A Rare Morphological Variant Accessory Posterolateral Cusp of Mitral Valve with Accessory Papillary Muscle in Left Ventricle in An Adult Cadaver in Middle East-A Case Report

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Abstract: Various cases of accessory leaflets of mitral valve or tissue around the opening have been reported in the past. In some cases they were the main etiological factor for sub aortic stenosis or regurgitation. Here we report a rare finding of accessory posterolateral cusp of mitral valve with accessory papillary muscle in left ventricle of an adult male cadaver. Such variation has not been reported earlier in our region. We noticed additional commissure (as deep as rest of the other two) with chordae tendonae attached from the tip of additional papillary muscle. Such Variations may cause regurgitation or may compress outflow tract causing stenosis which may require their immediate removal. Awareness of such variations on the part of the surgical team is extremely important before operating in cases of mitral valve prolapse or regurgitation, aortic or mitral stenosis and left ventricular outflow tract obstruction and various valvular repair and graft surgeries. Knowledge of these would help in proper designing and placement of valve prosthesis.

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1. Introduction

Heart is a king, who rules over all the organs of the body; the lung are his executives, who carry out his orders." was written by Huang Ti in 2600 B.C.(1)in his "Canon of Medicine" Human mitral valve is a very complex, dynamic, and highly variable structure. Rusted et al in 1952 reported that the mitral valve has two major leaflets. They identified the commissures using the tips of the papillary muscles on either side as a guide. The papillary muscles were used as a landmark for the commissure. (2)

Mitral valve is described as consisting of a continuous veil attached around the entire circumference of the mitral orifice with its free edge bearing several indentations; two are sufficiently deep and regular to be nominated as commissure. It is more usual, however, for these anteromedial and posterolateral extremities to be designated as two independent commissures. The official names for these cusps, anterior and posterior. The anterior cusp is seen to guard one-third of the circumference of the orifice and to be semicircular or triangular, with few or no marginal indentations. The posterior cusp guards two-thirds of the circumferential attachments. Further indentations usually divide the posterior cusp into a relatively large middle scallop and smaller anterolateral and posteromedial commissural scallops. (3). The term cleft for a division between scallops on either the anterior or posterior leaflet.

Current nomenclature describes it as a bileaflet valve with chordae tendonae connecting the leaflets to two ventricular papillary muscles. Kumar and colleagues went on to classify the leaflets of the mitral valve using a numbering scheme for the scallops (4); however, Carpentier's description remains the most commonly used.(5)

2. Case Report

The purpose of present commentary is to report a rare accessory posterolateral cusp (APLC) of mitral valve with and accessory posterolateral papillary muscle (PLPM) which was encountered incidentally during routine cadaveric dissection of 39 yr old male cadaver who died of pneumonia. The rare finding of APLC of mitral valve was seen in left atrio ventricular opening arising from posterolateral left ventricular wall. APLC was seen to be on posterolateral side (shown as white Star In Figure 1)and was 2.5 cm wide and 1.5 cm at narrowest point. On its free margin it had attached chordae tendonae arising from tip of accessory postero-lateral papillary muscle (PLPM). Strikingly, accessory posterior cusp had three sub scallops with two indentations. The rare finding consisted of three sub scallops with two indentations in between. It extended from antero-medial commissure (C1) to additional posterolateral commissure (C3). The posteromedial cusp extended from posterolateral commissure (C3) to posteromedial commissure (C2). Anteromedial cusp (shown with

black star) extended from C2 to C1.Cavity of left ventricle was measured to be 9.7 cm in length and 5.2 cm in width. Accessory posterolateral papillary muscle was 2.5 cm in length had separate base from posteromedial papillary muscle. Tip of both muscles gave rise to strut of chordae tendonae attached to the free margin of the cusps Left ventricular outflow tract was measured to be 3.5 cm in circumference with usual three aortic cusps at its margin. Left atrioventricular opening was 4.3 cm in circumference.

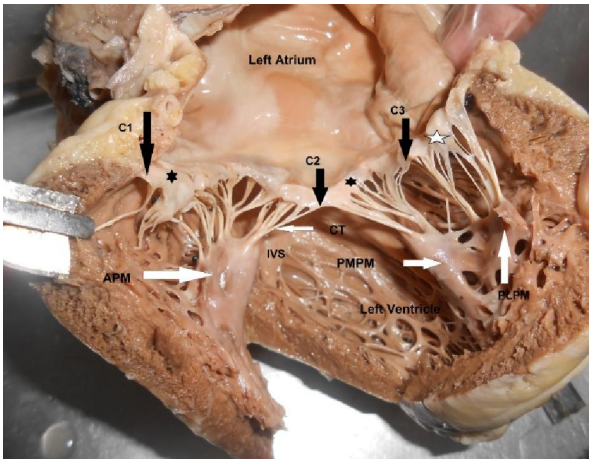

Figure 1 Showing-

White Star Shows Accessory Postero Lateral Cusp (APLC) of Mitral Valve.

Two Black Star Represents Anteromedial and Posteromedial Leaflet of Mitral Valve from C1 to C3.

Anterior Papillary Muscle (APM), Postero-Medial Papillary Muscle (PMPM); Postero-Medial Papillary Muscle (PLPM) Chordae Tendonae (CT); Interventricular septum (IVS).

C1-Anteromedial Commissure.

C2- Posteromedial Commissure.

C3- Additional Posterolateral Commissure.

3. Discussion

During the development of mesenchymal cells of cardiac tube around the right and left atrioventricular canals proliferate to form collars of endocardial cushions. The ventricular surface of the proliferated mass is excavated to form the atrioventricular valves which are attached to the ventricular wall by the trabeculae. (6) Leaflets of the tricuspid valve were considered to develop equally from the endocardial cushion tissues and the myocardium The leaflets and tensile apparatus of the atrioventricular valves was formed by a process of delamination of the inner layers of the inlet zone of the ventricles.(7)

Ranganathan et al, 1970 recognized unique commissural chordae tendonae that inserted in 98% hearts were useful definitive landmarks. The posteromedial commissural chorda was thicker and longer branches and a considerably wider spread than its anterolateral counterpart, valve chordae tendonae. They described posterior leaflet as one unit which is constantly notched and is divisible into three scallops in 92% of the hearts examined. They believed that the middle scallop has been mistaken as the entire posterior leaflet. They described an ‘‘edge zone,’’ a ‘‘clear zone,’’ and a ‘‘basal zone’’ based upon the thickness and location of these leaflet tissues. (8)

Most common and widely accepted configuration for leaflet anatomy was the description provided by Carpentier, 1976 described 3 posterior leaflet scallops with 2 clefts separating them, 2 commissures separating the anterior and posterior leaflet, and 1 anterior scallop. Carpentier nomenclature for locations of each cleft within the anterior and posterior leaflets. If a cleft was found in either of the 2 locations described by Carpentier, it was referred to as a ‘‘standard’’ cleft. Any clefts that were found in regions described as being scalloped regions by the current nomenclature were termed ‘‘deviant’’ clefts.(5). Our finding showed three prominent junction of three scallops and was considered as commissures and not clefts as they were deep and had attached chordae from variant papillary muscle.

Quill et al in 2009 proposed leaflet nomenclature for surgeons typically describing an anterior and posterior leaflet, each divided by 2 commissures into the A1, A2, and A3 and P1, P2, and P3 scallops. The term cleft was used for a division between scallops on either the anterior or posterior leaflet. The average number of anterior leaflet scallops was 1.2 ± 0.4, and the average number of posterior leaflet scallops was 3.0 ± 1.0. They described variation in mitral valve anatomy is to describe the clefts as either standard or deviant The standard cleft that separates P1 from P2 was found in 66% of the hearts and the standard cleft that separates P2 from P3 was present in 71% of the hearts Our case C3 was present at same region as of junction between P1 and P2. In contrast to their finding we found accessory posterolateral muscle with chordae attached to C3. They found anterior leaflets with fewer deviant clefts than the posterior leaflets, and deviant clefts appeared most often in the P2 regions. (9)

Monibi et al described 69% hearts having abnormal contour of the left ventricle with or without associated prolapsed mitral valve. These consisted of abnormality of contour and/or contractility 23 % of these had a dyskinetic and hypokinetic left ventricle. 38% patients were having moderate to severe prolapse of the mitral valve with abnormally functioning ventricle. The echocardiogram did not prove helpful in

detecting the left ventricular or mitral valve abnormalities (10).

Hartynski et al reported a case with parachute-like accessory mitral valve leaflet and tissue attaching to the anterior leaflet, ballooning into the subaortic ventricular septum associated with a discrete subaortic membrane causing stenosis. They emphasized intraoperative epicardial or transesophageal echocardiography for recognition of such defects.(11)

Rodrigo et al reported cases with double orifice mitral valve with anomaly of the tensor apparatus double-parachute mitral valve because each of the 2 mitral orifices inserts exclusively into fused papillary muscles.(12)

Meldrum et al reported three patients with accessory mitral valve tissue which caused left ventricular outflow tract obstruction in two children with congenitally corrected transposition and in one child with normally connected great vessels. This tissue was attached by chordae-like strands to pulmonary and atrioventricular tissue and to the superior edges of the ventricular septal defect. The accessory tissue obstructed both the ventricular septal defect and the subpulmonary region. Another patient, without ventricular septal defect, had valve tissue, without chordae, attached to the ventricular aspect of the atrioventricular valve.(13)

Levy et al in 1963 described three necropsy specimens from patients with congenitally corrected transposition in which two had "umbrella" like formations of accessory valvular tissue protruding into the subpulmonary area.(14)

MacLean et al reported subaortic stenosis caused by accessory tissue on the mitral valve which formed two cup-shaped "diverticula" on the ventricular surface of the anterior leaflet of a patient presenting with dextrocardia(15)

Junsono et al 1988 reported a case with arrhythmias, aortic regurgitation, and symptoms of severe intermittent ventricular outflow obstruction caused by an accessory mitral leaflet. (16)

Tanaka et al, 2006 reported report a case of accessory mitral valve that caused both left ventricular outflow tract obstruction and mitral insufficiency. The membrane insertion of the accessory mitral valve was to aortomitral continuity. Use of echocardiography was emphasized for the diagnosis of accessory mitral valve.(17)

Kuhne et al 2006 reported isolated cleft of the mildly thickened posterior mitral valve leaflet with a moderate late systolic mitral regurgitation. (18)

Mishra et al, 2014 described shape of both anterior and posterior cusps the most frequent shape encountered was triangular in 75-80% Three out of 120 cases, the mitral valve showed accessory triangular leaflet.(19)

A retrospective analysis of ten isolated cleft mitral valve leaflets with none of them affecting the posterior leaflet.(20). Cleft mitral valve leaflet may result in severe mitral regurgitation and the need for surgical repair (21). Admittedly we have not conducted histological evaluation of accessory cusp and papillary muscle.

Radiologists and surgeons should be aware of such variations as they could be main etiological factor in various clinical conditions due to compression and therefore may require resection or replacement. Such variations are commonly associated with other congenital malformations. They are important in various valvular repair and graft surgeries for better understanding of operative field. We suggest use of both Carpentier and Quills nomenclature with tip of papillary muscle and attachment of chordae tendonae to the cusps as landmarks for classifying cusps for both anatomists and surgeon so as to better define its morphology and variations. Particularly in light of the increasing number of cardiac interventions, knowledge of normal and variant anatomic features has become more important than in the past.

4. Conclusion

At present, every practicing radiologist and surgeon must have in-depth knowledge of such variations for better understanding of associated clinical conditions such as and mitral and aortic stenosis and regurgitation. Functional anatomical studies can provide new insights that are difficult to observe in conventional anatomy studies. Further anatomic studies of the mitral leaflets are typically analyzed with the valve in the systolic position should be conducted to better analyze cusps and commissures. Such diversity in the shapes of valves is of immense value in designing prototype of mitral valve prosthesis. Pre-operative radiological analysis such as catheterization of the left side of the heart with selective left ventricular cineangiocardiography or 3-D echocardiography should always be performed for better understanding of structures in operative field.

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