

Porcelain Aorta and it's Rheumatological Significance

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Received: May 05, 2017; **Published:** May 23, 2017

Abstract

Porcelain aorta is a harbinger of widespread atherosclerotic process and its identification may be associated with higher cardiovascular and surgical mortality. Patients with porcelain aorta requiring cardiac surgery need specific alterations in approach to the surgery especially with regard to aortic cross – clamping. However, its association with systemic inflammatory bone and joint disease is an orthopedic concern. Detection of asymptomatic porcelain aorta in rheumatoid arthritis may have therapeutic concerns also.

Keywords: Porcelain Aorta; Aortic Stenosis; Systemic Inflammatory Disorders; Rheumatoid Arthritis

Case Report

A 70 year old gentleman was admitted in the emergency with syncope. There was history of similar several episodes of syncope over the past 2 months. His ECG showed complete heart block with ventricular escape rate of only 30 beats per minutes. He was urgently given a temporary cardiac pacemaker under fluoroscopy. Under fluoroscopy, he also demonstrated the dense circumferential calcification outlining the thoracic and abdominal aorta (Figure 1 and Video 1). His subsequent entire workup for valvular heart disease, atherosclerotic coronary artery disease, peripheral arterial disease and systemic inflammatory bone and joint disease was found negative. He was given a permanent pacemaker and was discharged from the hospital.

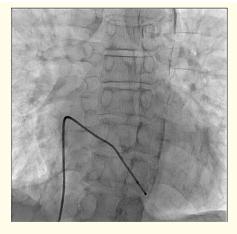


Figure 1: Fluoroscopic image showing porcelain aorta. Note the temporary pacemaker lead going inside the heart in the right ventricle. Aortic valve calcification is also present, however it is better appreciated in video 1. Some scoliosis is also there but it was positional.

Video 1: Cineangiographic run showing porcelain aorta in its entire length. Calcification of aortic valve is also better appreciated.

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Discussion

Porcelain aorta is a rare entity characterized by extensive partial or full circumferential calcification of aorta [1]. It is seen more common in elderly and is a marker of extensive atherosclerotic process. Its prevalence is has only continued to increase with increased prevalence of coronary artery disease. It is also associated with severe aortic stenosis and its relevance and complications during the surgical or percutaneous approach to replacement of calcific aortic valve.

Porcelain aorta is commonly diagnosed at the time of cardiac surgery on palpation of aorta before aortic cross clamping precluding its use if present [2]. More commonly it is diagnosed as an incidental finding on chest roentgenogram or as diffuse calcific outline of aorta on fluoroscopy while performing coronary angiography (Figure 1 and Video 1). Hence, most patients with porcelain aorta are asymptomatic and true incidence is difficult to quantify.

The pathophysiology of porcelain aorta is not fully understood; however, presence of extensive atherosclerosis is considered as a harbinger of calcification. Among the causes of such porcelain, traditional risk factors for atherosclerosis such as smoking, diabetes mellitus, hypertension, dyslipidemia and aging have been implicated as risk factors. Specific subsets have been found to be associated with porcelain aorta such as chronic kidney disease [3], radiation exposure [4].

Systemic inflammatory disorders like Takayasu arteritis, SLE and rheumatoid arthritis have also been associated with porcelain aorta [5]. One study by Paccou., *et al.* [6] demonstrated higher incidence of coronary and abdominal aortic calcification in patients with rheumatoid arthritis as compared to age and sex matched controls. Importantly, they also found that erosive arthritis is a major determinant of abdominal aortic calcification. These findings implicate that systemic inflammatory responses contribute to development to extensive calcification though further research is implicated in identifying the pathophysiologic mechanisms.

Increasing use of chimeric monoclonal antibodies like Rituximab for treatment of rheumatoid arthritis and other autoimmune diseases suggest investigations for rule out asymptomatic coronary artery disease as the use of Rituximab has been shown to be associated with cardiac adverse reactions in the form of myocardial infarction [7], coronary vasospasm [8], polymorphic ventricular tachycardia [9] and non-ischemic cardiomyopathy [10].

Specific skeletal abnormalities have not been found to be associated with porcelain aorta or dense aortic calcification.

Conclusion

Porcelain aorta is a harbinger of widespread atherosclerotic process and its identification may be associated with higher cardiovascular and surgical mortality. Patients with porcelain aorta requiring cardiac surgery need specific alterations in approach to the surgery especially with regard to aortic cross – clamping. However, its association with systemic inflammatory bone and joint disease is an orthopedic concern and demands its full workup.

Conflict of Interest Statement

There is no conflict of interest.

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