Anomalous Left Atrial Chorda and its Association with Mitral Regurgitation

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ABSTRACT

Anomalous left atrial chorda is associated with mitral regurgitation. A young woman presenting for mitral valve repair with the diagnosis of mid-segment (A2) of anterior mitral leaflet prolapse causing severe mitral regurgitation. Transesophageal echocardiography examination in pre-bypass period showed an anomalous chorda attaching A2 to the left atrial roof, tethering the anterior mitral leaflet toward the atrial wall. Surgical findings confirmed the abnormally attached chordae and an absence of normal chorda of A2 segment. The anomalous chorda was resected and neo-chordae placed between the A2 segment and papillary muscles and annulus strengthened with an annuloplasty ring.

Keywords: Anomalous chordae, left atrial chordae, mitral regurgitation

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INTRODUCTION

Causes of mitral regurgitation (MR) are varied, ranging from rheumatic heart disease, degenerative valvular disease, ischemic heart disease, infective endocarditis, dilated cardiomyopathy, or a part of congenital heart disease such as atrioventricular septal defect. We describe presence of a chorda-/band-like structure joining the roof of left atrium (LA) and A2 segment of anterior mitral leaflet (AML) and its role in the genesis of MR. The band was detected incidentally during pre-bypass transesophageal echocardiography examination. Earlier, 3 similar cases have been described in the literature.

CASE HISTORY

A 27-year-old woman weighing 62 kg presented with dyspnea on exertion since past 6 months. At an age of 8 years, she

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was incidentally diagnosed to have mitral valve prolapse but at that time she did not have any cardiac symptoms. There was no history of rheumatic fever. There were no comorbidities except history of coronavirus disease-2019, a year ago from which she recovered completely. Her transthoracic echocardiography (TTE) examination showed dilated LA and left ventricle–48 \times 52 \times 69 mm and 50 \times 35 mm, respectively; prolapse of the A2 segment of AML causing a posteriorly directed severe MR, and good biventricular functions. She was scheduled for mitral valve repair.

In the operating room, standard ASA (American Society of Anesthesiologists) monitors were attached and a radial arterial access was taken under local analgesia. Anesthesia was induced with fentanyl 2 μ g/kg and propofol 1.5 mg/kg; rocuronium 1 mg/kg was administered

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to facilitate tracheal intubation with a 7.5 mm (ID) endotracheal tube. A triple-lumen central venous catheter and a pulmonary artery catheter was placed as per the institute protocol. During pre-bypass period, transesophageal echocardiography (TEE) examination in mid-esophageal (ME) long axis, commissural and 4 chamber views confirmed the presence of MR [Video 1]; there was an evident prolapse of the A2 segment. The leaflet looked normal and it was not thickened (2 mm). No leaflet redundancy, cleft(s), annular dilatation, or calcification(s) was found in ME 4-chamber view, ME commissural view, ME 2-chamber view, ME long axis view, and a 3-dimensional zoom en face view. There was no mitral annular disjunction or ruptured chordae or myocardial wall motion abnormality. A chorda-/band-like structure, originating from the roof of the left atrial wall facing the ascending aorta and attaching to the atrial surface of the A2 segment of AML was noted and it appeared to tether it (A2) toward the roof of LA [Videos 2 and 3; Figure 1], the band was considered to be an anomalous accessory-chorda. The presence of the chorda was not reported in the TTE examination. On standard mild hypothermic cardiopulmonary bypass (CPB) and under cardioplegic arrest, the mitral valve was examined, the mitral valve leaflets were normal, one chorda to the A2 segment was absent, and the band noted during TEE examination was found to tether the A2 segment of AML to the roof of LA [Figure 2]. The valve repair included resection of the chorda originating from the roof of the LA and attaching to the A2 segment of AML, implantation of 3 artificial chordae, 1 between the edge of the prolapsing A2 and anterolateral papillary muscle, and 2 between the edge of A2 and posteromedial papillary muscle, and implantation of a 28-mm Physio II annuloplasty ring. Following repair, the patient was warmed and separated from CPB. On TEE examination, an eccentric wall-hugging residual MR jet was noted at the posteromedial commissure, which was absent earlier, in the pre-repair echo [Figure 3]. CPB was reinstituted and posteromedial commissuroplasty was performed. The patient was weaned from CPB. Repeat TEE examination showed trivial MR. Anticoagulation was reversed and after surgical completion, the patient was shifted to intensive care unit. Her postoperative recovery in the intensive care unit remained uneventful. TTE examination before discharge showed a trivial residual MR.

DISCUSSION

Patrianakos *et al.*^[11] reported 4 cases of anomalous left atrial chordae in 6500 autopsies. Using key words anomalous, accessory chordae, and LA yielded several papers describing development of normal chordae, its incidental presence,



Figure 1: Static image of the prolapsing A2 segment and the chorda arising from it attaching to the left atrium



Figure 2: Surgical image of the chorda attaching to the A2 segment of the anterior mitral leaflet and left atrial wall



Figure 3: Mitral valve navigation image showing the lone A2 prolapse, with rest of the segments and commissures normal

and treatment, however, embryological development of abnormal chorda was not found $^{\left[2,3\right] }$

MR generally occur as a consequence of rheumatic heart disease, or degenerative valvular heart disease or ischemic heart disease. Our patient presented with MR at an age of 27 years and did not have history of rheumatic heart disease or connective tissue disorder. Gurrieri et al., Vlassak et al., and Alghamdi et al.[4-6] described presence of accessory chorda and MR in young patients; however, the proposed mechanism of MR was different in all the 3 patients. Mechanisms described for MR included the following: leaflet prolapse, leaflet tethering, cleft, perforation, and annular dilatation, which may escalate tethering. In the case described by Gurrieri et al.[4] the anomalous chorda originated from the posteromedial papillary muscle, coursed through the valve, attaching onto the LA wall, and impaired coaptation of leaflets, causing MR. Vlassak et al. and Alghamdi et al.[5,6] described the chordal attachment between interatrial septum and A2 causing MR. We believe that in our patient, the MR was secondary to tethering of the A2 and pulling it toward the LA roof resulting in prolapse of the A2. Additionally, in our patient, one primary chorda to the A2 was absent, perhaps, enhancing the pulling effect of accessory chorda. In our patient, except A2 prolapse no other mechanism of MR was found to be operating.

On echocardiography, the anomalous chordae appear similar to normal chordae but in some instance, they have a reticular structure that can be misinterpreted as vegetations.^[7] Other possible conditions that may resemble the anomalous chordae are cor-triatriatum sinister, atrial tumors, and thrombus. The characteristics feature of independent mobility usually helps differentiate the tumors and thrombus. To summarize, in a young patient presenting with MR, during pre-bypass TEE examination, one should look for other causes of MR also including the presence of an anomalous chordal attachment even if they are not reported during TTE examination.

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Conflicts of interest

There are no conflicts of interest.

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