Chiari Network with Papillary Fibroelastoma in Rheumatic Mitral Stenosis (*)

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The echocardiographic appearance of the Chiari network has previously been described but clinical significance of this embryological remnant has not been emphasized sufficiently.

A 27-year old male referred to our echocardiography laboratory in order to decide whether his rheumatic mitral stenosis was suitable for balloon valvuloplasty or not. Echocardiographic examination revealed a curvilinear mobile structure attached to the right atrial wall.This coincidental finding was subsequently diagnosed as Chiari network and the patient has undergone open heart surgery.

The reason for open commissurotomy was the risk of pulmonary embolism from the trombus formation on it or disruption of the small pieces of the network. Furthermore entrapment of the right heart catheter within the network may cause manipulation more difficult.

At operation Chiari network(1x1 cm. diameter) has been totally excised which was attached to the lateral border of the right atrium just beneath to the opening of inferior vena cava.

Histopathological findings were consistent with a characteristic papillary fibroelastoma.

Identification of Chiari network was important for two reasons in this case: First, tumoral transformation may occur from this normal variant and secondly balloon valvuloplasty should not be performed in these patient owing to its potential risks.

The echocardiographic identification of Chiari network is important for differential diagnosis of right atrial pathologies and for its potential risks [1,2,3].

The clinical significance and histopathological structure of this embryologic remnant has not been reviewed sufficiently. Since Chiari network is almost always asymptomatic on routine clinical grounds, the diagnosis is usually coincidental.

In this report a coincidental finding of Chiari network in a case with rheumatic mitral stenosis is presented.

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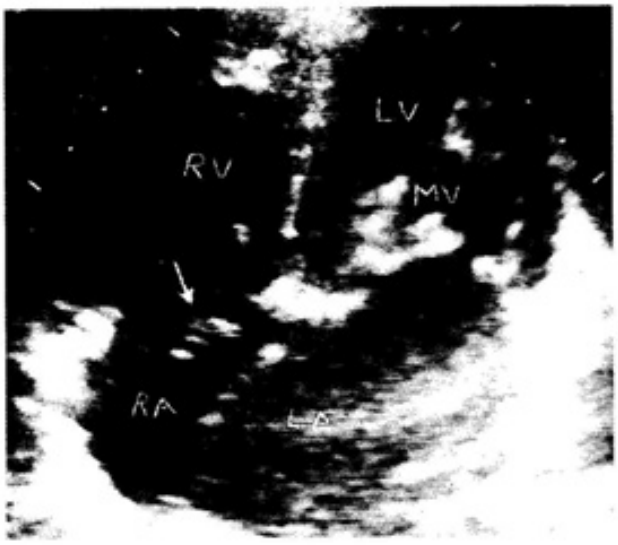

Fig. 1: Apical four chamber view of Chiari network with rheumatic mitral valve.

LV: Left ventricle, RV: Right ventricle,
MV: Mitral valve, RA: Right atrium,
LA: Left atrium.

Case

A 27-year old male with rheumatic heart disease was admitted to our center in order to decide whether he was suitable for balloon valvuloplasty. The clinical, electrocardiographic and echocardiographic studies revealed pure rheumatic mitral stenosis with a 16 mmHg pressure gradient. The patient was otherwise normal.

The apical four chamber view revealed a highly mobile, curvilinear structure attached to the right atrial wall (Fig. 1-2). Other routine echocardiographic findings were all within normal limits.

A preliminary diagnosis of Chiari network has been made. The patient was subsequently undergone to open commissurotomy. A network-like structure has been excised from the lateral border of the right atrium just beneath the opening of the inferior vena cava (Fig. 3). The histopathological examination revealed a characteristic papillary fibroelastoma (Fig. 4,5).

Discussion

Chiari network is considered to be a normal variant encountered in the right atrium[4,5]. It is an embryologic remnant which regresses during childhood but very occasionally it persists in adults. The differential diagnosis should exclude infectious right heart vegetations, thrombi, pedinculated tumours and ruptured chordae tendineae of the tricuspid apparatus[1]. In this case, the echocardiographic appearence was inconsistent with these possibilities. Since balloon valvuloplasty has potential risks of right atrial catheter into lesion[2,3], an open commissurotomy has been performed.

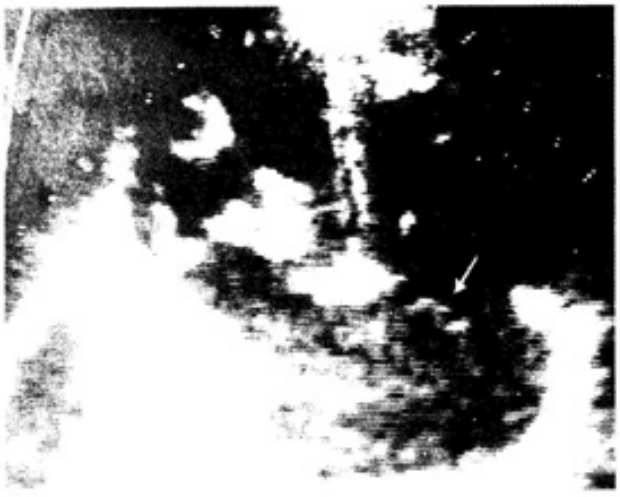

Fig. 2: Apical four chamber view of Chiari-network with rheumatic mitral valve.

Fig. 3: Chiari network within the right atrium

The histopathological evaluation of Chiari network is unavailable in the literature due to the lack of surgical indication. The excision of the network in our case was a part of open commissurotomy. A diagnosis of papillary fibroelastoma was made microscopically.

The histological examination of Chiari network should further be clarified in cases who undergone open heart surgery for another indication[5].

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Fig. 4: Microscopical view of papillary fibroelastoma.

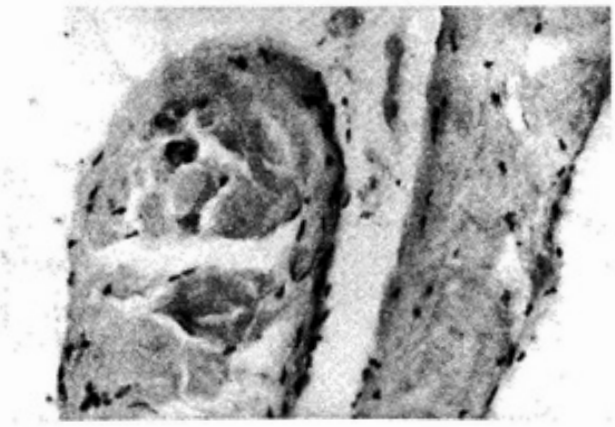

Fig. 5: Microscopical view of papillary fibroelastoma.

Small Aortic Annulus Problem in Aortic Valve Surgery (*)

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Small aortic root creates technical problems in aortic valve surgery. Effective orifices of 19 and 21 sized valves are the lower limits for a normal person and therefore should not be used unless under special circumstances. There have been various methods to enlarge the aortic annulus. Manouguian procedure is one of these.

Aortic valve surgery was performed in 254 patients between February 2, 1985 - April 1, 1989 at the Koşuyolu Heart and Research Hospital. In 11 of these patients (4.3%), we had to face the small aortic annulus problem. In these patients the aortic annulus had to be enlarged by Manouguian procedure so that AVR could be performed . The overall mortality rate was 9%.

In conclusion the Manouguian procedure seems a life saving method in the small aortic annulus problem. In selected patients it should be the first choice to select.

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Aortic valve replacement has become a simple procedure in most cardiac centers. However, the presence of small aortic root or hypoplastic left ventricular outflow tract may create technical problems to the surgeon in aortic valve surgery[4,6,10,26]. Because the major aim in aortic valve surgery is to relieve or not to augment the transvalvular gradient, so the effective orifice of the selected prosthetic valve should match the patient size and physical activity[3,4,6,10]. Effective orifices of 19 and 21 sized valves are the lower limits for a normal person, and therefore, valves under these sizes should not be used unless under special circumstances[3,13]. Otherwise, prosthetic valve mismatch will occur, resulting in left ventricular dysfunction, hemolysis, or necessity for reoperation with increased risk[10,18,20,24,25].

Procedures that can be used in cases with the small aortic root are as follows:

1- Supraannular placement of the prosthetic valve at the non-coronary cusp level (may not suffice, allows only one size larger valve to be implanted),

2- Supraannular enlargement of the aortic root with patch graft (used for supraannular stenosis),

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3- Aorto-ventriculo-plasties (The Konno's procedure),

4- Left ventricular apical-aortic valved conduits,

5- Supraannular - annular - subannular enlargement of the aortic root with patch graft (Manouguian procedure).

Between these techniques, the most suitable one seems to us the Manouguian procedure.

Material and Method

Between February 2, 1985 and April 1, 1989, 976 patients underwent valvular open heart surgery, and 254 of these were for the aortic valve. In eleven of these patients we encountered the small aortic root problem. Two of these patients were female and 9 were male; the youngest patient was18 years old, the oldest 42, and the mean age was 23 years old. Seven patients were in NYHA Class IV, and 4 in Class III. Cardiothoracic ratio was over 60% in 7 patients. Eight patients were admitted to the hospital in congestive heart failure. Also, 8 patients had pure aortic stenosis and the rest combined lesions. Three patients had mitral valve disease concomittanly and one had tripple valve disease.

Five patients were catheterized preoperatively. Transvalvular gradients were minimally 75 mmHg, maxi mally 120 mmHg with an average of 100 mmHg.

Surgical Technique

All patients prepared and anesthetized by our routine methods. Also, cardiopulmonary bypass were established and myocardial protection was achieved by our routine methods. Aortic valve exposure was provided with an oblique aortotomy on the anterior aorta, extending between left and non-coronary cusps annulus. Valvular stenosis was the predominant finding in all cases. All valves portrayed some degree of calcification. Leaflets were carefully resected, remaining calcification decalcified, and annulus diameter was measured[22,33]. Obtained diameters were 17 mm in 6 patients, 19 mm in 4, and 21 mm in 1 patient. The incision was extended from the intervalvular trigone towards the fibrous origin of anterior mitral leaflet, and when required, was further extended along the line of the leaflet[6,19,28,32]. However, care should be taken not to include the oppositional part of anterior mitral leaflet. Surgical view can be broadened by widening the atrial incision superiorly. 'U' shaped defect created between the anterior mitral leaflet and aortic root is closed with an appropriate graft material (pericardium, Gore-Tex, etc.)[5,19,21]. While implanting the prosthetic valve, pledgeted sutures were taken to outside along the non-coronary annulus[5,6]. The defect between the left atrium and the patch was closed with valve sutures. These sutures were pledgeted and tied in the extracardiac position: One patient received a Bjork Shiley prosthesis, while the rest St. Jude(SJM) valves. Pericardium in 1 patient and Gore-Tex in 10 patients were used as patch material. In all of the patients, the aortic root was enlarged by the Manouguian technique[19,21,32]. In one patient with segmental supraannular stenosis, supraannular Gore-Tex patch was used with a Bjork-Shiley sized 23 valve in the non-coronary supraannular position[1,24]. However, this patient was later reoperated due to paravalvular leak. Three patients

with mitral stenosis and insufficiency had concomittant mitral valve replacement, and 1 patient had concomittant mitral valve replacement together with tricuspid valvotomy. One patient received size 21, eight patients received size 23, two patients received size 25 prosthetic valves.

Results

One patient died at in-hospital period because of low cardiac output. Our hospital mortality rate is 9%. There was no serious complication in early postoperative period.

All living patients were followed-up from 6 to 47 months with a cummulative follow-up period of 22 patient-years. We have no late death. Also we have no late valve or procedure related complication. Eight of the survivors have become to functional class of I while the rest to class II. The survivors were examined by doppler echocardiography at the 2nd and 6th postoperative months.

Transaortic valvular systolic gradients decreased meanly from 75 mmHg to 10 mmHg. Also, it has been detected by these examinations that the ejection fraction increased meanly from 36% to 52%.

Discussion

Improvements in valve design have greatly diminished gradients in small sized valves. There are reports on the use of 19 mm, even 17 mm valves on appropriate patients[13,23].

When faced with a restrictive aortic root, the technically easiest solution is to implant the valve in the supraannular position. A second approach is to place a patch along the aortotomy incision[1,2,15,33]. Both approaches allow only one size larger valves to be implanted.

A different technique is enlargement of the root both at the annular and supraannular level. The method described by Nick and collagues[33], passes the aortotomy incision through the non-coronary sinus, devides the annulus, and extends the incision up to the fibrous origin of anterior mitral leaflet. Many modifications on this technique have been made, however, the enlargement can only be made up to two sizes[10,19,28,29,31].

In our patients we used the Manouguian technique. In our series, we used size 23 valves. Only one patient who was of small stature received size 21 SJM valve.

One of our patients died due to low cardiac output. This patient had undergone concomittant MVR and tricuspid valvotomy.

Three other patients had concomittant MVR.

In the other 7 patients, the incision on the mitral anterior leaflet and the placed patch did not result in mitral insufficiency[6,19,21,29].

One might feel the patch in Manouguian technique might cause prosthetic valve dysfunction, howe-ver, this complication is rare, because:

1- The patch area is relatively immobile during the cardiac cycle, and the anterior mitral leaflet is functionally passive.

2- The angle between the oppositional and nonoppositional faces of the anterior mitral leaflet reaches to 90 degrees during systol and finds more surface area with the posterior leaflet[19,21,32].

3- In patients with anterior leaflet prolapsus, the Manouguian technique gives good results.

4- A wide and improper patch which is inserted to the anterior mitral

leaflet position produces more prolapsus and mitral insufficiency. In the 7 non-MVR patients we did not have this problem.

5- If exploration of the mitral valve is planned, the Manouguian procedure should be the technique of choice.

The Manouguian technique should not be considered in minimal mitral insufficiency that does not require surgical correction and in presence of aorticomitral septal calcification that might increase the risk of surgical bleeding[7,8,19,21,26,30].

In aorticoventricular plasty, a longitudinal incision is made over the anterior ascending aorta, which in turn is extended between the free right ventricular wall and the septum, staying to the left of right coronary artery. With the placement of a patch into this incision, the annulus can be enlarged close to two fold[3,5]. The annulus and subvalvular outflow tract can be enlarged with aortoventricular plasties, however these techniques are unusually complex for a valve replacement procedure, which at the end increases morbidity and mortality rates.

In our clinic, we avoid to use of left ventricular apico-aortic valve conduits for valve replacements in small aortic root problem[4,13,14,17]. Some surgeons find the associated morbidity and mortality too high in the small aortic root enlargement procedures, and advocate to use of SJM size 19 valves instead[2,3]. In our clinic, the surgical mortality rate is 9%. The mortal case in our series was in NYHA Class IV, and underwent concomittant mitral valve replacement and tricuspid valvotomy. It is, therefore, difficult to blame the Manouguian procedure as the cause of death in this case.

Conclusion

The Manougiuan procedure in small aortic root problem could be performed in a significant number of patients undergoing aortic valve replacement. It could be technically easier, effective to enlarge the aortic root and could be performed with no significant increase in mortality and morbidity.

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