Accessory mitral valve leaflet causing aortic regurgitation and left ventricular outflow tract obstruction

Case report and review of published reports

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From the Royal Liverpool Children's Hospital

SUMMARY Arrhythmias, aortic regurgitation, and symptoms of severe intermittent ventricular outflow obstruction developed in a 14 year old boy with a heart murmur who had been followed from infancy. These were caused by an accessory mitral leaflet, which was successfully removed at open heart operation. A review of 21 previously reported cases found a high incidence of associated cardiac malformations, appreciable subaortic obstruction in most patients, and a consistent attachment of the accessory tissue to the ventricular aspect of the anterior mitral leaflet. The characteristic echocardiographic appearance of a mobile mass arising from the area of aortic-mitral continuity is sufficient for the diagnosis of accessory mitral leaflet and echocardiographic examination will facilitate the surgical management of this condition.

Among the more rare causes of subaortic obstruction are several mitral valve abnormalities, including abnormal insertion within the left ventricular outflow tract, prolapse of redundant chordae or a leaflet, and accessory tissue in the subaortic area. Such accessory tissue is uncommon in normally connected hearts. It is a condition singularly amenable to repair without valve replacement. We report a case with several unusual clinical features that illustrates the value of real time echocardiography in diagnosing left ventricular outflow tract obstruction with aortic regurgitation caused by an accessory mitral leaflet.

Case report

A 14 year old boy was admitted to hospital with a two month history of a central stabbing chest pain. He had been seen first at 11 months for an asymptomatic heart murmur that was present at birth, and right heart catheterisation subsequently confirmed the clinical diagnosis of a small ventricular septal defect. Routine outpatient follow up found no change in his physical signs until an early diastolic murmur was heard when he was 14. The patient then recalled several episodes of sharp precordial pain precipitated by strenuous exercise, as well as palpitation and giddy spells unrelated to effort.

The relevant physical findings included an irregularly irregular waterhammer pulse at 80 beats/minute, a systemic blood pressure of 130/70 mm Hg, and an easily palpable thrill along the left sternal border. This corresponded to a harsh systolic ejection murmur radiating widely over the precordium as well as to the patient’s neck and back. In addition, there was a grade 2/4 immediate diastolic murmur. The electrocardiogram showed a bizarre pattern of multifocal ventricular extrasystoles, some periods of sinus rhythm with intermittent right bundle branch block, and other periods of junctional rhythm. There was no evidence of ventricular hypertrophy or ischaemia. Chest x ray showed a cardiothoracic ratio of 0.53 with pulmonary vasculature. The remainder of the patient's physical examination and laboratory investigations were within normal limits.

Cross sectional echocardiography showed a pedunculated mass originating just beneath the non-coronary cusp. The mass had prolapsed through the aortic valve during systole (fig 1). In addition, there was a large aneurysm of the membranous septum. Pulsed Doppler examination showed moderately

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beneath the commissure between non-coronary and right cusps there was a wide-mouthed aneurysm (20 mm in diameter) of the membranous septum extending into the right ventricle. There was no ventricular septal defect; but, at the lower margin of the aneurysm, a discrete ring of fibrous tissue extended into the outflow tract for about 5 mm and around half its circumference. This did not correspond with the echocardiographic appearances, however, and accordingly a search was made deeper within the left ventricle. This disclosed a mass of soft white tissue attached in several places to the ventricular surface of the anterior mitral leaflet and, on the other side, by well defined chordae, to a small papillary muscle near the ventricular septum (fig 2). This tissue was completely separate from both the subaortic membrane and the aneurysm of the membranous septum (fig 3).

severe aortic regurgitation and a velocity of 2.3 metres/second across the left ventricular outflow tract, indicating a gradient of approximately 21 mm Hg. Repeat cardiac catheterisation and angiography confirmed the echocardiographic findings and excluded any intracardiac shunt.

Despite the absence of clinical or laboratory findings to suggest bacterial endocarditis, the echocardiographic appearances were thought to be consistent with a large vegetation and the patient was treated with intravenous antibiotics. Four days after catheterisation his giddy spells recurred, and urgent transaortic exploration of the left ventricular outflow tract was carried out on cardiopulmonary bypass with moderate hypothermia and cardioplegic arrest. The aortic valve was normal, and immediately

Fig 1  Cross sectional long axis echocardiograms in systole (a) and diastole (b) showing the prolapse of a large mass of tissue (small arrows) through the aortic valve during ventricular ejection. This tissue fell back against the aneurysm of the membranous septum (large arrow) during diastole. VS, ventricular septum; LV, left ventricle; AO, aorta; LA, left atrium; AM, accessory mitral leaflet.

Fig 2  Transluminated surgical specimen showing the aortic (a) and ventricular (b) sides of the accessory mitral leaflet. The papillary muscle arose near the normal anterolateral papillary muscle and the upper lip was attached to the back of the anterior mitral leaflet in several places. A rudimentary chordal architecture and cusp differentiation is apparent but the leaflet was imperforate.
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The discrete subaortic membrane was enucleated, and the accessory mitral leaflet was excised by sharp dissection. The aneurysm of the membranous septum was plicated on its right ventricular side through the tricuspid valve. Weaning from cardiopulmonary bypass was smooth and uneventful, as was the patient’s subsequent postoperative recovery. He was discharged from hospital seven days after operation, when pulsed Doppler echocardiography showed only a trace of aortic regurgitation. The electrocardiogram had returned to normal sinus rhythm.

Discussion

Accessory mitral valve tissue producing subaortic obstruction was first described by MacLean et al in 1963; and, in his precise classification, Edwards included this anomaly as a rare cause of subaortic stenosis. During the past decade one or two cases have been reported each year, and it is now possible to characterise this unusual condition in some detail.

The condition usually presents as an asymptomatic heart murmur, although the two youngest patients had low cardiac output from aortic obstruction at three days of age, and congestive heart failure at 14 months. Signs of left ventricular outflow tract obstruction generally develop during the first decade (fig 4) but these have been accompanied by complaints of exercise intolerance, chest pain on exertion, or syncope in less than one third of patients. Usually, the obstruction is severe, with gradients of >50 mm Hg measured by cardiac catheterisation or Doppler echocardiography. There were major associated cardiac defects in 13 of 21 previously described cases and in our patient, but these did not suggest any particular developmental pattern (table).

Our patient was unusual because he presented with dominant aortic regurgitation and arrhythmias, which, in view of his past history of ventricular septal defect, suggested bacterial endocarditis. It is possible that the aneurysm of the membranous septum provided a bypass around the obstructing accessory mitral tissue until this tissue became sufficiently large to prolapse through the aortic valve. The only other patient to have such a mild gradient across the left ventricular outflow tract also had an aneurysm of the membranous septum. Sudden complete blockage of aortic outflow would account for the patient’s chest pain and giddy spells, but the aetiology of his arrhythmias is less clear. One possibility is that increased left ventricular pressure caused tension on the aneurysm and adjacent conduction tissue, resulting in bundle branch block and the episodes of nodal rhythm. Alternatively, the accessory leaflet itself may have damaged the conduction tissue as it moved within the subaortic area. In either event, the electrocardiogram would be expected to revert to normal, as it did, after operation.

The diagnosis of this and other types of subaortic obstruction has been facilitated greatly by real time echocardiography. Although M mode studies showed multiple echoes in the subaortic area, sector scanning clearly differentiates between fixed fibrous tissue and movement of an accessory leaflet into the outflow tract. Whereas a tumour mass or vegetation, theoretically, might produce a similar appearance,
### Table: Summary of reported cases of accessory mitral leaflet or tissue in normally connected hearts

<table>
<thead>
<tr>
<th>Author</th>
<th>Age (yr)/sex of case</th>
<th>Presentation</th>
<th>Gradient (mm Hg)</th>
<th>Surgical approach</th>
<th>Accessory tissue</th>
<th>Histology</th>
<th>Associated lesions</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maclean et al 2(^{1963})</td>
<td>29/M</td>
<td>Exercise intolerance, murmur since childhood</td>
<td>70</td>
<td>Aortotomy</td>
<td>2 parachute-like structures (5 mm and 15 mm) attached to anterior mitral leaflet</td>
<td>Situs inversus, right SVC</td>
<td></td>
<td>Good</td>
</tr>
<tr>
<td>Deal et al 3(^{1963}) Sellars et al 4(^{1964})</td>
<td>11/F</td>
<td>Seizures; heart murmur, &quot;Known to have subaortic stenosis&quot;</td>
<td>70</td>
<td>Aortotomy</td>
<td>5 balloon-like masses of spherical tissue</td>
<td>Connective tissue similar to normal mitral valve</td>
<td>Membranous subaortic stenosis. Supravalvar mitral ring; bicuspid aortic valve; single sinus origin of coronary</td>
<td>Died POD4. Accessory tissue not excised</td>
</tr>
<tr>
<td>Kelly et al 5(^{1972})</td>
<td>7/M</td>
<td>130</td>
<td>Ventriculotomy</td>
<td>On anterior mitral leaflet</td>
<td></td>
<td></td>
<td>Dextrocardia, subaortic ring</td>
<td>Good</td>
</tr>
<tr>
<td>Cooperberg et al 6(^{1976})</td>
<td>9/M</td>
<td>Asymptomatic</td>
<td>80</td>
<td>Aortotomy</td>
<td>Parachute-shaped, attached to anterior mitral valve ring; chordae attached to septum and papillary muscles</td>
<td></td>
<td></td>
<td>Good</td>
</tr>
<tr>
<td>Mathewson et al 7(^{1976})</td>
<td>3 d/F</td>
<td>Murmur; low cardiac output</td>
<td>62</td>
<td>Aortotomy</td>
<td>Pedunculated mass originating from midpoint of junction of mitral annulus and anterior leaflet</td>
<td></td>
<td></td>
<td>Died in operating theatre</td>
</tr>
<tr>
<td>Freedom et al 8(^{1977})</td>
<td>8 months/ not known</td>
<td>40</td>
<td>None (Postmortem specimen)</td>
<td>40</td>
<td>Nodular gelatinous mass of undifferentiated endocardial tissue from aortic leaflet of mitral valve and contiguous membranous septum</td>
<td>Ventricular septal defect, coarctation</td>
<td></td>
<td>Died</td>
</tr>
<tr>
<td>Kohda et al 9(^{1979})</td>
<td>2/F</td>
<td>Heart murmur at 14 months</td>
<td>50</td>
<td>Aortotomy</td>
<td>Accessory tissue attached to anterior mitral leaflet with 3 chordae</td>
<td>Fibrous tissue</td>
<td></td>
<td>Mild mitral regurgitation</td>
</tr>
<tr>
<td>Kuribayashi et al 10(^{1979})</td>
<td>3/M</td>
<td>Heart murmur at 1 month; syncope and chest pain on exertion</td>
<td>100</td>
<td>Aortotomy</td>
<td>White tissue with chordae attached to anterior leaflet of mitral valve and septum</td>
<td>Fibrous tissue</td>
<td></td>
<td>Good</td>
</tr>
<tr>
<td>Nanton et al 11(^{1979})</td>
<td>9/F</td>
<td>Murmur from 1st year</td>
<td>100</td>
<td>Aortotomy</td>
<td>Balloon-like, 2-2 cm x 1-3 cm. Attached in concave arc below left coronary leaflets and to LV wall. Chordae to anterolateral papillary muscle and lateral wall of LV</td>
<td>Fibrous tissue</td>
<td>Discrete subaortic ridge, muscular VSD, accessory tricuspid tissue, 2 coronary ostia in left sinus</td>
<td>Died in low cardiac output. Accessory tissue not excised</td>
</tr>
<tr>
<td>Gomes et al 8(^{1980})</td>
<td>14 months/ M</td>
<td>Murmur from birth, congestive heart failure</td>
<td>90</td>
<td>Aortotomy</td>
<td>1.2 cm x 4 cm nodular, flap-like fibrous tissue attached to base of anterior mitral leaflet</td>
<td>Valve-like structure</td>
<td>Partial AV septal defect; left SVC</td>
<td>Mitral insufficiency, otherwise satisfactory</td>
</tr>
<tr>
<td>Tanimoto et al 12(^{1981})</td>
<td>10 months/ F</td>
<td>Heart murmur from birth</td>
<td>26</td>
<td>Aortotomy</td>
<td>Chordae attached to mitral valve ring, aortic annulus, and LV free wall</td>
<td>Fibrous tissue</td>
<td>Supravalvar aortic stenosis; septal aneurysm</td>
<td>Good</td>
</tr>
<tr>
<td>Hatem et al 13(^{1981})</td>
<td>10/F</td>
<td>Asymptomatic heart murmur at 5 weeks</td>
<td>195</td>
<td>Aortotomy</td>
<td>Sheet-like mass under aortic valve with chordae to septum and papillary muscles</td>
<td>Fibromyoid tissue consistent with valve tissue</td>
<td>Bicuspid aortic valve</td>
<td>Residual 110 mm Hg gradient; reoperated via left atrium; similar mass removed from base of mitral leaflet</td>
</tr>
</tbody>
</table>
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</tr>
</thead>
<tbody>
<tr>
<td>Furuta et al 17</td>
<td>5/M</td>
<td>Murmur from birth. Tumour-like mass LVOTO on echo</td>
<td>100</td>
<td>Aortotomy then left atriotomy</td>
<td>25 cm × 30 mm accessory leaflet with multiple chordal attachments to anterior mitral leaflet and free wall of LV</td>
<td>Myxoid dysplasia. Normal endothelium of mitral cusp</td>
<td>Ring of fibrous tissue on atrial side of anterior mitral leaflet</td>
<td>Good</td>
</tr>
<tr>
<td>Hotta et al 21</td>
<td>14/M</td>
<td>Easy fatigability; heart murmur at 2½ years</td>
<td>50</td>
<td>Aortotomy then left atriotomy</td>
<td>Attached to anterior mitral leaflet and ventricular septum. Chordae connected to mitral valve chordae</td>
<td>Collagen fibre with myxoid stroma and endothelium</td>
<td></td>
<td>Mild aortic regurgitation</td>
</tr>
<tr>
<td>Zin'kovsky and Ignatow 19</td>
<td>6/F</td>
<td>Breathless on exertion; 100 temporary heart pains</td>
<td>100</td>
<td>Aortotomy</td>
<td>20 × 15 mm, arising from anterior leaflet of mitral valve</td>
<td>Like a mitral valve</td>
<td></td>
<td>Good</td>
</tr>
<tr>
<td>Losay et al 14*</td>
<td>9/M</td>
<td>Aortic stenosis and regurgitation</td>
<td>34</td>
<td>Aortotomy</td>
<td>1.5 × 2 cm “membrane”</td>
<td>Situs inversus, mitral regurgitation</td>
<td></td>
<td>Mild aortic regurgitation Good</td>
</tr>
<tr>
<td>Binet et al 22*</td>
<td>4/M</td>
<td>Aortic stenosis and regurgitation</td>
<td>112</td>
<td>Aortotomy</td>
<td></td>
<td>VSD</td>
<td></td>
<td>Good</td>
</tr>
<tr>
<td>Alboliras et al 16</td>
<td>15/M</td>
<td>Exertional dizziness and dyspnoea; mass on echo</td>
<td>100</td>
<td>Aortotomy</td>
<td>“Hood” of accessory tissue on anterior mitral valve leaflet</td>
<td>Discrete subaortic membrane</td>
<td></td>
<td>Residual 23 mm Hg gradient; accessory tissue not excised</td>
</tr>
<tr>
<td>Meldrum-Hanna et al 23</td>
<td>7/F</td>
<td>Asymptomatic murmur</td>
<td>80</td>
<td>Aortotomy</td>
<td>Attached around leftward part of anterior mitral leaflet and on to septum. Short chordae on free edge</td>
<td>Myxomatous dysplasia; abnormal mitral valve tissue</td>
<td></td>
<td>Reoperated 2 years later for discrete subaortic stenosis</td>
</tr>
<tr>
<td>Ascuitto et al 13</td>
<td>4/M</td>
<td>Heart murmur, mass on anterior mitral leaflet on echo</td>
<td>100</td>
<td>Aortotomy</td>
<td>Folds of leaflet-like fibrous tissue attached to base of anterior mitral leaflet; chordae to both papillary muscle</td>
<td>Trace of aortic regurgitation. Doppler gradient 25 mm Hg</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Three cases reported in two papers.

AV, atrioventricular; SVC, superior vena cava; VSD, ventricular septal defect; LVOTO, left ventricular outflow tract obstruction; POD, postoperative day.

such lesions more commonly originate from cardiac muscle or build up directly upon the low pressure side of a heart valve respectively. The degree of subaortic obstruction may be estimated from Doppler echocardiography, which may avoid the need for catheterisation. Indeed, recently operation has been recommended without invasive studies. Angiography can visualise a mass in the subaortic area but adds little to the diagnosis, and cardiac catheterisation probably is required only to investigate associated cardiac malformations.

The indication for operation was severe left ventricular outflow obstruction in eighteen patients and explorations of an intracardiac mass in two. One neonate who had repair of aortic coarctation subsequently died and was found to have accessory mitral valve tissue at necropsy. In another patient the accessory leaflet was discovered during reoperation for residual subaortic obstruction.17

All operations were performed on cardiopulmonary bypass, with a variety of approaches to the accessory leaflet. While aortotomy provides good access, identification of the accessory tissue may be difficult when it is accompanied by subaortic stenosis or has collapsed into an empty left ventricle. This is because the accessory mitral valve tissue always lies below any discrete fibrous obstruction. When the relation of the accessory leaflet to the mitral valve cannot be clearly seen through the aorta, or when additional malformations are present on the atrial side of the valve, a left atrial approach has been useful. Although a systemic ventriculotomy also provides good access, it is likely to compromise cardiac function and is probably unnecessary.

The gross appearances of the subaortic accessory mitral leaflet have been remarkably constant and are
not unlike those of accessory mitral valve tissue described in ventriculoarterial discordance with\textsuperscript{13,14} or without\textsuperscript{15,16} atrioventricular discordance. In all patients the accessory tissue was attached on the ventricular surface of the anterior mitral leaflet and in most it extended to the septal surface or free wall of the left ventricle. Both in relative and real terms, the accessory leaflet tends to be large. In our patient its diameter exceeded 25 mm, compared with an aortic annulus of 20 mm; and in all other operated cases the width was > 15 mm. Occasionally, a second mass of tissue originates from the membranous septum.\textsuperscript{8,11}

The extent of differentiation has been variable and may reflect haemodynamic forces applied to the accessory tissue within the left ventricular outflow tract. The most primitive, nodular, gelatinous mass of undifferentiated tissue was found in an eight month old infant;\textsuperscript{12} but most leaflets are parachute or sail shaped with a variable number of well defined chordae anchoring the ends to either or both papillary muscles of the mitral valve, to normal chordae, to the left ventricular free wall, or, as seen in our case, to an accessory papillary muscle. The hood-like structure thus produced presents a concave surface towards the left ventricle, so that in ventricular systole this pouch is distended with blood and is carried into the subaortic area. Histological examination of the accessory leaflet has generally shown fibrous tissue with myxoid dysplasia, analogous to dysplastic mitral valve tissue.\textsuperscript{7,13,16}

Because the leaflet is accessory tissue, its removal should in no way compromise the function of an otherwise normal mitral valve, and the results of operation usually have been satisfactory. In fifteen patients in whom the mass was removed completely as a primary procedure, there was one death. This was a three day old baby who had a left ventriculotomy after the tissue could not be identified through the aorta.\textsuperscript{4} Two patients had mild mitral insufficiency attributed in one to simultaneous repair of a partial atroventricular septal defect\textsuperscript{18,19}; and three patients (including the one reported here) had trivial aortic regurgitation.\textsuperscript{13,22} All survivors remained or became symptom free and none has shown signs of recurrent subaortic obstruction. Failure to remove the tissue, however, has been associated with considerable early mortality\textsuperscript{8-10} and a high number of reoperations,\textsuperscript{15,16} as has partial or incomplete relief of the subaortic obstruction.\textsuperscript{11,12} Since the development of real time echocardiography to demonstrate reliably the accessory leaflet before operation, there have been no operative deaths attributed to this lesion.

Little is known about the aetiology and course when an accessory mitral leaflet is present. It has been suggested that anomalies of the mitral valve and fibrous subaortic stenosis both result from abnormal development of endocardial cushion tissue.\textsuperscript{27} Yet, considerable evidence supports the view that discrete subaortic stenosis is an acquired malformation,\textsuperscript{28-30} while the anlaga of an accessory leaflet probably is present at birth, since the murmur is present early in life, even when there are no other cardiac lesions.\textsuperscript{37} It is likely that the degree of left ventricular outflow obstruction is progressive, because a gradient of 65 mm Hg at five years increased to 196 mm Hg by 10 years in the only patient who underwent serial left heart studies.\textsuperscript{11} Whether this results from enlargement of the accessory leaflet or from narrowing of the outflow tract by a discrete membrane is a question that echocardiography may answer in the future; certainly when the leaflet eventually prolapses through the aortic valve it produces symptoms of severe left ventricular outflow tract obstruction and aortic regurgitation, as seen in our patient.

More widespread availability of sector scanning and Doppler echocardiography may lead to the identification of an accessory mitral leaflet before the development of symptoms or important obstruction to left ventricular outflow. While elective, prophylactic removal of the tissue does not seem to be justified, careful excision of the accessory leaflet can be accomplished without morbidity or mortality. Accordingly, accessory tissue probably should be removed during open heart surgery for any associated cardiac defects to avoid residual and, possibly, progressive left ventricular outflow obstruction.\textsuperscript{16}

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Accessory mitral valve leaflet causing aortic regurgitation and left ventricular outflow tract obstruction


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