CASE REPORT

Severe mitral regurgitation due to anterior mitral leaflet perforation after surgical treatment of discrete subaortic stenosis

Arda Ozyuksel, ¹ Ozgur Yildirim, ² Ibrahim Onsel, ³ Mehmet Salih Bilal²

¹Department of Cardiovascular Surgery, Medipol University, Istanbul, Turkey ²Department of Cardiovascular Surgery, Medicana International Hospital, Istanbul, Turkey ³Department of Anesthesiology, Medicana International Hospital, Istanbul,

Correspondence to Dr Arda Ozyuksel, ozyukselarda@yahoo.com

Turkey

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SUMMARY

Congenital subvalvular aortic stenosis may be associated with anomalies of the mitral valve. In this case, we present a patient with severe mitral valve regurgitation due to a perforation in the anterior mitral leaflet detected 4 months after an operation for relief of subaortic stenosis. A 10-year-old male patient who was operated for subvalvular aortic stenosis in another clinic was admitted to our hospital, and transthoracic echocardiography revealed severe mitral valve regurgitation due to a defect that was demonstrated at the anterior valve leaflet. The perforated area at the mitral valve zone A1 was repaired with a PTFE patch. The patient was successfully operated for the mitral valve perforation and the postoperative course was uneventful. In our case, the perforation in the anterior mitral leaflet implies a possible implementation of inappropriate surgical technique which necessitated a second surgical intervention after the initial operation.

BACKGROUND

Congenital aortic stenosis is traditionally classified as supravalvular, valvular and subvalvular. Discrete membranous subaortic stenosis (DMSS) is the most common form of congenital subvalvular aortic stenosis. Besides a localised discrete shelf, additional subaortic anomalies may be encountered which may be the accompanying factors in cases with recurrent subaortic stenosis. DMSS may be associated with anomalies of the mitral valve. In this case, we present a patient with severe mitral valve regurgitation due to a perforation in the anterior mitral leaflet 4 months after surgical intervention for relief of DMSS.

CASE PRESENTATION

A 10-year-old male patient was admitted to our clinic with dyspnoea and palpitation. He was diagnosed with DMSS and operated on in a cardiovascular surgery clinic in a foreign country 4 months ago, and at that time, any mitral valve regurgitation was not noted in the preoperative evaluation. In the follow-up, moderate-to-severe mitral as well as moderate aortic regurgitation was determined and the patient was referred to our clinic. Physical examination revealed pansystolic murmur (4/6°) at the left fifth intercostal space. Transthoracic echocardiography revealed severe mitral and mild aortic regurgitation with moderate dilation of the left atrium and ventricle (left atrium diameter: 38 mm, left ventricle end diastolic diameter: 44 mm; figure 1). Residual



Figure 1 Echocardiographic image demonstrates the perforation and regurgitant jet on the anterior leaflet (arrow).

subaortic stenosis was not encountered. Mitral regurgitation was due to the defect which was demonstrated at the anterior valve leaflet. The patient was operated on electively. Following midline sternotomy and institution of standard hypothermic cardiopulmonary bypass (32°C), the aorta was cross clamped. Antegrade multidose hypothermic blood cardioplegia was administered. Mitral valve was exposed through transseptal approach following right atriotomy. A perforated area was demonstrated at the anterior leaflet zone A1, adjacent to anterior leaflet annulus and anterolateral commissure with a diameter of 10 mm (figure 2). The defect was repaired with a PTFE patch using continuous 7/0 PTFE suture. In order to reduce the annular dilation, annuloplasty was performed with the aid of two single 5/0 polypropylene sutures at two sides of the posterior annulus (figure 3). Subvalvular apparatus was morphologically normal. Following saline injection into the left ventricle, mild central mitral regurgitation was noted. Interatrial septum was primarily sutured. Following the deairing and closure of the right atrium, the cross clamp was released. Cardiopulmonary bypass was terminated and mitral valve function was evaluated with transoesophageal echocardiography which did not reveal any residual regurgitation at the mitral valve. Aortic cross clamping and cardiopulmonary bypass times were 44 and 65 min, respectively. The postoperative course was uneventful and the patient was discharged 10 days after operation.



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Unusual presentation of more common disease/injury

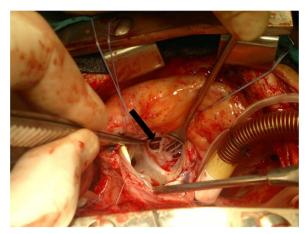


Figure 2 Exposure of the anterior mitral valve and area of perforation at zone A1 (arrow).

DIFFERENTIAL DIAGNOSIS

In this patient, any congenital or acquired aetiology of mitral valve insufficiency should be ruled out. Such perforations in the mitral valve leaflets can be encountered as 'drop lesions' in infective endocarditis of the aortic valve.

DISCUSSION

Congenital subvalvular aortic stenosis includes a wide spectrum of conditions ranging from DMSS to tunnel-like stenosis and may be accompanied with mitral valvular anomalies. Cohen $et\ al^3$ studied the anatomy of mitral valve in patients undergoing surgical resection of discrete subaortic stenosis and grouped mitral valvular anomalies into five categories: (1) insertion of a papillary muscle into the aortic leaflet, (2) insertion of a papillary muscle into the ventricular wall, (3) muscularisation of the subaortic portion of the aortic leaflet, (4) anomalous insertion of the valvular tissue into the ventricular wall and (5) accessory valvular tissue. The incidence of one or more mitral valvular anomaly accompanying discrete subaortic stenosis was 48% in their patient group.

Although rare, mitral leaflet tears may be encountered as complications during surgical resection of subaortic membrane that may necessitate mitral valve repair.⁴ Mitral valve regurgitation may be encountered early in the postoperative course as in our case, or may be complicated by mitral valvular lesions even years after initial intervention.⁵ Valvular pathologies in these

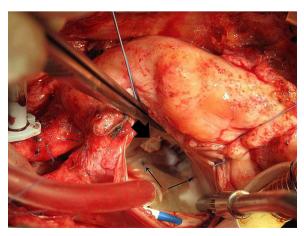


Figure 3 Area of perforation is repaired with a PTFE patch (arrow head) and annuloplasty is completed with two polypropylene sutures (arrows).

patients should be considered as 'multivalvular' diseases; therefore, close follow-up for the previously 'normal' valves is mandatory in the postoperative period.

In our case, mitral regurgitation was due to a perforation in the anterior mitral leaflet without any other intrinsic valvular anomalies. In some cases, accompanying papillary muscle and chordae tendineae anomalies may result in traction at the mitral leaflet. Allwork et al demonstrated microscopic and macroscopic anomalies in the resected mitral valve tissues in patients operated for mitral valve replacement following surgical relief of subaortic stenosis. They suggest that myocytolysis, vasculitis and fibrosis observed in the leaflets may be the accounting factors leading to mitral valvular malfunction in these patients. In our patient, mitral valve and subvalvular apparatus was explored and any intrinsic gross anomaly was not encountered. This condition implies a possible implementation of inappropriate surgical technique resulting in valvular perforation. Rather than mitral valve replacement, we preferred to repair the defect with a PTFE patch. We also preferred suture annuloplasty instead of a rigid ring annuloplasty. We believe this surgical technique resulted in a more physiological valve and annulus function.

Learning points

- ➤ Valvular pathologies may be encountered as isolated or combined clinical presentations in children.
- ► The previously 'normal' valves should be carefully evaluated in the follow-up period in children with a history of cardiac surgery.
- Infective endocarditis and iatrogenic aetiologies must be ruled out when a recent valvular pathology is encountered in a patient with a medical history of cardiac surgery.
- Meticulous surgical technique is mandatory in order not to result in stretching of the leaflet tissue, altered three-dimensional geometry and function.

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