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A Treatise on the Surgical Management of Porcelain Atrium and Coconut Atrium

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Abstract

The present perspective is a synthesis of 119 published investigations in the setting of porcelain atrium and coconut atrium in the literature. We identified 60 suitable cases (porcelain atrium, n=44; coconut atrium, n=16) from 34 investigations, and reviewed the clinical presentation, diagnostic modalities utilized, surgical techniques employed and outcomes. Roentgenography, cross-sectional transthoracic and transesophageal echocardiography, computerized tomography and fluoroscopy provided the necessary diagnostic information and defined the disease entity before surgery in all patients. Magnetic resonance imaging and selective coronary angiography had been used for further clarification of the presence of intraluminal left atrial thrombus. We then grouped the lesions into two categories, namely "porcelain atrium" and "coconut atrium" that have a bearing on the appropriate surgical approach, discussing appropriate surgical or non-surgical techniques for each group. For the overall group, taking into consideration of the patients subjected to surgical intervention, the operative mortality remains high at 20.6% [porcelain atrium: hospital death 6/30 (20%), coconut atrium: hospital death 1/4 (25%). We submit that an increased appreciation of different types of left atrial wall calcification may well contribute to improved surgical management.

Keywords: Balloon mitral valvotomy; Closed mitral valvotomy; Coconut left atrium; Endoatriectomy; Giant left atrium; Left atrial calcification; Mitral valve replacement; Neovascularized left atrial thrombus; Open mitral commissurotomy; Porcelain left atrium

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Introduction

Massive calcification of the left atrium is a relatively uncommon complication of long-standing rheumatic mitral stenosis, and is frequently revealed in patients with a previous operation on the mitral valve [1-31]. It is reported rarely in patients with end-stage renal disease requiring hemodialysis and in patients requiring chest radiotherapy for neoplastic diseases [32-35] Left atrial calcification was first described in 1898 [3]. In the less severe forms, calcification may involve the atrial appendage, the free wall of the left atrium, or the mitral valve apparatus. The most severe form here-to-fore described- "Porcelain atrium"- has been reported infrequently; it involves the left atrial appendage, the free wall of the left atrium, and the mitral valve apparatus except for interatrial septum, thus provides surgical access to the mitral valve. However, involvement of dense calcification to the interatrial septum which is called "coconut atrium" prevents surgical correction of mitral stenosis or replacement of dysfunction prosthetic valve [31-64].

Since the first postmortem description of "porcelain atrium" on a 44-year-old male by Oppenheimer in 1912, there have been isolated case reports of this disease entity, highlighting the problems of establishing the diagnosis and surgical techniques [36]. The first x-ray of left atrial calcification observed antemortem by Bedford and was published by Shanks and associates in 1938 in a patient affected by long-standing mitral stenosis [4]. Rheumatic heart disease involving the mitral valve remains the dominant causative factors for its development. Conclusions are difficult to draw from much of the published literature on the actual incidence and outcome of calcification of the left atrium because of small number of patients in individual series and sparsity of reported clinical details [1-65]. With these deficiencies in mind, we have analyzed the published literature to identify the described instances of porcelain atrium and coconut atrium and evaluated all clinical studies describing their clinical presentation, the methodology of diagnosis, indications of surgical intervention and outcomes. The search engines employed were PubMed, Medline, Google scholar, Cochrane database and Embase. The search included literature in all languages. This strategy yielded 119 investigations addressing specifically the etiopathogenesis, diagnostic criterions, perioperative mortality and management of these difficult surgical entities. We have then synthesized all these features to outline the issues of concern and trends of various surgical strategies of the porcelain and coconut atrium.

Patients and Methods

As far as we could establish there have been 115 cases of "porcelain atrium" and 16 cases of "coconut atrium" described in the published literature [1-64]. For the purposes of this review, we have selected 44 patients of porcelain atrium and 16 patients of coconut atrium from 34 published series in whom an accurate description was provided that might possibly have a bearing on the clinical presentation, diagnosis, surgical techniques and perioperative mortality (Tables 1A, 1B and 2) [31-64]. In making this selection, we excluded 71 patients from the world series in whom we deemed the anatomical description and surgical techniques utilized to be inadequate for our purpose [1-30].

Table 1A: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
1.	Lee JW et al[38]	2010	67 years female	RHD, mitral stenosis, chronic atrial fibrillation; MVR 23 years back, CTR 82%, transthoracic echocardiography- good prosthetic valve function; mild MR, severe TR, PAP 70 mmHg, CT-massive LA calcification	Endoatriectomy 2008- Tricuspid annuloplasty 33 mm ring	Survived Follow-up at 2 years NYHA class II
2.	Ha JW et al[39]	2001	67 years female	RHD, severe mitral stenosis, chronic atrial fibrillation, chest x-ray, cardiomegaly, large left atrium, echocardiography- heavy calcification, left atrium thrombi, CT scan- same findings	Refused surgery	-
3.	Vallejo JL et al[40]	1995	N=8 Age: mean 55.4± 19.6 years (range: 35-69 years)	RHD, severe mitral stenosis, chronic atrial fibrillation, porcelain atrium=7, coconut atrium=1; closed mitral valvotomy (n=2), open mitral commissurotomy (n=3), aortic valve replacement and open mitral commissurotomy (n=1), mitral valve replacement (n=2), interval between surgical procedures 15.4±7.7 years (8-29 years), pulmonary artery pressure 67±23.1 mmHg (35-100 mmHg)	Transseptal superior approach posterior to interatrial groove extending superiorly; endoatriectomy + MVR=4 MVR+tricuspid annuloplasty=2 Aortic and AVR+MVR=2	Hospital death=1 (12.5%) Late death=2 (25%)
4.	Ruvolo G et al[41]	1994	56 years male	RHD, severe mitral stenosis, chronic atrial fibrillation; chest x-ray- extensive mild like calcification; echocardiography, catheterization- moderate tricuspid regurgitation; moderate pulmonary hypertension; LA size 60 mm	Wide endoatriectomy, MVR 29 mm carbomedics	Survived; doing well at 18 months follow-up; NYHA I

Table 1A: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
5.	Vilacosta I, et al[42]	1994	71 years female	RHD, severe mitral stenosis, chronic atrial fibrillation, OMC at 54 years age; left hemiplegia at 54 years; transesophageal echocardiography- massive calcification, prevented visualization of left atrium	Not mentioned	-
6.	Leacock K et al[43]	2011	71 years female	RHD, severe mitral stenosis, chronic atrial fibrillation, chest x-ray- curvilinear cardiac calcification; CT- left atrial distribution of calcification; porcelain atrium	Surgery denied Discharged home	Surviving at discharge
7.	Prestana G et al[34]	2018	59 years male	Chronic renal failure, on hemodialysis Transthoracic echo- long-mobile mass contiguous with LA appendage; Transesophageal echocardiography- 2 mobile masses in left atrium 21mm disappeared on repeat echo; CT- multiple LA parietal calcification surrounding the pulmonary venous ostia, extending to left atrial appendage ostium; foci of ectopic calcification in kidney disease	Discharged on oral anticoagulation	Surviving at the time of reporting
8.	Edwards JM, Chrisholm RJ[44]	2006	69 years female	Postoperative MVR, permanent pacemaker, chest x-ray cardiomegaly, porcelain atrium, fluoroscopy confirmed the extent of calcification	Not mentioned	Surviving at the time of reporting
9.	Lahey T, Horton S[32]	2002	22 years female	Chronic idiopathic glomerulonephritis at 10 years of age, subtotal parathyroidectomy at 14 years; transthoracic echocardiography- massive mitral annular calcification extending to left atrium, mobile echogenic material, recurrent myocardial and cerebrovascular ischemia at 22 years of age	Declined surgery (endoatriectomy)	Surviving at the time of reporting

Table 1A: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
10.	Peces R et al[33]	2000	28 years female	Chronic renal failure on hemodialysis for 20 years; severe secondary hyperparathyroidism, x-ray, CT and TEE-massive LA calcification corresponding to multiperforated membrane of cortriatriatum; multiple vascular calcification; superior mesenteric artery occlusion	Small bowel resection	Surviving at the time of reporting; 20 years follow-up-several episodes of septicemia because of central venous catheters
11.	Popescu BA et al[45]	2010	64 years female	Starr Edwards MVR 1975, chest x-ray, cardiothoracic ratio 0.90, widening-tracheal bifurcation, porcelain atrium, transthoracic echocardiography, left atrium 12/13cm, area, 127 cm^2, hyperechogenic walls, intraluminal thrombus, CT-confirmed findings	Treated conservatively; refused surgical treatments	Discharged home
12.	So YH et al[46]	2002	6 patients (29-74 years); median 49 years	Previous MVR with failure; systolic PA pressure 37-97 (mean 62 mmHg); diameter LPA 22.5-36.3 mm (mean 22.8 mm); interval between surgery and CT-16 years	Endoatriectomy	All survived
13.	Goel AK et al[47]	1997	17 years female	RHD; severe mitral stenosis; normal sinus rhythm; chest x-ray; circumferential, heavy, left atrial, wall calcification, transesophageal echocardiography- severe mitral stenosis; calcified left atrium; no thrombus	Not mentioned	

Table 1A: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis					Treatment	Results
14.	Koroglu M et al[48]	2005	40 years male	End-stage renal disease; on hemodialysis; aortic calcification; severe aortic stenosis; severe mitral annular calcification; CT-calcified left atrium; dura-spinal cord					Oral anticoagulation; medical treatment;	Discharged home
15.	Sankhyan LK et al[50]	2018	58 years female	RHD, severe calcific mitral stenosis, chronic atrial fibrillation, large left atrial clot, chest x-ray, porcelain atrium, giant left atrium, coronary angio, vascularized left atrial clot					Mitral valve replacement (29 mm-St. Jude Medical, Mechanical), Endoatriectomy	Survived, 4 years postoperative NYHA-I
16.	Roberts WC et al[49]	1970		Diagnosis	Duration of symptoms (years)	PA pressure (mm Hg)	LA-LV gradient (mm Hg)	Previous operation (6-9 years prior)		
			57 years female	MS, TR, AF, RVH	24	52/15	8	OMC	Re OMC, TVR, thrombectomy	Died
			32 years female	MS, TR, AF, RVH	11	59/41	5	OMC	Nil	Died before operation
			39 years female	MS, TR, AF, RVH	39	82/32	15	OMC	MVR, tricuspid annuloplasty; thrombectomy	Died

Table 1B: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management (contd…).

S.No.	Authors	Year	Age/Sex	Diagnosis	Duration (years) prior to recognition of calcification	Means by which calcification was diagnosed	Valve calcification	Operative or postmortem findings	Preoperative embolic events	Results
17.	Harthrone JW et al[31]	1966	52/Female	MS, atrial fibrillation- 16 years	21 years	X-ray, postmortem findings	No	Porcelain atrium, valve fibrous, large friable LA clot, operation 1-CMV and postmortem	Embolic phenomenon, CVA at 47 years, saddle embolus during surgery	Died
			48/Female	MS, AR, paraoxysmal atrial fibrillation	5 years	X-ray and surgery	No	Closed mitral valvotomy, thrombosed left atrial appendage, porcelain atrium, fibrous valve	Pulmonary emboli	Living, improved after surgery
			56/Female	MS, AR, chronic atrial fibrillation-13 years	20 years	X-ray only	No	Operation 1-CMV, no calcium, no clot Operation 2-Re-CMV, clot in the area of old atriotomy, no calcium	CVA at 43 years ? renal; BP 200/104	Living improved after surgery
			44/Female	MS, TR, chronic atrial fibrillation-6 years	7 years	X-ray and surgery	No	Operation 1-Atrial wall diffusely calcified, sparing interatrial septum, no clot, difficulty in entering atrium; CMV→ moderate mitral regurgitation PA pressure (systolic/diastolic) - 100/40 (mean 50); pulmonary vascular resistance 13 (Woods unit/m^2)	Saddle embolus at 40 years	Living, improved after surgery

Table 1B: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management (contd…).

S.No.	Authors	Year	Age/Sex	Diagnosis	Duration (years) prior to recognition of calcification	Means by which calcification was diagnosed	Valve calcification	Operative or postmortem findings	Preoperative embolic events	Results
			54/Female	MS, AR, chronic atrial fibrillation-20 years	22 years	x-ray, surgery and postmortem findings	Yes	Operation 1- Thrombosed left atrial appendage, calcified wall, organized LA clot, fibrous valve Operation 2- Diffusely calcified wall PA pressure (systolic/diastolic) - 120/50 (mean 50)	Cerebral, splenic, pulmonary emboli at 40 years	Died after surgery
			50/Male	MS, AR, chronic atrial fibrillation-11 years	9 years	Surgery and x-ray in retrospect	No	Porcelain atrium, no LA clot, CMV- adequate valve opening, fibrous mitral valve PA pressure (systolic/diastolic) - 75/25 (mean 40)	Pulmonary emboli	Living, improved after surgery
			56/Female	MR, MS, AR, AS, TR, chronic atrial fibrillation-20 years	20 years	x-ray, surgery and postmortem findings	No	Operation and postmortem- calcified left atrium with clot	Pulmonary bifurcation embolus	Died after surgery
			50/Female	MS, AS, TR, PS, chronic atrial fibrillation-12 years	12 years	x-ray and postmortem findings	No	Operation and postmortem- calcified left atrium with clot	Splenic, pulmonary and renal emboli, CHF	Dead
			56/Female	MS, , chronic atrial fibrillation-20 years	25 years	X-ray and surgery	No	Operation 1- “Great deal” of adherent clot, calcified wall, but not LAAA, valve fibrous	Cerebral and renal emboli? BP 170/100mmHg	Living improved

Table 1B: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management (contd…).

S.No.	Authors	Year	Age/Sex	Diagnosis	Duration (years) prior to recognition of calcification	Means by which calcification was diagnosed	Valve calcification	Operative or postmortem findings	Preoperative embolic events	Results
			62/Female	MS, TR, , chronic atrial fibrillation-14 years	No	S-ray only	No	None	Pulmonary emboli	Living, chronic CHF
			49/Female	MS, , chronic atrial fibrillation-30 years	No	X-ray only	No	None	CVA at 48 years, suspected pulmonary embolism	Living, refuses surgery
			57/Female	MR, MS, TR , chronic atrial fibrillation-27 years	27 years	x-ray and postmortem findings	No	Postmortem- Giant left atrium, calcified left atrial appendage and posterior left atrial wall	None	Died
			56/Female	MS, AS, normal sinus rhythm	11 years	x-ray, surgery and postmortem findings	Yes	Operation 1- Age 49 - CMV Operation 2- Age 51 - OMC, open atrial commissurotomy Operation 3- AVR, MVR, calcified LA, no LA clot,	None	Dead of table
			50/Male	MS, AR, , chronic atrial fibrillation-19 years	19 years	X-ray and surgery	No	Operation 1- Age 38 - CMV, no calcium, no clot Operation 2 - Age 40 - OMC, calcified LA, mural thrombus, interatrial septum spared	Pulmonary emboli at 33, 34; left femoral embolus at 45 year, right femoral embolus at 50 years	Surviving, doing well

Table 1B: Summary of the published investigations documenting the diagnosis of porcelain atrium and its management (contd…).

S.No.	Authors	Year	Age/Sex	Diagnosis	Duration (years) prior to recognition of calcification	Means by which calcification was diagnosed	Valve calcification	Operative or postmortem findings	Preoperative embolic events	Results
			61/Female	MS, TR, , chronic atrial fibrillation-17 years	2 years	X-ray only	No	Age 49 years, closed mitral valvotomy, no calcium, no clot	Left femoral embolus-49 years	Surviving, doing well
			64/Female	MS, chronic atrial fibrillation-22 years	22 years	x-ray and postmortem	Yes	Postmortem-porcelain atrium, severe calcific MS, massive LA thrombus	Cerebral, renal , splenic emboli	Died

AF= Atrial fibrillation, AR= Aortic regurgitation, AS= Aortic stenosis, AVR= Aortic valve replacement, Cath= Catheterization, CMV= Closed mitral valvotomy; CT-scan= Computerized tomographic scan, CTR= Cardiothoracic ratio; CVA= Cerebrovascular accident, IABC= Intra-aortic balloon counterpulsation LA= Left atrium, LAAA= Left atrial LPA= Left pulmonary artery, LAAA= Left atrial appendage, LV= Left ventricle, LVEDP= Left ventricular end-diastolic pressure, LVOT= Left ventricular outflow tract, MR= Mitral regurgitation, MS= Mitral stenosis, MVR= Mitral valve replacement, NYHA= New York Heart Association, OMC= Open mitral commissurotomy, PAP= Pulmonary artery pressure, PCWP= Pulmonary capillary wedge pressure, RHD= Rheumatic heart disease, RVH= Right ventricular hypertrophy, TEE= Transesophageal echocardiography.

Table 2: Summary of the published investigations documenting the diagnosis of coconut atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
1.	Radhakrishnan BK et al[51]	2018	N=1 (57 years female)	Rheumatic heart disease with mitral stenosis; chronic atrial fibrillation; closed mitral valvotomy 29 years back; balloon mitral valvotomy 8 years ago; chest x-ray-coconut atrium	Endocardectomy; mitral valve replacement	Not mentioned

Table 2: Summary of the published investigations documenting the diagnosis of coconut atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
2.	Tsumaru S et al[52]	2015	N=1 (58 years female)	Three prior open heart surgeries; redo mitral valve replacement; CT scan; Coconut atrium	3rd sternotomy for prosthetic mitral valve stenosis; transseptal superior approach; endoatriectomy and bileaflet mitral valve replacement; left atrial wall reconstruction using bovine pericardium	Survived uneventful postoperative recovery
3.	Reddy D et al[53]	2013	N=1 (75 years male)	RHD with mitral stenosis; chronic atrial fibrillation for 30 years; previous closed mitral valvotomy; open mitral valvotomy; balloon mitral valvotomy; unenhanced/computed tomography- coconut atrium	Surgery was not performed because of dense calcification	Surviving at the time of reporting; on medical treatment
4.	Panayiotou A and Holloway B[54]	2018	27 years male	End-stage renal failure, failed renal transplant, long-term hemodialysis, severe hyperparathyroidism, parathyroidectomy, serial CT image- coconut atrium	Medical follow-up	Surviving at the time of reporting; on medical treatment
5.	Anandan PK et al[55]	2015	37 years female	Rheumatic heart disease; mitral stenosis; mitral regurgitation; chronic atrial fibrillation; balloon mitral valvotomy 13 years back; computed tomography-coconut atrium	Contraindication for mitral valve surgery; medical follow-up	Surviving at the time of reporting; on medical treatment

Table 2: Summary of the published investigations documenting the diagnosis of coconut atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
6.	Del Campo C et al[56]	2000	67 years female	Rheumatic heart disease; mitral stenosis; chronic atrial fibrillation; open mitral commisurotomy (right thoracotomy)-26 years back; moderate pulmonary arterial hypertension. Transthoracic, transesophageal echocardiography-coconut atrium	Advised urgent mitral valve replacement; refused surgery. Emergency surgery-inaccessible surgical approach through Dubost incision; MVR abandoned; Devega's tricuspid annuloplasty for tricuspid regurgitation, IABC	Died 48 hours later
7.	Meyners W, Peters S[57]	2003	65 years female	Rheumatic heart disease; mitral stenosis; at 42 years of age, MVR (Starr Edwards mechanical prosthetic valve), left atrial thrombectomy for massive left atrial thrombus; chronic atrial fibrillation; echo-severe pulmonary artery hypertension, tricuspid regurgitation; well functioning prosthetic valve; cine-mural left atrial calcification; LA 6.5 x 8.3cm	Conservative medical treatment	Surviving at the time of reporting

Table 2: Summary of the published investigations documenting the diagnosis of coconut atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
8.	Pullikal G, Marshall A[58]	2006	71 years male	Rheumatic heart disease; mitral stenosis; chronic atrial fibrillation; closed mitral valvotomy- 37 years back; MVR (Starr Edwards mechanical prosthesis) 21 years back; massive calcification- inter-atrial septum; transthoracic echocardiography- coconut atrium; LVOT gradient 30mmHg (cath); oral anticoagulation	Medical treatment Surgery contraindicated because of involvement of interatrial septum	Surviving at the time of reporting
9.	Onishi T et al[59]	2015	76 years male	Open mitral commissurotomy followed by aortic and mitral valve replacement; chest x-ray; diffuse calcification, coconut atrium; CT- circumferential calcification	Medical treatment Autopsy-dilated coconut atrium Histopathology- intensive calcification of endocardium	Died
10.	Funada A et al[60]	2012	78 years male	Rheumatic heart disease; mitral stenosis; chronic atrial fibrillation; aortic and mitral valve replacement; left atrial thrombectomy- 21 years ago; CT- coconut atrium; cardiac cath- severe pulmonary arterial hypertension	Surgery was not recommended because of absence of cleavage plane and low success probability	Surviving at the time of reporting

Table 2: Summary of the published investigations documenting the diagnosis of coconut atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
11.	Santini F et al[61]	1998	61 years male	Rheumatic heart disease; mitral stenosis; aortic and mitral valve replacement (Starr Edwards mechanical prosthesis)-28 years ago ; paravalvular leak, dysfunctional prosthetic valve	Total prosthetic replacement of left atrium and mitral valve enbloc leaving 2 pulmonary vein cuff free of calcification; repair by means of a valved T graft (29 Carbomedics)	Survived the operation at 1 year NYHA II; CT scan functioning successful anastomotic reconstruction-T-shaped conduit
12.	Lin YS et al[62]	2008	47 years male	Rheumatic heart disease; severe mitral stenosis; mitral regurgitation; moderate pulmonary hypertension; age 22 years; MVR-Carpentier Edwards 29 mm bioprosthesis; age 29 years- degenerated bioprosthesis; redo MVR (31 mm Sorine Mechanical Bioprosthesis, USA) for degenerated bioprosthesiss; chest x-ray coconut atrium	Redo MVR (St. Jude 31 Mechanical Valve) for paravalvular leak and ventricular rupture	Survived 1 year postoperative surgery NYHA II
13.	Jorgensen M et al[63]	2014	49 years female	CT scan- coconut calcification; moderate coronary calcification		
14.	Kawakami R et al[64]	2015	84 years female	AVR, MVR- 35 years ago Chest x-ray; severe cardiomegaly; left atrial enlargement with curvilinear calcification; CT chest- dense thick plate like calcification, coconut atrium	Difficulty in finding cleavage plane; erosive calcification; renal infarction due to emboli calcified left atrium	Died 14 days after admission.

Table 2: Summary of the published investigations documenting the diagnosis of coconut atrium and its management.

S.No.	Authors	Year	No. of patients	Diagnosis	Treatment	Results
15.	Jenkins NP et al[35]	2004	45 years male	Previous surgery + radiotherapy- thoracic neuroblastoma; CT- coconut atrium; cath and fluoroscopy confirmed the findings; normal coronaries; mean PA pressure 34 mmHg; mean PCWP 20 mmHg; transpulmonary gradient 14mmHg; LVEDP 12 mmHg; non-compliant left atrium	Not mentioned	Unknown

AF= Atrial fibrillation, AR= Aortic regurgitation, AS= Aortic stenosis, AVR= Aortic valve replacement, Cath= Catheterization, CMV= Closed mitral valvotomy; CT-scan= Computerized tomographic scan, CTR= Cardiothoracic ratio; CVA= Cerebrovascular accident, IABC= Intra-aortic balloon counterpulsation LA= Left atrium, LAAA= Left atrial LPA= Left pulmonary artery, LAAA= Left atrial appendage, LV= Left ventricle, LVEDP= Left ventricular end-diastolic pressure, LVOT= Left ventricular outflow tract, MR= Mitral regurgitation, MS= Mitral stenosis, MVR= Mitral valve replacement, NYHA= New York Heart Association, OMC= Open mitral commissurotomy, PAP= Pulmonary artery pressure, PCWP= Pulmonary capillary wedge pressure, RHD= Rheumatic heart disease, RVH= Right ventricular hypertrophy, TEE= Transesophageal echocardiography.

Incidence

Due to rarity of these disease entities, the actual incidence of "porcelain atrium" and "coconut atrium" is unknown. Mention of these findings in contemporary cardiology text books is uncommon. Literature search till date revealed 44 cases of porcelain atrium and 16 case reports of "coconut atrium" [1-67]. Massive calcification of the left atrium is three times more common in women (74%) [1-67]. It took an average of 19.7 years to diagnose massive left atrial calcification after rheumatic mitral stenosis [1-67]. The incidence of systemic or pulmonary embolism is difficult to estimate but they were present in 15 of the 60 cases in which a specific note was made [31-64].

Demographics

The age of the patient at initial diagnosis ranged from 17 to 84 years (mean 55.5±14.3 years, median 56 years) and massive calcification of left atrium in 3 times more common in females (74%) [31-64]. It was not possible to identify any regional or ethnic predominance. From a review of cases, one can see that the clinical characteristics of majority of these patients are rheumatic heart disease of long duration. Other rarer causes include patients with chronic renal failure on long-term hemodialysis and those undergoing thoracic radiotherapy for neoplastic diseases [31-64]. Atrial fibrillation of many years duration is almost universal, averaging more than 10 years in several series. Mitral valve involvement, usually of a severe degree is

present in all patients, although the incidence of valvular calcification seems unrelated. Combined or multivalvular lesions are common. The predominance of female (74%) is striking and follows the higher involvement of the mitral valve. Published literature documents symptoms referable to the cardiovascular system for an average of 19.7 years prior to recognition of left atrial calcification. Considerable left atrial enlargement and dysphonic symptoms are frequently present. The diagnostic accuracy is improving with the use of deep voltage roentgenography, transthoracic and transesophageal echocardiography, computerized-tomographic scan and fluoroscopy. The diagnosis was confirmed by postmortem examination or surgical intervention in 42 of the 86 cases reported in the world literature till 1966 [1-31].

Pathophysiology

Calcification of the left atrium is an uncommon complication of long-standing valvular rheumatic heart disease, end-stage renal disease requiring long-term hemodialysis and radiation injury. Calcification of the atrial endocardium is even more uncommon [1-67]. The following pathophysiological mechanisms have been proposed for the genesis of calcification of the wall of the left atrium in patients with mitral valvular disease: i) a response to chronic strain forces in the setting of mitral valvular disease [68]. Such long-term hemodynamic forces in the left atrium stretches the walls of the left atrium and induce endocardial dysfunction. During atherogenesis, endothelial dysfunction plays an important role in vascular calcification. Several investigators have demonstrated that the endothelium is a source of osteoprogenitor cells in vascular calcification [31-50]. Further inflammation during the healing process of rheumatic carditis may be involved in the pathogenesis of rheumatic heart disease. Histologically, there are fibrotic changes and few inflammatory cells in the left atrial wall, suggesting the occurrence of active inflammation during very early phase of the rheumatic fever. These factors induce structural and electrical remodeling of the atrium, which lead to the development of atrial fibrillation. Indeed, atrial fibrillation of many years duration is almost universal. The advanced electro anatomical remodeling of the left atrium exacerbates the hemodynamics of heart failure; ii) the consensus of other group of investigators is that left atrial calcification is the end-result of repeated and extensive rheumatic auricularis beginning with focal patches of calcification in areas of rheumatic endocardial ulceration and progression to chronic fibrotic changes with plaques of subendocardial calcium [31-50]. The incidence of atrial calcification appears to be related to the severity of the original rheumatic attack and the associated valvular damage. Adherent mural thrombi are a common finding [31-50]. Accurate description of the atrial calcification is missing in most published reports; iii) recent research on the mechanisms of ectopic calcification such as arterial calcification indicates that well-regulated processes, similar to those that drive osteogenesis, mediate vascular calcification [69-77]. It is hypothesized that a subpopulation of vascular cells retains the potential to differentiate into osteoblasts and may play a pivotal role in vascular calcification by forming mineralized nodules and expression of osteoblast-specific proteins such as osteocalcin, bone morphogenetic protein-2 and alkaline phosphatase [76,77] However, currently there are no published studies that define the mechanisms of calcification in the myocardium. Other mechanisms seem to play a role as well. Secondary hyperparathyroidism in patients with chronic renal failure, endothelial progenitor cells, or calcifying vascular cells have been shown to secrete substances that foster calcium deposition. Transforming growth factor-" may trigger this process. Some authors suggested a link between aluminum intoxication and extra skeletal calcium deposition, whereas others raised the possibility that hypomagnesemia might foster the development of mitral annular calcification [68-

77]; iv) radiation injury has also been incriminated as one of the etiological factor in selected cases; v) reports of a small, calcified left atrium and a giant right atrium in a subset of patients led Roberts and associates to conclude the following: calcific deposits prevent the left atrium from dilating and overstretching, decreased its compliance and caused the elevated left atrial pressure to be transmitted to the pulmonary vessels and right ventricle, leading to tricuspid insufficiency [49] and vi) calcification of the left atrial wall is frequently associated with adherent mural thrombi, which may also calcify. 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 [50]. Associated valvular calcification bears no predictable relation to atrial calcification [31-64,78].

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. 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 [79].

Published literature does not provide a conclusive answer on the incidence of intracardiac 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 [80-84]. 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 [85]. 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 [81,86-88].

Diagnosis

Diagnosis of this entity is a rewarding clinical experience. Roentgenographic recognition of this entity has received much comment in radiological literature. Radiography of the left side of the chest is recommended to assess long-standing mitral valve disease. Careful roentgenographic techniques are necessary to identify left atrial calcification. High kilovoltage and well-penetrated films are essential. The lateral or oblique projections are most useful. Laminography may be helpful for better definition of the extent of the calcification. Cineradiotherapy because of movement of the deposits, often reveals calcification not apparent on routine films and is a valuable diagnostic tool to evaluate mechanical heart valve prostheses and to identify the presence and extent of cardiac calcification [89-92]. Mural calcification appears as a thin, curvilinear density tracing the outline of the left atrium in part or completely. In the frontal projection, a round or oval shell of calcium 8 to 10 cm in diameter is usually seen lying in the center of the cardiac silhouette beneath the carina and the main stem bronchi. In the lateral or oblique projections, a completely calcified wall appears as a C-shaped curvilinear density with the opening of the C lying anteriorly in the region of the mitral annulus. Lesser degrees of calcification may be confined to streaks along one or more margins

or may outline the entire posterior wall. Occasionally the calcification extends into the pulmonary veins. If the calcification closely approaches or forms the left heart border in the frontal projection, the atrial appendage is probably calcified, and this may occur as an isolated finding. It must be remembered, however, that the calcification is frequently more extensive than revealed by x-ray [31, 89-92].

Left atrial calcification must be differentiated from calcification occurring in the cardiac valves, pericardium, coronary arteries, ventricular aneurysms or infarctions, intracardiac tumors, mediastinal cysts, hilar nodes, and costal cartilages. All of these have characteristic locations and motion, and present little difficulty to the experienced observer. The image of computed tomography can display massive atrial calcification rather than the plain chest x-ray. However, even with high-resolution techniques, computed tomography does not reveal with certainty the degree of calcification of the interatrial septum [56, 93, 94]. The feasibility of transthoracic and transesophageal echocardiography for the diagnosis of porcelain atrium / coconut atrium has not been adequately addressed. In general, transesophageal echocardiography permits adequate visualization of the left atrium compared with transthoracic echocardiography because of anatomic accessibility of the esophagus and the proximity of the left atrium close to esophagus. Vila Costa and associates reported a case wherein they found that quality of transesophageal imaging is significantly impaired in the presence of calcified walls of the left atrium [42]. Goel and associates were able to obtain satisfactory image quality with multiplane transesophageal echocardiography in spite of heavy circumferential calcified left atrial walls. Adequacy of transesophageal echocardiographic imaging possibly depends on the extent and density of calcification. There may have been some less dense areas through which a satisfactory window could probably be obtained [47]. However, transesophageal echocardiography suffers from the following drawbacks: (i) in patients with an enlarged left atrium the gastroscope 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 [42,56].

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 < 1cm in the left atrial cavity [95-98]. 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 [95]. 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” [99,100]. Left atrial angiography can demonstrate an intracavitary thrombus [101, 102]. However, transseptal atrial septostomy used for left atrial angiography has been associated with significant risk to the patient [101-105]. Primary arteriography levophase left atrial angiography is safer, but is not sensitive enough to detect the thrombus [101-105]. Some investigators have demonstrated that the specificity and sensitivity of left atrial angiography for thrombi are comparable to those of 2D-echocardiography study [102,104,106]. Standen using selective coronary angiography in 1975, described “tumor 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 [107]. Coronary neovascularization with fistula formation as a specific sign for the presence of left atrial thrombi has been documented by some investigators [108-114]. Coronary neovascularization with fistula formation has a sensitivity of 58%, specificity of 98% and positive predictive accuracy of 95% [108-115]. Magnetic resonance imaging may demonstrate the presence of thrombus, but its ability to depict the atrial calcification is suboptimal [116-118].

Surgical approach and management

Surgery is the mainstay of management of porcelain atrium and coconut atrium and has evolved with time. Massive calcification of the left atrium entails surgical problems. Since calcification of the left atrial wall will usually indicate thickening, fibrosis and difficulty in entering the left atrium, it is important for the surgeon to be aware of its presence prior to surgery [31-51,119]. Surgery must be tailored to the individual patient and in the patient with isolated, noncalcific mitral stenosis with complicating pulmonary hypertension is probably best carried out as a closed procedure (closed mitral valvotomy, balloon mitral valvotomy) recognizing the attendant hazards. For the patient without pulmonary hypertension or with mixed or combined lesions, an approach through a right thoracotomy or median sternotomy and open valvuloplasty through the posterior interatrial groove or atrial septum seems preferable [31-64]. The massive calcification of the left atrium entails the following major surgical problems: (i) a complex approach to the left atrium, (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) hemorrhage from suture holes upon closure of the friable left atrial wall [31-64]. It is a common practice to approach the mitral valve through left atrium posterior to interatrial groove, through the right atrium and in the interatrial septum. In cases of “porcelain atrium”, the interatrial septum is usually free from calcification. During surgery on this subset of patients with “porcelain atrium”, 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. There have been no cases of rupture of the atrial wall, or need of pericardial or prosthetic patches to close the atriotomy among patients subjected to surgical intervention. The great majority of porcelain atrium have been managed through the superior approach of Tucker. Surgical access to the mitral valve in cases of complete calcification of the left atrium, including the interatrial septum becomes very difficult. Out of 16 reported cases in the published literature only 4 patients could be operated on [51-64].

Based on the available preoperative imaging, several investigators envisaged an impenetrable left atrium, with the risk of perioperative morbidity and mortality exceeding the perceived benefit of surgical intervention. In a case of coconut atrium, Ruvolo and colleagues were able to make one small incision behind the interatrial groove and then a transseptal incision, which made possible a total endoatriectomy and mitral valve replacement.[41] A complete left atrial endoatriectomy was undertaken through the two little breaches opened in the calcified cell after fragmenting the latter at many points with a costotome. Using the traditional approaches to the mitral valve (transseptal, interatrial groove, atrial roof) simultaneously, it was possible to remove the calcified shell along its cleavage plane. In cases of difficult exposure for cases of coconut atrium, some investigators have exposed the interatrial septum through a Dubost incision. Santini F and colleagues have described a 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" [61].

Surgical Results

The overall mortality for patients undergoing surgery for porcelain and coconut atrium is 20.6% (porcelain atrium 6/30, 20%; coconut atrium, 1/4, 25%). There were 5 more non-surgical deaths who died of diverse causes including multiple cerebral, splenic, renal, pulmonary emboli and congestive cardiac failure. Many patients refused surgery. The high mortality rate in the published series may be explained taking into account the period of time within which the patients were operated, the antecedent of at least one previous surgical procedure and the presence of severe pulmonary artery hypertension. The high mortality is related to patients advanced diseases process rather than the operative technique. Although total endoatriectomy of a "coconut left atrium" remains the procedure of choice, the possibility of performing total prosthetic replacement of the left atrium and mitral valve enbloc in complicated cases as described above should be kept in mind in the surgeon's armamentarium as an acceptable solution for a potentially dreaded condition. Cardiovascular surgeons should carefully consider the contemporary benefits of various surgical options when managing porcelain and coconut atrium.

Conclusions

On the basis of the published literature enunciated in the manuscript including ours, we conclude that total endoatriectomy and en bloc left atrial thrombectomy is an expedient, safe and effective technique in patients with "porcelain atrium" that facilitates both the approach to the mitral valve and the suture of the atrial wall. The surgeon should take extreme precaution during endoatriectomy not to injure the posterior left atrial wall, ostia of the pulmonary veins, coronary sinus and atrioventricular groove. Although, it may not be possible to establish preoperatively that the atrium is completely calcified, the surgeon should suspect this disease entity (porcelain atrium) in the presence of predisposing factors i.e. a woman with a long history of mitral stenosis or have undergone previous closed mitral valvotomy / open mitral valvotomy / percutaneous mitral valvotomy / mitral valve replacement, has high pulmonary artery pressure, suffer congestive cardiac failure and evidence of massive calcification of the left atrial wall on computerized tomography. If there is evidence of complete transmural calcification (i.e. the septum cannot be visualized by transesophageal echocardiography and appears calcific on computerized tomography), the surgeon should suspect "coconut atrium" and should weight carefully the decision to operate. Knowledge of this approach should contribute to the armamentarium of the cardiac surgeon faced with such findings as enunciated above.

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