Correspondence

British Heart Journal, 1975, 37, 438–439.

Congenital mitral regurgitation due to ‘posterior reinsertion’ of chordae tendineae

Sir:

Our experience of the ‘floppy’ mitral valve (redundant cusp syndrome) shows chordal rupture to be a not infrequent complication of this common condition. We were, therefore, surprised to see the article by Dr Bevilacqua with the above title (British Heart Journal, 1974, 36, 520) illustrating what to us is a typical example of the redundant posterior cusp with chordal rupture. The only unusual features are the number of chordae that have ruptured and their adhesion to the ventricular surface of the valve leaflet, though the latter feature is well recognized. The absence in the discussion of any reference to the redundant cusp syndrome suggests to us that the author is unaware of this condition, resulting in the misinterpretation of an acquired lesion as a congenital one. The clinical history also is typical of the redundant cusp syndrome.

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Sir:

I am surprised that Dr. Bevilacqua should conclude that the ‘posterior reinsertion’ of chordae tendineae illustrated in his paper is a congenital lesion. Surely a patient with such an extensive congenital lesion would develop symptoms of mitral insufficiency long before age 53? An alternative explanation is that the man ruptured the chordae when 53, the free ends recurved, as is common, came into contact with the ventricular surface of the posterior leaflet and, in the years before his death, fused with that surface. Dr. Bevilacqua should have excluded this possibility by indicating whether or not chordal stumps were found in the papillary muscle – such stumps may resorb leaving only greyish elevations. Furthermore, he should detail the morphological findings at the junction of the reinserted chordae and leaflet. One might expect a difference between a congenital and an acquired lesion. Until he provides answers to these points his conclusion must remain questionable.

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These two letters were shown to Dr. G. Bevilacqua who replies as follows:

Sir:

I thank Drs. Davies and Pomerance and Professor Silver for giving me this opportunity of clarifying certain details of my paper.

First, I should like to summarize the most important pathological data:

Macroscopical examination: the abnormal chordae are inserted by their ends only; there is no sign of fusion; inflammatory changes are absent; the posterior leaflet has a hood-like appearance and is thickened; the papillary muscles are normal in number and situation and their tips show normal chordal attachments; no chordal stumps are present.

Microscopical examination: the thickness of the valve leaflet is due to superimposition of collagen tissue; the collagen component of the valve shows some degeneration particularly near the tip, but even there the fibrils are mostly regularly arranged; mucoid degeneration is absent; the site of reinsertion of the chordae is normal, without intervention of altered collagen tissue or inflammatory infiltrates, and collagen fibres are seen to pass regularly from the chordae to the leaflet.

These findings suggest two diagnostic hypotheses: a) congenital abnormality of the chordae, or b) rupture of the chordae with their successive fusion on the ventricular surface of the leaflet. I support the former hypothesis since there are no data which may reasonably support the latter.

Rupture of the chordae may result from a) endocarditis; b) collagen degeneration such as the mucoid degeneration of the floppy valve syndrome; c) trauma; d) no apparent cause.
In the present case, there is no history of trauma, and the macroscopical and microscopical findings exclude the diagnosis of either endocarditis or degenerative pathology of the mitral valve.

In respect to the presence of a large hood-like leaflet, this is not pathognomonic of the floppy valve, since it has often been observed without mucoid degeneration; it has also been described to result from blood regurgitation in cases of rupture of the chordae tendineae (Osmundson, Callahan, and Edwards, 1961; Edwards, 1971).

The morphology of the papillary muscles and of the site of chordal reinsertion seems to exclude the possibility of ruptured normal chordae.

It would not be exceptional to find morphological resemblance between the appearances described and that of ruptured chordae (hood-like leaflet and jet lesions on the interatrial septum) in view of the identical haemodynamics of regurgitation present in both conditions.

The long survival is not strange if we consider that patients suffering from rheumatic fever causing valvular involvement die on the average 40 years after the beginning of such disease. Furthermore, some morphological findings may explain such behaviour: the presence of normal chordae at the extreme ends of the leaflet and the reinserted chordae should have prevented the overturning into the atrium of the cusp or of its free margin only; moreover, the size of the hood-like leaflet (height, 2 cm; width, 5.5 cm) might have partly compensated for the valve regurgitation.

In my opinion the fusion of ruptured chordae is an improbable event considering that cardiac activity during systole moves them continuously and can even make them vibrate into the atrium.

In 1953, Prior described a similar case with some differences in the arrangement of the chordae, considering it a congenital anomaly. This pathogenetic conclusion by the author has been quoted without comment by other authors such as Keith (1962) and Hudson (1965).

In conclusion, the absence of signs of rupture of chordae and the lack of consistent changes in the valvular tissue and at the site of reinsertion excluded the possibility of a refusion of ruptured chordae and suggested the diagnosis of a congenital anomaly.

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References