Introduction

The left atrial calcification is a relatively uncommon complication of long-standing rheumatic valvular heart disease [1]. Porcelain atrium is a massive calcification involving the left atrial appendage, the free wall of the left atrium, and the mitral valve apparatus except for interatrial septum [2,3]. Whereas, coconut atrium involves all areas of left atrium including the interatrial septum [4-14].

Giant left atrium is usually the result of rheumatic heart disease. It implies primary rheumatic involvement of the left atrial wall as part of the rheumatic pancarditis and always associated with atrial fibrillation [15]. Calcification of such an extent is very rare as is the co-existence of a very large vascularized thrombus in a patient of rheumatic mitral stenosis.

Due to the rarity of this association, we present here-in the angiographic appearance of a large vascularised left atrial thrombus, and the surgical details in a surviving patient who underwent mitral valve replacement using a St. Jude mechanical prosthesis and a brief review of the literature.

Case Presentation

A 58-year-old woman was admitted at our institution in February 2014 with chronic atrial fibrillation and congestive heart failure (New York Heart Association Class-IV). She had two episodes of transient loss of consciousness with no neurologic deficits.

Notable clinical findings included an intermittently irregular pulse, blood pressure of 90/60 mmHg, cardiomegaly with a single and accentuated second heart sound and a low-pitched diastolic murmur at the apex consistent with mitral stenosis. Also, there was a holosystolic murmur at the tricuspid area, features of pulmonary edema and hepatomegaly. The electrocardiogram demonstrated atrial fibrillation, left atrial enlargement, right axis deviation and right ventricular hypertrophy.

The chest roentgenogram revealed a massively enlarged cardiac silhouette with a cardiothoracic ratio of 0.8, enlargement of the left atrium and the right ventricle, equalization of the pulmonary vasculature and massive, curvilinear left atrial calcification forming a shell around the circumference of the left atrial chamber. The carinal angle was wide.

Transthoracic and trans esophageal two dimensional color flow and Doppler echocardiography...
was performed using a Phillips iE33 with 2.0 to 5.0 MHZ transducer and HP Sonos 5500; Hewlett Packard, Andover, MA machine. Doppler echocardiography suggested severe mitral stenosis (Mitral valve area 0.8 cm²) with gross sub valvular fusion, moderate tricuspid regurgitation and systolic pulmonary artery pressure of 100 mmHg. The left ventricular ejection fraction was 30% and the left atrium and mitral valve were severely calcified. It confirmed the presence of giant left atrium (14 cm × 13 cm, area 182 cm²) with hyper echogenic walls, mostly occupied by left atrial thrombus. Computerized tomographic scan demonstrated the presence of a thrombus localized in the atrial appendage and on the roof and the posterior wall of the left atrium; it spared the anterior and the most inferior portions of the atrial chamber. Selective coronary angiography demonstrated normal coronary arteries. Within the left atrial cavity, there was neovascularity and blush in the area of left atrial appendage with fistula formation arising from the left circumflex coronary artery (Figures 1A,1B).

After initial medical stabilization, she underwent total endoatriectomy, thrombectomy and concomitant mitral valve replacement on an emergency basis. The operation was performed under moderately hypothermic cardiopulmonary bypass using angled venous cannula (Edwards Life Sciences Research Medical Inc., West Midvale, and Utah) into the superior and inferior caval veins and aortic cannulation. St. Thomas-II cold blood cardioplegic solution (1:4) and topical hypothermia was used for myocardial preservation.

The left atrium was approached through the interatrial groove, following the axis of the caval veins. A search for a cleavage plane to achieve total removal of the calcified endocardium with the Intracavitary clot was initiated, in an attempt to perform total endoatriectomy. The endocardium was carefully excised from the underlying left atrial wall, and mitral valve annulus, taking extreme precaution not to injure the pulmonary venous openings, posterior wall of the left atrium, coronary sinus, base of the left atrial appendage, atrioventricular groove and the adjacent circumflex coronary arterial system.

The chordo-papillary apparatus was preserved using the technique described by Miki and colleagues [16]. Every attempt was made to ensure that the preserved chordopapillary apparatus allowed implantation of 29 mm SJM bi leaflet prosthesis (St. Jude Medical, St. Paul, Minnesota, USA) without causing prosthetic valve entrapment or left ventricular outflow tract obstruction. Interrupted pledged 2-0 coated braided polyester sutures (Ti-Cron, M/S Covidien, Paul, Minnesota, USA) without causing prosthetic valve entrapment or left ventricular outflow tract obstruction. Interrupted pledged 2-0 coated braided polyester sutures (Ti-Cron, M/S Covidien, Santo Domingo, Dominican Republic) were used for mitral valve replacement.

Para-annular plication parallel to the posterior mitral annulus was performed using interrupted pledged mattress sutures between the ostia of the superior and inferior pulmonary veins and the posterior mitral annulus in a semilunar fusion as described by Kawazoe and colleagues [17].

The superior plication was performed in the area between the right and left pulmonary veins. The left atrium was closed in 2 layers by double breasting the redundant left atrial wall. The base of the left atrial appendage was doubly ligated using No.2 Stupak braided silk suture (Ethicon LLC, Johnson & Johnson Pvt. Ltd., San Lorenzo, USA).

A Kay’s annuloplasty was performed to correct the tricuspid insufficiency. After releasing the aortic cross clamp, the heart resumed atrial fibrillation with a heart rate of 60/min. The patient was weaned off cardiopulmonary bypass on atrioventricular sequential pacing, dopamine 5µg/kg/min and epinephrine 0.01µ/kg/min. She was extubated on the first postoperative day.

She was anti-coagulated with low-dose subcutaneous heparin (2500 units three times a day) starting 24 hours after the operation and continued till the target Prothrombin time international normalized ratio of 2.5-3.0 was achieved with warfarin.

Four years after the operation, she is in New York Heart Association functional class-I; follow-up echocardiography showed normally functioning mitral prosthesis with a left ventricular ejection fraction of 60%, no tricuspid insufficiency and no evidence of intracavitary thrombosis.

Discussion

Since the first postmortem description of “porcelain atrium” by Oppenheimer in 1912, there have been isolated case reports of this disease entity, highlighting the problems of establishing the diagnosis and surgical techniques [18]. In 1966, Harthorne and colleagues reviewed the literature and reported 16 cases of porcelain atrium [19-23]. Published literature documents only 11 case reports of massive calcification of the entire left atrial wall including the inter-atrial septum “the coconut atrium” [4-14].

Massive calcification of the left atrium is three times more common in women [19]. It took an average of 19.7 years to diagnose massive left atrial calcification after rheumatic mitral stenosis [6,19]. Our patient was 58-year-old at the time of surgical intervention with history of rheumatic fever in childhood.

Vallejo and colleagues postulated the following pathophysiological mechanisms for the genesis of calcification of the wall of the left atrium in patients with mitral valvular disease: (i) a previous ulceration of the atrial wall as the origin of calcification, and (ii) a response to chronic strain forces in the setting of mitral valvular disease [1]. Some investigators have reported the occurrence of left atrial calcification in patients with chronic renal failure undergoing hemodialysis and following radiotherapy to chest [7,20-22]. We concur with observations of other investigators that the calcified left atrial wall prevents the left atrium from dilating, decreases it compliance and causes the elevated left atrial pressure to be transmitted to the pulmonary vessels causing pulmonary hypertension and tricuspid insufficiency [22].

Among patients with left atrial thrombus, those with massive...
intra-atrial clot form a unique subgroup. The massive atrial clot may be attached to the wall with a broad base or pedunculated or smooth and float freely within the left atrium as a “ball thrombus”. In each instance, the thrombus generally is occlusive to left atrial emptying. “Ball thrombus” as defined by Wood in 1814 (cited by Evans and Benson) is an unattached clot whose cross-sectional diameter is greater than the orifice of the chamber containing it [24]. Evans and Benson modified Woods definition to call a “mass thrombus”. Mass thrombus was defined as thrombus which by reasons of its large size or peculiar location impedes the flow of blood through the valve orifice [24].

Published literature does not provide a conclusive answer on the incidence of intra cardiac thrombus in rheumatic heart disease. Methods of patient selection i.e. necropsy, angiography, echocardiography and surgery markedly affect the prevalence. Review of 5 necropsy studies of embolic disease in patients with rheumatic heart disease revealed a 58% incidence of emboli [25-29]. The incidence of intra-cardiac thrombus in the necropsy review was 42%. Using angiography techniques, between 9 and 19% of patients with rheumatic heart disease have arterial emboli [30]. Preoperative systemic embolism occurred in 18% of patients undergoing cardiac surgery.

Five studies provided information about the location of LA thrombus. Of 156 patients with mitral stenosis and left atrial thrombus, the thrombus was found in left atrial appendage in 33% of cases, body of the left atrium in 58% of cases and both left atrium and left atrial appendage in 6% of cases [26,31-34].

Atrial wall calcification represents long-standing and extensive rheumatic mitral valve disease. Sometimes the calcification is the result of a thrombus that is adherent to the atrial endothelium [35]. Chest radiography with a penetrating view is recommended to assess long standing rheumatic mitral valve disease [1-6,35,36]. Computerized tomography of the chest is a superior imaging modality in displaying massive atrial calcification compared to chest radiography. However, even high-resolution techniques do not reveal with certainty the involvement of interatrial septum [2-6].

Magnetic resonance imaging may demonstrate the presence of thrombus, but its ability to delineate septal calcification is suboptimal [37-39]. Because of the success of M-mode echocardiography in demonstrating LA myxoma, it was hypothesized that left atrial clots might be demonstrable non-invasively [40,41]. However, M-mode echocardiography is unreliable for diagnosing relatively fixed immobile left atrial masses like thrombi [40,41].

Although two-dimensional echocardiography has a sensitivity of 75% to 78% and the specificity of 99% to 100% in detecting left atrial cavity thrombi, it is insensitive in detecting thrombi in the left atrial appendage and small thrombi with a diameter of <1 cm in the left atrial cavity [42-45]. Using a modified short axis, parasternal cross-sectional view at the aortic valve level, Herzog and colleagues reported the first 3 cases in which left atrial appendage thrombus could be visualized by two dimensional echocardiography [42]. However, other investigators could not duplicate their observations, possibly due to the following reasons: (i) the modified short-axis view is not readily obtainable and not necessarily the same in all patients, (ii) the left atrial appendage was visualized optimally only during late ventricular systole when the left atrium was maximally distended, and (iii) thirdly, this part of the cardiac anatomy has been described as “blind” and “inaccessible” to the current techniques of echocardiographic study [46,47].

Trans esophageal echocardiography can readily assess the structures in and about the left atrium because of the proximity to the probe [48,49]. However, trans esophageal echocardiography suffers from the following drawbacks: (i) in patients with an enlarged left atrium the gastro scope manipulation may induce arrhythmia due to mechanical irritation of the atrial myocardium, (ii) may cause esophageal perforation, and (iii) the imaging may be significantly impaired in the presence of densely calcified left atrial wall [5,49]. MRI may demonstrate the presence of thrombus, but its ability to depict the atrial calcification is suboptimal [37-39].

Left atrial angiography can demonstrate an intra caviary thrombus [50,51]. However, transcatheter atrial septostomy used for left atrial angiography has been associated with significant risk to the patient [50-55]. Primary arteriography levophase left atrial angiography is safer, but is not sensitive enough to detect the thrombus [50-55]. Some investigators have demonstrated that the specificity and sensitivity of left atrial angiography for thrombi are comparable to those of 2D-echocardiography study [51,54,55].

Standen using selective coronary angiography in 1975, described “tumour vascularity” with abnormal vessels arising from the left circumflex artery to the left atrium in a patient with severe mitral stenosis and left atrial thrombus [56]. Coronary neovascularization with fistula formation as a specific sign for the presence of left atrial thrombi has been documented by some investigators [57-63]. Coronary neovascularization with fistula formation has a sensitivity of 58%, specificity of 98% and positive predictive accuracy of 95% [57-63].

Giant left atrium is characterized by the following two echocardiographic findings: (a) large left atrium depicted by M-mode echocardiography with diameter >65 mm, and (b) left ventricular postero-basal wall bent inward and lying between the dilated left atrial cavity, and left ventricular cavity. The postero-basal segment usually bends more than 30 mm (in the long axis view on the two-dimensional echocardiography) after the onset of diastole [17,64,65].

Because of its potential to complicate the atriotomy, severe calcification of the left atrial wall should be anticipated before surgery. The massive calcification of the left atrium entails the following major problems from the surgical point of view: (i) a complex approach to the left atrium [2-14], (ii) difficult access to mitral valve due to rigid atrial wall (iii) embolization of the particulate calcific plaque, clots especially within the pulmonary veins and left ventricular cavity, and (iv) suture holes upon closure of the atriotomy.

During surgery on this subset of patients, we look for the best cleavage plane to remove the entire calcified “cortex” (total endoatriectomy) with the contained massive thrombus en bloc, if possible. In this way, we avoid fragmentation of the thrombus and subsequent embolization of thrombotic particles. Once the calcified “cortex” is removed, the atrial wall is still thick enough to be sutured without problems. In our experience, there have been no cases of rupture of the atrial wall, or need of pericardial or prosthetic patches to close the atriotomy.

In cases of difficult exposure, some investigators have exposed the interatrial septum through a Dubost incision [2]. Santini F and colleagues have described the technique of total replacement of the
left atrium and mitral valve utilizing a valved, T-shaped graft as a possible alternative when a more conventional approach is not feasible in cases of “coconut atrium” [66]. Published literature records the surgical mortality of this disease entity as high as 25% [11].

Conclusion

We conclude that total endoatriectomy and en bloc left atrial thrombectomy is an expedient, safe and effective technique that facilitates both the approach to the mitral valve and the suture of thrombectomy is an expedient, safe and effective technique that facilitates both the approach to the mitral valve and the suture of

References


