Mitral Valve Prolapse

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Mitral regurgitation is now much more commonly due to prolapse of the leaflets than to rheumatic disease. Its numerical importance had escaped recognition until recently because the volume of reflux is usually small, therefore the heart chambers are little increased in size, and also because the abnormality may not be recognised when the valve is cursorily inspected at postmortem. Our ability to follow the movements of the mitral leaflets with echoes has been the chief reason for the rapid growth in knowledge of this syndrome.

Previous generations of physicians recognised auscultatory signs at the mitral area which did not imply the presence of rheumatic mitral disease. Thus a systolic murmur that began late or a click in mid-systole were not associated with progressive disease and were considered innocent[1]. It is not possible to state with confidence the exact prevalence of imperfect mitral valve closure due to prolapsing leaflets because there are no undisputed criteria for its presence. However, a combination of the auscultatory and echocardiographic signs appears to be reliable. Using these criteria, Proccoci and co-workers[2] studied 1,169 young women with an average age of 32 years and found evidence of mitral prolapse in 6.3 per cent. Hickey and Wilckem[3] performed echocardiograms on 200 volunteers, half of whom were women, and found 4 cases in each sex (4 per cent). It is probable that a degree of mitral leaflet prolapse can be recognised in about 5 per cent of the population.

Pathogenesis

Functional competence of the mitral valve cannot be satisfactorily tested after death, so necropsy recognition of potential prolapse is difficult. Nevertheless, a number of structural and histological features have been recognised that allow a partial understanding of causative mechanisms.

Becker and de Wi[4] found imperfect chordal support of the areas of valve cusps that bulged unduly when the left ventricle was distended. Irregular arrangement or deficient branching of chordae left small or large portions of the leaflets poorly supported and would allow prolapse and reflux. Ballooning with increased cusp area can easily be recognised in the more severe cases, especially those submitted to surgical treatment. Such cusps may be thinned and white, or thin, transparent and of bluish tinge; their chordae are usually elongated and may be considerably thickened. Rupture of chordae is frequent and easily identified.

Mucinous degeneration may accompany elongation and perhaps thickening of the chordae (Fig. 1a, b).

Fig. 1(a). Normal mitral valve leaflet showing dense collagen.

Fig. 1(b). Mitral valve leaflet from mitral prolapse coming to valve replacement showing a few remaining fragments of collagen separated by non-staining mucopolysaccharide material.

Extensive replacement of the normal structure by mucoid material may be associated with rupture of chordae. As mucoid material is also present in normal valves, its detection does not imply abnormality. When excessive mucoid degeneration is present it might be the cause or
the result of the elongation and excessive tension. Thus imperfections of chordal support, redundant cusp tissue, mucoid degeneration and rupture of chordae are all implicated. The sequence of events and the time course of the changes remains to be elucidated.

Is the abnormality of the mitral valve an isolated lesion or part of a more generalised disturbance? Bon Tempo[5] and Solomon[6] and their co-workers reported a high incidence of pectus excavatum, straight back and scoliosis in their patients with signs of mitral leaflet prolapse. Davies et al.[7] examined 31 subjects with straight backs and 27 similarly affected relatives. Of the 58 subjects, 39 had clinical and/or echocardiographic evidence of mitral prolapse. The suspicion of Marfan's syndrome may arise in some. Davies et al.[8] found increased amounts of cross-linked procollagen and large amounts of single chain procollagen in floppy mitral leaflets, and postulated a block in the synthesis of mature collagen in these patients.

Physical Signs

In the great majority of patients recognised as having leaflet prolapse, auscultatory signs at the mitral area, with or without ectopic beats, are the only clinical evidence of the condition. However, echocardiographic evidence of mitral prolapse may be found in the absence of any auscultatory evidence, so the condition may be entirely silent.

Recognition of mitral prolapse almost always follows the hearing of a mitral systolic murmur or mitral systolic click. Characteristically, the mitral murmur does not begin with the first heart sound but follows a clear interval, the first sound being of normal intensity. Accentuation of the murmur in late systole is common. It is an easily heard murmur, usually of blowing quality although it may be honking, and, in some young people, occasionally audible to bystanders[9]. The presence of a silent interval after the first heart sound implies that no reflux occurs in early systole, but in those patients in whom there is echocardiographic evidence of immediate prolapse of the cusps the murmur may be pansystolic. This is likely to be the case whenever chordae have ruptured. Accentuation of the murmur in the left lateral position is usual but, even in those with gross mitral reflux, diastolic murmurs are uncommon.

Mid-systolic clicks may be the only auscultatory sign of prolapse or they may introduce the murmur. They are usually quite loud and easily distinguished from an aortic ejection click because they occur later in systole—usually near the mid-point. More than one click may be heard and recorded. Simultaneous echocardiograms and phonocardiograms show that the sound coincides with the onset of prolapse and leave little doubt that the click occurs as the prolapsing leaflet is abruptly tensed later than the rest of the valve.

Auscultatory signs may not always be present but can sometimes be brought out by making the patient stand or squat; these manoeuvres are recommended if mitral prolapse is suspected.

Displacement of the left border of the heart and an unduly prominent cardiac impulse implying a considerable increase in left ventricular stroke volume will be found only if reflux is gross. In these cases a systolic thrill may be felt.

Symptoms

Nearly all the subjects are symptomless. Breathlessness and heart failure will occur with massive mitral reflux only. Symptoms often follow medical discovery of the lesion or are engendered by anxiety, palpitations and chest pain being the most common.

Palpitations are usually due to the occurrence of ectopic beats—of ventricular origin in most instances. Sustained ventricular tachycardia and fast atrial arrhythmia also occur.

Chest pain may be troublesome and is always difficult to evaluate. In some patients the pain closely mimics ischaemic angina pectoris. More often the pain is maximal around the left breast, and is less clearly related to exertion and longer lasting than in angina pectoris. The mechanism is not understood.

Coronary artery disease may occasionally accompany prolapse but coronary arteriography usually shows normal vessels. Increased tension on papillary muscles is a possible cause of the pain. Not surprisingly, there is no specific medication, but the most important aspect of management is not to raise the suspicion of coronary disease if it is not present. The benign nature of the symptom should be stressed and the patient encouraged not to allow it to limit activity.

In some patients there are ECG abnormalities which increase the suspicion of coincidental coronary disease. It is reported that treadmill exercise tests may induce ECG changes but these should be considered false positives, as exercise thallium scintigraphy of the myocardium does not show perfusion defects[10]. Where any doubt exists it is wise to perform coronary arteriography with the expectation that coronary disease will be excluded and the patient reassured.

Investigations

The ECG and Chest Radiograph

No abnormality is to be expected in the ECG or chest X-ray in the presence of mild mitral reflux. Gross reflux will be accompanied by cardiomegaly and pulmonary venous congestion. The ECG will remain normal unless atrial fibrillation has occurred. However, a minority of patients (usually with chest pain) exhibit changes which suggest previous myocardial infarction and remain unaltered for many years. There is no agreement as to the cause. Ischaemic or inflammatory changes at the base of the papillary muscles or in the nearby inferior wall of the left ventricle have been postulated[11], as has a more generalised cardiomyopathy[12].

Echocardiography

Ultrasound cardiography best demonstrates prolapsing leaflets. M mode echocardiograms have been extensively
studied. The transducer should be placed perpendicular to the chest wall over the anterior cusps of the mitral valve. The least disputed abnormality is abrupt posterior sagging of the cusps in the second half of systole. Both cusps normally move forward together during systole but with prolapse there is abrupt backward movement beginning at the time of the systolic click (Fig. 2). If the prolapse is holosystolic the cusps sag backwards from the beginning of systole. Multiple echoes may be present. In diastole the prolapsing posterior cusp may move anteriorly rather than in its usual mode, mirror-imaging the anterior cusp. Gross redundancy of a cusp may give multiple echoes in diastole, as does a tumour[11]. Convincing echo evidence is the best currently available evidence of leaflet prolapse but in a proportion of patients it is impossible to record satisfactory echoes and other methods must then be used.

Angiocardiography

Mitral prolapse can be visualised by left ventricular angiography. The left anterior oblique position is preferred. Reflux of contrast into the left atrium is easily recognised but visualisation of the prolapse is not always beyond dispute. In this plane systolic bulging of the cusps is most obvious in the inferior and lateral portion. Prolapse in the middle section may be recognised by the ascending aorta and prolapse of the superior segment is less common. In some patients prolapse is best seen in the right anterior oblique view and if there is doubt both views should be taken.

Left ventricular angiography is only indicated in patients with severe reflux who are being evaluated for surgery, as part of the investigation of chest pain by coronary arteriography. As proof of the diagnosis of prolapse it is rather less reliable than echocardiography. It is superior, however, for visualising the reflux with certainty, and essential for confirming the volume of reflux. It also allows the state of left ventricular contractility to be assessed.

Complications

Heart Failure

Heart failure will only occur in patients with gross reflux. Even gross reflux with considerable cardiomegaly may be tolerated for ten years or more without obvious deterioration. In these circumstances the onset of atrial fibrillation frequently precipitates failure and mitral valve replacement is then called for. Rapid deterioration with heart failure may also be caused by rupture of the elongated chordae tendineae. A further cause of heart failure is infection of the valve. Infective endocarditis seems to be a greater risk in patients who have a mitral systolic murmur. This complication is uncommon and there may often be doubt as to whether antibiotic prophylaxis for dental treatment should be advised. In the absence of a systolic murmur it may be considered unnecessary.

Arrhythmia

Awareness of rhythm disturbance is a common mode of presentation. Isolated or frequent ventricular ectopic beats are usually responsible for the symptoms, but ventricular tachycardia and ventricular fibrillation may occur. The rhythm disturbance is usually benign and needs no medication, but a few patients have attacks of syncope or near syncope that call for careful evaluation. In these cases 24-hour ECG tapes are mandatory and may reveal repetitive ventricular tachycardia. A short PR interval may give evidence of an atrioventricular bypass. Sudden death is known to occur. Repetitive ventricular tachycardia or rapid supraventricular tachycardia may call for medication. The choice lies between beta-blockade, disopyramide, quinidine and mexiletene. In resistant arrhythmia amiodarone may be required.

Systemic embolism

Major systemic embolism is considered a rare complication. However, Barnett et al.[13] performed echocardiograms on 60 patients who had suffered cerebral ischaemic events before the age of 45. Mitral prolapse was thought to be present in 24 of the 60, as opposed to 5 of 60 controls. No other recognisable cause for cerebral ischaemia was found in 18 of the 24 and they postulated that fissuring around myxomatous valves might favour thrombus formation there.

Clinical Course

In the symptomless patient with a mitral click and late systolic murmur prognosis for life is excellent. Thus Harris and Leatham[14] followed 62 patients with late systolic murmurs for an average of 13 years. One patient...
died of infective endocarditis and another at the age of 75 from worsening mitral reflux. In 10 there was evidence of minor worsening of mitral reflux. Five patients developed infective endocarditis. One patient required mitral valve replacement for ruptured chordae. There were no sudden deaths. Patients with chest pain or abnormal ECGs were excluded and thus the study may over-emphasise the benign nature of the condition. Other studies have reported serious ventricular arrhythmias and an occasional fatality.

Worsening mitral reflux which requires surgery is relatively uncommon unless chordal rupture occurs or infection supervenes but, because of the frequency of the condition, mitral prolapse now figures prominently in surgical series. Replacement of the valve has usually been thought necessary but repair operations, which are clearly preferable if effective, are being reported[15].

Management

Patients with mitral leaflet prolapse are now most likely to be referred for assessment because a systolic murmur has been heard at routine examination or because ectopic beats have been detected by the patient or doctor. Characteristic physical signs and echocardiographic appearances suffice to confirm the diagnosis and with a heart of normal size it will usually be clear that reflux is slight. Life is then little affected by the lesion and the fact that about one in 25 normal people have the lesion justifies the view that the heart is really normal. There is a very considerable risk that troublesome anxiety symptoms will affect the life of the patient and there is much to be said for discharging the patient from further hospital attendance, the heart being considered normal. This is the author's practice. If the volume of mitral reflux appears to be more than slight, hospital follow-up is desirable, for if heart failure is allowed to develop the heart may become so dilated that surgery will not be able to achieve a favourable long-term result.

The dilemma is whether to recommend antibiotic prophylaxis for dental treatment. In highly anxious subjects with trivial signs it is not unreasonable to omit this measure, which is at present very poorly complied with. The risk of endocarditis has to be recognised.

In those with persistent ectopic activity there is a major therapeutic problem. In those who are symptomless the overall good prognosis makes it reasonable to ignore the complication. The continuous prescription of anti-arrhythmic tablets because the physician lacks the courage to accept rhythm disturbance on the patient's behalf is to be deprecated. The case is even worse if tablets are prescribed without evidence that they are being effective. In those whose arrhythmia causes troublesome symptoms, and above all in those with syncope or near syncope, 24-hour ECG taping is essential. Trials of beta blockade, disopyramide or mexiletene are appropriate, ECG monitoring of the effect being the yardstick of effectiveness. Invasive electrophysiological studies may be needed.

The problem of chest pain has already been considered.

Worsening mitral reflux should be assessed with care. Increasing left ventricular size recognised by X-ray, or preferably by echocardiography, is the best indicator of deterioration. Continuing heart failure, though it may be temporarily controlled by anti-failure treatment, soon leads to such dilatation of the left ventricle that poor function will persist even if the valvular fault is corrected[16].

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