerim Buğra[1], Aytul Buğra[2], Ersin Kadirogullari[1], and Burak Onan[1]

[1]Mehmet Akif Ersoy Thoracic and Cardiovascular Research and Education Hospital
[2]Council of Forensic Medicine

March 30, 2022

Abstract

Aortitis, in its simplest definition, is an inflammation of the aorta. It can be divided into two groups as infectious and non-infectious. Noninfectious aortitis can be an involvement of multisystemic and autoimmune diseases as well as being diagnosed incidentally. In our case, in which we planned elective coronary bypass surgery, we encountered an ascending aorta with a pearlish color, dilated and firm consistency intraoperatively. Histopathological examination showed extensive lymphoplasmocytic infiltration and strotiform fibrosis. In the laboratory tests performed for etiology in the postoperative period, no abnormalities were observed in the early and long term. Even when detected isolated and incidentally, aortitis may be a component of a multisystemic and/or autoimmune disease. The time of diagnosis may coincide with the asymptomatic period of the systemic disease. We wanted to present this case because it was detected incidentally during coronary bypass surgery and was diagnosed histopathologically immunoglobulin-G4 related aortitis, although it was not found in clinical and laboratory evaluations.

1.Introduction

Aortitis is aortic wall inflammation and histopathological examination is the gold standard in diagnosis. The incidence is about between 2.1 and 12%. Infectious and non-infectious conditions plays role in the etiology. The most common known causes are giant cell (temporal) and Takayasu arteritis. However, there may be autoimmune noninfectious causes such as Behçet's disease, Reiters and Cogan syndrome. In addition, involvement that causes aortitis can be seen in staphylococcal, salmonella, mycobacterial and syphilis infections[1–3].

Immunglobulin (Ig) G4 associated aortitis and isolated thoracic aortitis, which are characterized by IgG4-dominated lymphoplasmocytic infiltration, are also frequently diagnosed recently[4]. It is a member of the family 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, and it has components such as lymphoplasmocytic infiltration, lymphoid follicle formation, obliterative phlebitis and strotiform fibrosis. Increased serum IgG4 levels, positron emission tomography imaging and 18-fluorodeoxyglucose involvement in scintigraphic examination play an important role in diagnosis and follow-up[4–6].

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

The treatment usually involves initial systemic steroid use, and secondary chemotherapeutic agents and monoclonal antibodies. Surgical planning is made in accordance with cardiac pathology during the disease process[1,2,4,6,7].

2.Case

A 53-year-old male patient was admitted to the cardiology clinic with a complaint of stable angina. Three-vessel coronary artery disease were detected as a result of angiographic evaluation and elective coronary artery bypass operation was planned. No co-morbid systemic disease was detected in his preoperative preparations. Chest roentgenogram showed no abnormality of aortic silhouette. In echocardiographic evaluation, it was observed that the ejection fraction was 55%, the valve structures were normal and normofunctional, and the diameters of the aortic root, ascending aorta, and heart chambers were within normal limits. No additional imbalance was detected in laboratory tests.

In the operation, the pericardium was opened after the median sternotomy. It was observed that the ascending aorta was firm, fibrotic, dilated and of a pearlish 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 performed. It was observed that the aortic wall was approximately 15 mm thick and had continuity including the coronary ostia(Figure 1b-c). Three-vessel coronary bypass surgery and Bentall procedure performed with 23 no mechanical composite valve graft conduit (St Jude Medical Inc., USA)(Figure 1d). There were no complications in the postoperative period.

Post operative 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 normofunctioned prosthetic aortic valve.

In histopathological examination, an intense lymphoplasmocytic infiltration accompanied by strotiform fibrosis and accompanying lymphoid follicles were observed (Figure 2a). In addition, endothelial swellings in the vaso vasorum and lymphoplasmocytic infiltration in the vascular walls were detected(Figures 2b, 2c). Histochemically, strotiform fibrosis was highly remarkable in staining with Masson trichrome(Figure 2d). Immunhistochemical examination with IgG showed diffuse staining in plasma cells(Figure 3a). IgG4 staining in this area was evaluated as less than 50 cells at x400 magnification(Figure 3b).

No surgical pathology or clinical complaints were found in the 5-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 another organ involvement.

3.Discussion

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

IgG4-related diseases have an indolent character, may take a long time to manifest symptoms, and may present myriad different clinical manifestations. Primary and secondary IgG4-related vasculopathies may present with aortic aneurysm and dissection[6]. IgG4-associated aortitis is a rare form of aortitis and usually occurs after 50 years of age. It can cause sialadenitis, pancreatitis, lymphadenopathy, sclerosing cholangitis, nephritis, retroperitoneal fibrosis with systemic inflammatory response. For diagnosis an increase in the level of serum IgG4 and, histopathologically, confirmation of fibrosis, IgG4/IgG ratio should be more than 50%, more than 50 IgG4 positive plasma cells should be seen at x400 magnification[8].

Because 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. Sometimes, nonspecific increases in new lesions may explain the progression of IgG4 with the process. Delay in diagnosis may lead to underestimation of the disease[7].

There is some evidence that the presence of IgG4 is a premalignant or paraneoplastic condition[9]. Therefore, cancer and infections should be considered in differential diagnosis[10].

Isolated thoracic aortitis etiopathogenesis is unclear. Laco[4] et al. did not detect any IgG4-related disease in their series of 11 cases. Histopathologically, lymphoplasmocytic infiltration and fibrosis were found varying intensity. Obliterative or obstructive phlebitis was not detected in the vasa vasorum. The IgG4/IgG ratio of the cases ranged from 0.07 to 0.98. Increased fibrosis seen in advanced or chronic disease stages may cause IgG4 + plasma cells to appear less frequently. Even if the diagnosis gets difficult histopathologically, may also indicate an end stage disease. For these reasons, it is thought that idiopathic thoracic aortitis may be a subtype / variant of IgG4-related diseases[4,5]. In our case, fibrosis was widespread, IgG4 (+) staining ratio may be less 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 accompanying other organ involvement, the diagnosis of IgG4-related disease remained in the background. However, histopathological examination revealed dense lymphoplasmocytic infiltration with lymphoid follicles, endothelial swelling in the vasa vasorum and lymphoplasmocyte infiltration that infiltrated the vessel walls, accompanied by stroriform fibrosis. According to international consensus guideline for pathological diagnosis of IgG4 related aortitis, the diagnosis of IgG4-associated aortitis was considered in the foreground[8].

Löfler et al.[10] stated in their study that IgG4 positivity may not always be seen in aortitis, dissection and aneurysm development increased in the presence of IgG4, and sampling from adventitia should definitely 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 may be prone to develop aortic aneurysm, dissection or rupture and replacement of the aorta can be recommended to prevent fatal complications.

5.Conclusion

Thanks to the developing medicine and technology, rare diseases are diagnosed more frequently and can be treated more effectively. 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. It is not known exactly what triggers or slows them down. Surgical resection of diseased segment of the aorta can be beneficial for the differential diagnosis and prevention of severe complications.

Declaration of conflicting interests: None

Funding: None

Author contributions

All authors have an equal contribution to the design of the study, the interpretation of the data, the preparation of the draft, and the final approval of the article.

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

References

1. Erbel R, Aboyans V, Boileau C, Bossone E, Di Bartolomeo R, Eggebrecht H, et al. 2014 ESC guidelines on the diagnosis and treatment of aortic diseases.Eur Heart J.2014;35(41):2873–926.

2. Gornik HL, Creager MA. Aortitis. Circulation. 2008;117(23):3039–51.

3. Pérez-García CN, Olmos C, Vivas D, Ferrera C, García-Arribas D, Enríquez-Vázquez D, et al. IgG4-aortitis among thoracic aortic aneurysms. Heart. 2019;105(20):1583–9.

4. Laco J, Steiner I, Holubec T, Dominik J, Holubcova Z, Vojacek J. Isolated thoracic aortitis: Clinicopathological and immunohistochemical study of 11 cases. Cardiovasc Pathol. 2011;20(6):352–60.

5. Agaimy A, Weyand M, Strecker T. Inflammatory thoracic aortic aneurysm (lymphoplasmacytic thoracic aortitis): A 13-year experience at a German Heart Center with emphasis on possible role of IgG4. Int J Clin Exp Pathol. 2013;6(9):1713–22.

6. Perugino CA, Wallace ZS, Meyersohn N, Oliveira G, Stone JR, Stone JH. Large vessel involvement by IgG4-related disease. Med (United States). 2016;95(28).

7. Weiss MA, Aubry MC, Pflederer BR. A Case of IgG4-Related Aortitis and Pericarditis: Diagnostic Challenges and Natural History. Am J Case Rep. 2018;19:1232–6.

8. Stone JR, Bruneval P, Angelini A, Bartoloni G, Basso C, Batoroeva L, et al. Consensus statement on surgical pathology of the aorta from the Society for Cardiovascular Pathology and the Association for European Cardiovascular Pathology: I. Inflammatory diseases. Cardiovasc Pathol. 2015;24(5):267–78.

9. Tioni C, Antonutti M, Nora C Di, Proclemer A. Never forget the aorta: A case report of IgG4-related disease causing aortitis. Eur Hear J - Case Reports. 2018;2(4):1–5.

10. Löffler C, Hoffend J, Rebel M, Waldherr R, Uppenkamp M, Bergner R. A rare cause for lower back pain: a case of an IgG4-related periaortitis. Clin Rheumatol. 2016;35(1):265–70.

Figure legends:

Figure 1: A.Intraoperative view of heart. Ao: aorta,RA:right atrium,RV:right ventricule, B.Intraluminal view of aorta. Ao:aorta, C.Macroscopically view of excised ascending aorta and coronary ostia(asterisk), D.Postoperative view of heart. Composite valve graft conduit(star) and saphenous vein graphs(arrow heads).

Figure 2: A.(H&E, x100):lymphoid follicule formation and storiform fibrosis, B.(H&E, x200), C.(H&E, x100):endothelial swellings and lymphoplasmocytic infiltration in the vascular wall, D.(Masson trichrome, x40):storiform fibrosis

Figure 3: A.(IgG immunstaining, x400), diffuse IgG staining in plasma cells, B.(IgG4 immunstaining, x400):IgG4 staining was less than 50 cells in HPF