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Original Article

An analysis of 24 autopsied cases with supramitral rings

Pradeep Vaideeswar, Milind M. Baldi, Sandeep Warghade

Department of Pathology (Cardiovascular & Thoracic Division), Seth G. S. Medical College, Mumbai, India

Abstract The supramitral ring is a rare congenital malformation formed by presence of a ridge of connective tissue, usually attached at or above the mitral annulus. The incidence and clinical presentation is highly variable due to difficulty in diagnosis. A review of autopsied congenital heart diseases at our institute over a 17-year-period revealed 24 cases of supramitral ring. These were classified with respect to the morphology of the ridge and the presence of associated cardiac lesions. The ring was found in 1.5% of the autopsied specimens of congenitally malformed hearts, and in 37.5% of those with obstructed left-sided inflow tracts. The majority of the specimens came from children (79.2%). A clinical diagnosis had been made in only two. In one-third of the cases, the ring was associated with incomplete Shone's complex. Varied anomalies were seen in others, chiefly ventricular septal defects. An interesting association was the presence of rheumatic mitral valvar disease, found in 3 cases. There was no difference in the completeness or width of the supramitral ridge in the hearts from those with or without Shone's complex. Circumferential rings were fleshy and stenosing, while incomplete rings had variable locations and stenosis. The presence of a supramitral ring may be underestimated due to association with other cardiac anomalies, both congenital and acquired. Since the ridge need not always produce stenosis, the correct designation would be simply a supramitral ring.

Keywords: Congenital heart disease; congenital mitral stenosis; supravalvar mitral stenosis

In developing countries, rheumatic mitral stenosis continues to be the commonest cause of obstruction to the left ventricular inflow tract. Besides, by virtue of accelerated fibrosis, the disease is capable of producing crippling deformity in the very young.[1] But, there exist rare congenital lesions that can produce a spectacular variety of stenosing lesions. One among them is the so-called supramitral ring, characterized by a ridge of connective tissue that encroaches into the inflow tract at or above the mitral valvar annulus.[2] The clinical presentation of this rare anomaly is variable owing to the associated cardiac defects, as well as the morphology of the ridge, often leading to inadequate collection of data. We sought to shed light on the morphological aspects by presenting our experience of 24 such supramitral ridges encountered at autopsy.

Correspondence to: Dr Pradeep Vaideeswar, Department of Pathology (Cardiovascular & Thoracic Division), Seth G. S. Medical College, Parel, Mumbai 400 012, India. Tel: 91-22-24136951, ext 2550; Residence – 91-22-32501327; Fax: 91-22-22-24143435; E-mail: shreeprajai@yahoo.co.in

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Materials and methods

Over a 17-year period, from 1991 through 2007, we came across 64 cases of various congenital obstructions of the left ventricular inflow tract, excluding cases of mitral atresia (Table 1). These cases accounted for 3.9% of the 1633 autopsied examples of congenital cardiac disease, and amongst them were 24 examples of supramitral ring. The age and gender, mode of clinical presentation, findings on cross-sectional echocardiography, interventions if any, and causes of death were noted. All the hearts were carefully examined using sequential segmental analysis. Particular attention was paid to the morphology of the supramitral ridge, taking note of its completeness, location, and nature, as well as

the mitral valve. The hearts with the supramitral ridges were then classified into two groups, specifically into those with or without associated Shone's complex, which is the combination of a supramitral ridge, parachute mitral valve, sub-aortic stenosis, and aortic coarctation.[3] The complex was further categorized into complete and incomplete forms, depending on the presence of all four, or two or more of the lesions, respectively.[2]

Results

Our 24 cases of supramitral ring accounted for 1.5% of the autopsied cases of congenital cardiac disease, and 37.5% of those with obstruction to the left-sided inflow tract. Most of them came from infants and children, with only 3 obtained from adults. There was a male predominance (58%).

Table 1. Details of 64 cases with congenital obstructions of the left atrial & left ventricular inlet tracts.

Study period	1991–2007
Total number of autopsied congenital heart disease	1633
Divided left atrium	09 (0.6%)
Supramitral ring	24 (1.5%)
Congenital mitral stenosis with hypoplastic left heart syndrome	09 (0.6%)
Congenital mitral valvar hypoplasia	14 (0.9%)
Reduplicated mitral valve	04 (0.2%)
Parachute mitral valve	04 (0.2%)

In 8 cases (33.3%), there was an incomplete form of Shone's complex, with the hearts exhibiting 2 or 3 of the 4 lesions described initially (Table 2). There were 7 children with an age range from 10 months to 14 years. They had a history of failure to thrive, delayed milestones, dyspnoea, easy fatigability, frequent infections of the respiratory tract, and/or repeated episodes of cardiac failure. A circumferential supramitral ridge was identified on echocardiography in 2 of them, which was excised (Fig. 1), combined with patch closure of a ventricular septal defect and excision of a sub-aortic fibrous shelf, respectively. In 3 children, coarctectomy had been undertaken. Among them, one (case 7) sustained a fatal traumatic head injury with bilateral carotico-cavernous fistulas, and had reduplication of the mitral valve with superimposed rheumatic heart disease (Fig. 2). The only adult was a 43 years male (case 8), who was diagnosed as a case of aortic stenosis. He had undergone repeated admissions in the past for cardiac failure, but had refused surgical intervention, leading to his present admission in a gasping state.

The remaining 16 patients (66.7%) had lesions other than Shone's complex (Table 3). Ventricular septal defects were present in 9 (56.3%, perimembranous in 3, muscular inlet in 2, doubly committed and sub-arterial in 2, and multiple in 2). Their ages ranged from 5 months to 25 years. Of the children, 5 males and 3 females presented with failure to thrive,

Table 2. Supramitral ring with Shone's complex.

S. no.	Age/gender	Shone's components	Morphology of supramitral ring	Associated mitral valvar pathology	Other associations
1.	10 months, Female	Coarctation (Coarctoplasty)	Complete; Annular; Thick and flap-like	Reduplicated mitral valve	Bicuspid aortic valve
2.	2 years, Female	Sub-aortic shelf	Incomplete & continuous; Supra-annular; Ridge-like	Dysplastic mitral valve	–
3.	4 years, Female	Coarctation	Complete; Annular; Thick and flap-like	Miniaturized mitral valve	–
4.	4 years, Male	Parachute mitral valve, Coarctation (Previous coarctoplasty)	Complete; Annular; Thick and flap-like (Excised)	–	Sutured patent foramen ovale, Patched perimembranous ventricular septal defect, Bicuspid aortic valve
5.	6 years, Male	Sub-aortic shelf (Excised)	Complete; Annular; Thick and flap-like (Excised)	Dysplastic mitral valve	Dysplastic bicuspid aortic valve
6.	10 years, Male	Sub-aortic shelf, Coarctation (Previous coarctoplasty)	Incomplete & continuous; Annular & valvar; Ridge-like	Miniaturized mitral valve	Bicuspid aortic valve
7.	14 years, Male	Coarctation (Previous coarctoplasty)	Incomplete & continuous; Annular; Ridge-like	Reduplicated mitral valve, Rheumatic mitral valvulitis (Previous balloon valvotomy)	Bicuspid aortic valve, Rheumatic tricuspid & aortic valvitis
8.	43 years, Male	Subaortic shelf	Incomplete & continuous, Supra-annular, Ridge-like	–	Bicuspid calcific aortic stenosis

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Figure 1.
Excised pieces of the supramitral ring, which are flap-like and translucent.

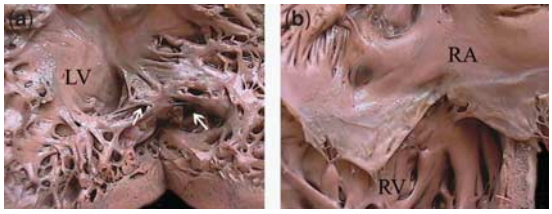

Figure 2.
Case 7 ***a)*** *Reduplicated mitral valve showing two orifices (arrows). There are hardly any intercordal spaces due to concomitant rheumatic mitral valvitis. (LV left ventricle),* ***b)*** *There was mild rheumatic affection of the tricuspid valve. (RA right atrium, RV right ventricle).*

breathlessness, and repeated respiratory tract infections for varying durations of time. In 4, the defects had been surgically closed. Cyanotic spells were an additional feature in the child where an inlet defect was associated with pulmonary infundibular stenosis. The defect was closed, and a Glenn shunt was performed. The adult was a female aged 25 years with 8 months amenorrhoea, who was admitted with respiratory distress. An emergency balloon valvotomy was performed for critical mitral stenosis. Autopsy revealed a supramitral ring (Fig. 3), as well as multiple ventricular septal defects.

In 4 others, there was associated complex congenital cyanotic heart disease in the form of tricuspid atresia, tetralogy of Fallot (Fig. 4), transposition, and double outlet right ventricle. The anomalies of the outflow tracts were corrected, but all patients died within 24 hours of operation. In 1 child, the supramitral ring was associated with congenital mitral valvar hypoplasia that was subjected to balloon valvotomy. There were no associated anomalies. Rheumatic mitral stenosis and degenerative aortic stenosis were the clinical diagnoses made in two males, aged 17 and 45 years, respectively. The first patient had undergone balloon mitral valvotomy and died subsequently of Penidure-induced anaphylaxis. The aortic valve was replaced in the second.

There was no difference in the completeness or width of the supramitral ridge in those with or without Shone's complex. Circumferential or complete ridges were identified in 7 patients, and these appeared thick, fleshy and fibrous (Figs 1 and 5). Incomplete rings were seen in the rest, as continuous (13) or discontinuous ridges (four). None of these were diagnosed on echocardiography. All of these appeared glistening, grey-white, and cord-like (Fig. 4). The rings were supra-annular in nine, located 0.2 to 0.7 centimetres above the mitral annulus. In 1 patient, the ring extended on to the atrial aspect of the aortic leaflet of the mitral valve (case 5, Fig. 6). In 13 patients, there was concomitant involvement of the mitral valve, either as congenital and/or acquired deformities.

Discussion

The supramitral ring, also known as supravalvar mitral stenosis, supravalvar stenotic mitral ring, supravalvar ring of the left atrium, supravalvar stenosing ring, or mitral supravalvar ring, was first described Fisher[4] in 1902, when the ridge of connective tissue above the mitral valve was likened to the diaphragm of the microscope. The defect has to be differentiated from the rare divided left atrium, where the partition is fibromuscular, separating the left atrium into the proximal and distal chambers.[5] Up to 2002, less than 100 cases of this rare malformation have been reported.[6] Consequently no data of the exact incidence or predilection for race or gender are available. The incidence is reported to range from one-eighth to two-fifths among congenital mitral valvar anomalies that were not necessarily stenotic.[2,7–10] All were children. In our analysis, the ring had an incidence of 1.5% among all congenital cardiac defects at autopsy, and accounted for just over one-third of all congenital lesions producing obstruction of the left ventricular inflow tract. Interestingly, 3 of our specimens were obtained from adults (20.8%). There was also distinctly a male predominance.

Most of the previous series do not particularly comment on the completeness or nature of the ring.[2,7–10] In a recent report of 15 children,[11] all had been circumferential, though incomplete rings have been described.[5] Circumferential ridges were seen in only 7 of our 24 hearts (29.1%), and these were entirely located on the valvar leaflets adjacent

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Table 3. Supramitral ring without Shone's complex.

S. no.	Age/gender	Main association	Morphology of supramitral ring	Associated mitral valvar pathology	Other associations
9.	5 months, Male	Perimembranous ventricular septal defect	Incomplete & continuous, Supra-annular, Ridge-like	Miniaturized mitral valve	Secundum atrial septal defect
10.	7 months, Female	Inlet ventricular septal defect	Incomplete & continuous, Supra-annular, Ridge-like	–	–
11.	1 year, Female	Inlet & perimembranous ventricular septal defects (patched)	Incomplete & discontinuous, Supra-annular, Ridge-like	–	Left ventricular non-compaction
12.	1 year, Male	Sub-arterial ventricular septal defect	Incomplete & discontinuous, Supra-annular, Ridge-like	–	Persistent left superior caval vein
13.	1 year, Male	Perimembranous ventricular septal defect (patched)	Incomplete & continuous, Supra-annular, Ridge-like	–	–
14.	2 years, Female	Perimembranous ventricular septal defect (patched)	Incomplete & continuous, Annular, Ridge-like	–	–
15.	3 years, Male	Sub-arterial ventricular septal defect (patched)	Incomplete & continuous, Annular, Ridge-like	Dysplastic mitral valve with accessory orifice	Dysplastic tricuspid valve
16.	5 years, Male	Inlet ventricular septal defect (patched)	Incomplete & continuous, Annular, Ridge-like	–	Infundibular pulmonary stenosis (Glenn's shunt), straddling tricuspid valve
17.	25 years, Female	Perimembranous & trabecular muscular ventricular septal defects	Incomplete & continuous, Annular, Ridge-like	Calcified rheumatic mitral stenosis (emergency balloon valvotomy)	–
18.	6 months, Male	Tricuspid atresia	Incomplete & discontinuous, Supra-annular, Ridge-like	Cleaved anterior mitral leaflet	
19.	33 years, Female	Tetralogy of Fallot	Incomplete & continuous, Annular, Ridge-like	Dysplastic mitral valve	–
20.	3 months, Female	Transposition	Incomplete & discontinuous; Supra-annular; Ridge-like	–	–
21.	6 years, Male	Double outlet right ventricle with sub-aortic ventricular septal defect (corrected)	Complete, Annular, Thick and flap-like	–	Persistent left superior caval vein
22.	4 years, Female	–	Complete, Annular, Thick and flap-like	Congenital mitral valve hypoplasia (Balloon dilatation performed)	–
23.	17 years, Male	–	Complete, Annular, Fleshy	Rheumatic mitral stenosis with balloon valvotomy-induced regurgitation	–
24.	45 years, Male	–	Incomplete & continuous, Annular, Ridge-like	–	Calcific aortic stenosis, Abnormal origin of left circumflex artery

to the annulus. On the other hand, the incomplete rings (70.9%) were found to be either continuous or discontinuous, and either annular or supraannular.

Even the morphology was different. Complete rings were always fleshy and fibrous, while the incomplete rings appeared akin to a tautened twine thread. The morphology also plays an important role in the clinical presentation as well as preoperative diagnosis. Though complete rings were identified in the study of Collison et al,[11]

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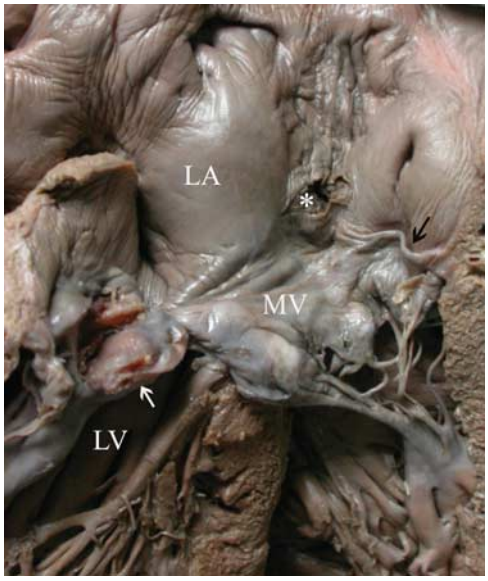

Figure 3.
Incomplete ridge (arrow) at the mitral annulus in case 17. Patient had undergone balloon mitral valvoplasty. Note atrial septostomy (*) and calcified postero-medial commissure. (LA left atrium, MV mitral valve, LV left ventricle).

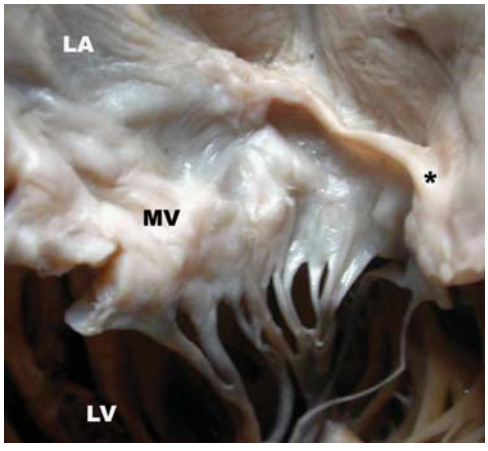

Figure 4.
The patient with Fallot's tetralogy showing an incomplete cord-like ridge (*) and dysplastic looking mitral valvar (MV) leaflets. (LA left atrium, LV left ventricle).

pre-operative diagnosis was made in three-quarters of the patients, the symptomatology largely being related to associated anomalies. These anomalies produce altered patterns in the flow of blood that may camouflage the ring. Similar observations were also made by Mychaskiw et al.[6] In our study, the ring was clinically identified in only 2 patients (8.3%). Hence, it is possible that the incidence of the ring may be underestimated, since all need not produce clinically overt stenosis, and all may not always be identified on routine echocardiography. Transoesophageal echocardiography is advocated for

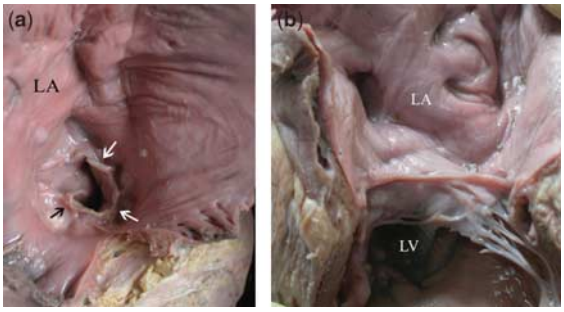

Figure 5.
***a)** View from the left atrium (LA), showing a circumferential annular mitral ring (arrows). Panel **b** shows the lesion after opening out the atrium and ventricle (LV).*

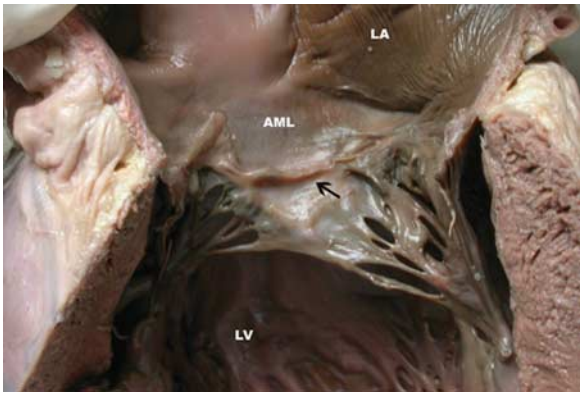

Figure 6.
A ridge which is annular at the attachment of the mural leaflet runs on to the mid-portion of the atrial aspect of aortic mitral leaflet. (LA left atrium, AML anterior mitral leaflet, LV left ventricle).

diagnosis.[12] Since stenosis is an inconstant feature,[11] the term supramitral ring is appropriate as used in isolation. We believe that all circumferential rings, as seen in three-tenths of our cases, were clinically significant. An abnormal mitral valve and/or an annular location of an incomplete ring, may lead to clinical relevance.

Isolated occurrence of the supramitral ridge was first reported by Chung and associates.[13] In most instances, nonetheless, the lesion occurs with other varied anomalies.[2,6,9,11,14] We did not find a single case of isolated ring. We found it useful to categorize the associations into those with or without Shone's complex. As has been the experience with others, the commonest association was a ventricular septal defect. Others also included malformations of the ventricular outflow tracts. Among those with involvement of the mitral valve, 13 hearts in all, we identified associated rheumatic mitral valve abnormalities in 3. As far as we are aware, this association has not previously been reported. There exists a possibility for the ridge to

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develop as a post-inflammatory phenomenon, which would mean recognition of such lesions in greater frequency. Apart from these reported cases, we have not seen them in our series of autopsied cases of rheumatic heart disease.[1] More interestingly, one of them also existed in a reduplicated valve. It is important to remember, therefore, that both congenital and rheumatic disease can occur in unison in countries with a high prevalence of rheumatic heart disease.[15] Additionally, we found the ring to be associated with even acquired aortic stenosis.

Excision, first performed by Lynch and associates,[16] has been the best therapeutic option,[11] as balloon valvoplasty may not always be successful.[17] The outcome also depends on the presence of simple or complex associations. Follow-up is essential since, at times, the lesion can recur, as seen in 4 of the 23 patients reported by Tulloh et al.[18]

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