A Thoracoabdominal Aorta Aneurysm Patient with Severe Calcification in Arch of Riolan

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Abstract

Background: The co-existence of thoracoabdominal aortic aneurysm and arch of Riolan has not been reported yet. This article is to describe a rare phenomenon of severe calcification in arch of Riolan, as well as other anatomic variation in arteries and veins in the same patient.

Methods and findings: A 53 year old man with abdominal wall varicosis was reported with a thoracoabdominal aorta aneurysm and severe stenosis of celiac trunk and superior mesenteric artery. There were multiple calcification and non-calcified plaques inside the wall of abdominal aorta and its branches, including arch of Riolan. A Budd-Chiari Syndrome was also reported. The intra-hepatic inferior vena cava was narrow and infra-hepatic inferior vena cava was enlarged with multiple intra-cavity calcifications. His right renal vein was drained into supra-hepatic inferior vena cava. Two cover stents were implanted near ostia of renal vessels. The aneurysm was cured and patient recovered uneventfully.

Conclusion: A combination of thoracoabdominal aorta aneurysm and arch of Riolan is rarely reported. Hemodynamic factor may explain why severe calcified plaques only exist in arch of Riolan but not in other branches of aorta. Calcification of inferior vena cava is extremely rare in adults, and a possibly relevant situation is structural anomalies.

Keywords: Thoracoabdominal aorta aneurysm; Arch of Riolan; Inferior vena cava; Vascular calcification; Budd-Chiari syndrome.

Abbreviations

AOR: Arch of Riolan; BCS: Budd-Chiari Syndrome; IVC: Inferior Vena Cava; IMA: Inferior Mesenteric Artery; SMA: Superior Mesenteric Artery; TAAA: Thoracoabdominal Aortic Aneurysm

1. Introduction

The thoracoabdominal aortic aneurysm (TAAA) is a kind of aneurysm extending from the thoracic aorta to the abdominal aorta [1]. Anastomosis of Riolan or arch of Riolan (AOR) has been well described anatomically [2]. However, the co-existence of AOR and TAAA has not been reported in published articles. The case we presented here was quite unique, featuring the severe calcification in AOR and Inferior Vena Cava (IVC) and numerous anatomic variations in arteries and veins. All these observations possibly indicated new etiology of aortic aneurysms.

2. Case Report

2.1 Methods

A 53 year old man with abdominal wall varicosis for 20 years was reported with a Thoracoabdominal Aorta Aneurysm (TAAA) during pre-operative assessment of an aortic valve replacement surgery. He complained no abdominal pain or any other discomfort. Computed Tomography Angiography (CTA) revealed an aneurysmal dilatation at level of 12th thoracic vertebrae and the maximum diameter was 5 cm. Severe stenosis of celiac trunk and Superior Mesenteric Artery (SMA) was illustrated as well. A curve communicating branch connecting SMA and Inferior Mesenteric Artery (IMA) was reported as well. A curve communicating branch connecting SMA and Inferior Mesenteric Artery (IMA) was reported as well, which is also known as AOR or “The Meandering Mesenteric Artery” (Figure 1) [2]. There were multiple
calcification and non-calcified plaques inside the wall of abdominal aorta and its branches. A Budd-Chiari Syndrome (BCS) was also reported in CTA. The intra-hepatic IVC was narrow and infra-hepatic IVC was enlarged with multiple intra-cavity calcification (Figures 2A and 2B). Anatomic variation in veins was also observed. The left renal vein was drained into left hemi-azygous vein, and the right renal vein was drained into supra-hepatic IVC.

As for history, the patient was a heavy smoker and was diagnosed with coronary heart disease and severe aortic valve insufficiency three years ago. The aortic valve was replaced recently. Family history of aorta aneurysm and other congenital vascular diseases was negative.

3. Results

According to patient history and CT scan, we diagnosed the patient with TAAA and BCS. Pre-operative evaluations showed no main indication for intervention or surgical treatment, therefore the patient took an endovascular graft exclusion therapy for TAAA. Two cover stents (Microport 36-36-80, 34-34-80) were implanted near ostia of renal vessels under digital subtraction angiography (DSA) (Figure 3). CTA at 6 month follow-up indicated the stent in proper position and blood flowing smoothly inside the stent and renal arteries.

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

4. Discussion

There are several points worth discussing in this case: 1) the reason why the severe calcification plaques perform only in AOR but not in iliac arteries; 2) possible association between calcification of arteries and veins; and 3) the reason of the co-existence of BCS.

4.1 Calcified plaques in TAAA and AOR

As demonstrated in the CTA image, AOR formed due
to severe stenosis of celiac trunk and SMA, which provided blood supply to colon and rectum. It is well accepted that atherosclerosis is one of the main risk factors of TAAA, which explains the atherosclerosis inside the aneurysm. The calcification of AOR and part of abdominal aorta that forming aneurysm indicate chronic inflammation of arteries, which is supported by both animal model study and human imaging study [3,4]. Atherosclerosis is the most common source of chronic vascular inflammation, and accumulation of oxidized lipids is the main underlying reason. Pro-oxidant lipids positively regulate vascular calcification, and vice versa [4]. As for the location of calcification, it occurs in arteries of all sizes including the aorta. Types of vascular calcification include intimal calcification relevant to atherosclerosis, medial calcification secondary to diabetes mellitus and chronic kidney disease, and rare genetic disorders [5]. Calcification of the aorta is correlated with atherosclerosis, coronary artery disease and carotid atherosclerosis [6].

However, what it does not explain is that atherosclerosis of AOR seems more severe than that of aorta where aneurysm occurred. Besides, other main arteries such as iliac arteries are quite smooth with no calcification. One possible explanation is hemodynamic factor. The AOR is long and tortuous, forming turbulence in vascular walls, and causing more atherosclerotic and calcium deposit inside. This hypothesis is supported by some flow studies. One shows that secondary aortic flow patterns (vortex or helix flow) induce shear force alterations, which induces the development of atherosclerosis [7].

Another study about Fas system may explain the association of turbulence and atherosclerosis. Turbulence condition leads to expression of Fas-receptor, which increases the amount of apoptotic cells, and thus plays a role in progression of atherosclerosis [8].

4.2 Calcification in IVC

IVC calcification is extremely rare in adults, and is associated with recurrent pulmonary embolism and antiphospholipid syndrome [9]. IVC calcification was mostly described in newborns and can hardly explain the situation of this patient and the mechanism remains unclear. Among those cases, a possibly relevant situation is structural anomalies. As described before, the patient has anatomic variation in veins. His right renal vein was drained into suprahepatic IVC. Whether this kind of variation lead to intra-cavity calcification of IVC is doubtful.

4.3 BCS and TAAA

As for the co-existence of BCS, there is rare association between TAAA and BCS and no evidence was put forward to explain the combination. Batur et al. [10] reported a rare association of BCS and renal arterial aneurysms and the disease underlying was behcet. The case we presented, however, did not fit the diagnosis of behcet by any measure. Thus, we state that this is an interesting coincidence, considering the patient’s history and the pathogenesis of the two diseases.

5. Conclusion

A combination of thoracoabdominal aorta aneurysm and arch of Riolan is rarely reported. Hemodynamic factor may explain why severe calcified plaques only exist in arch of Riolan but not in other branches of aorta. Calcification of inferior vena cava is extremely rare in adults and a possibly relevant situation is structural anomalies.

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