Case report

Aortic regurgitation in rheumatoid arthritis necessitating aortic valve replacement

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Abstract

Four cases of patients with aortic incompetence secondary to rheumatoid arthritis are presented. All survived aortic surgery (two having bioprosthetic aortic valve replacement and two a homograft aortic root replacement). A review of the pathophysiology and pathology of this rare cause of aortic regurgitation is presented. A rationale for the choice of aortic valve prosthesis is discussed. © 1999 Elsevier Science B.V. All rights reserved.

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1. Introduction

Rheumatoid arthritis commonly affects the heart with haemodynamically insignificant valve lesions often found in asymptomatic rheumatoid sufferers [1]. In comparison, severe aortic incompetence secondary to rheumatoid arthritis needing valve replacement is very rare.

Here we report four cases of rheumatoid associated aortic incompetence of rapid progression requiring valve replacement.

2. Case reports

Four patients underwent aortic valve surgery for rheumatoid aortitis. The patients (one male, three female) ranging in age from 37 to 68 all had a long history of severe debilitating seropositive rheumatoid arthritis. One patient (a 44-year-old woman) had undergone previous surgical treatment for cervical myelopathy (complicated by mild quadriplegia). The mode of presentation was acute left ventricular failure in all cases. In two of the patients (37- and 44-year-old women) there was an initial suspicion of bacterial endocarditis but blood cultures were negative in both. In two cases (a 68-year-old man and a 46-year-old woman) moderate aortic incompetence had been diagnosed 3 and 4 years earlier, respectively. In the other two cases (those initially suspected of having endocarditis) the incompetence had been noted only 3 months earlier in one and in the last the regurgitation was noted for the first time at presentation in left ventricular failure. Investigations showed diminished left ventricular contractility with severe aortic incompetence in all. All patients were on steroidal and non-steroidal medication prior to surgery and one received six pulses of Cyclophosphamide pre-operatively in an attempt to reduce any active aortitis.

At operation grossly oedematous, adherent pericardium was found in two patients. The aortic valve was found to be tricuspid with thickened cusps and a normal diameter annulus in all cases. In one case the actual cusps were small and retracted with a grossly thickened fibrous annulus. In another, with an initial diagnosis of suspected endocarditis, the right coronary leaflet was noted to be grossly sclerosed with retraction towards the cusp line. On excision of the leaflets (in this case) there was no sign of vegetative material but on sectioning small amounts of pus were found (microscopy demonstrated pus cells but no organisms). His-
Aortic incompetence died in the early post-operative phase in a patient with mechanical aortic valve replacement after acute post-operative complications. One patient (a 37-year-old woman who had a bioprosthesis inserted) was lost to follow-up. One patient (the eldest) died 6 months post-operatively of pneumonia. The remaining patients (who had homografts inserted) are well 2 and 3 years later, respectively.

3. Discussion

Rheumatoid arthritis has many cardiovascular manifestations, ranging from a severe vasculitis to discrete valvular heart lesions. Histopathologically there are two varieties of valvular lesions; the more common granulomatous involvement of the valve leaflets and ring and the rarer non-granulomatous inflammation of the valve with thickening and fibrosis of the leaflets [2,3]. Pathophysiologically aortic incompetence can be considered to be caused by a number of different mechanisms [4]: aortitis with dilatation of the aortic ring, a destructive process of the valve leaflets themselves or finally prolapse of actively inflamed degenerate cusps.

Aortic incompetence secondary to connective tissue diseases including rheumatoid arthritis has a relatively accelerated course rapidly leading to severe left ventricular failure. Our patients are typical of this accelerated natural history. In a previous group of young patients with seropositive arthritis and aortic incompetence, three of the four patients reported rapidly deteriorated within 4 years of diagnosis [5]. Other cases reported have had correspondingly rapid rates of deterioration with one case deteriorating from normality (asymptomatic with a normal aortic valve) to severe left ventricular failure and aortic regurgitation in 10 days [6].

Pre-operatively immunosuppressive regimes (pulse Cyclophosphamide [7]) are indicated to treat any active aortitis (which may even cause the aortic incompetence to regress avoiding surgery) [8] and may well make the procedure technically easier by reducing aortic root inflammation. The oedematous and friable nature of the tissues made the procedures technically demanding in all our cases. In our eldest patient Teflon® buttressing of the aortotomy had to be carried out to aid haemostatic closure, as friability of the autologous pericardium prevented its use as a buttress. Considering the severe immunosuppression and the multi-systemic nature of the disease found in these patients few post-operative complications have been reported. One patient with mechanical aortic valve replacement after acute aortic incompetence died in the early post-operative phase [8], an unexplained cerebrovascular accident occurred after mechanical aortic valve replacement [9] and a post-operative left ventricular pseudo-aneurysm [10] have been reported. Interestingly, coronary vasculitis (as complicated our first case) and severe wound problems have not been described.

The logic for different valve replacement strategies has not been discussed in the literature to date. A review of the literature would suggest that of the 28 previously described procedures 19 have had a mechanical prosthesis implanted. Considering the youth of many of the patients the use of biological valve replacements may be complicated by early failure with implicit high risk re-replacement. Two reported pulmonary autografts in adolescents with juvenile chronic arthritis have failed with associated mortality within 6 months of surgery. In comparison we feel that the necessity for long-term anticoagulation implicit in the use of prosthetic valves is relatively contra-indicated in such patients who often are on polypharmaceutical regimes, including steroids and non-steroidal anti-inflammatory agents that may predispose to peptic ulceration. Further long-term follow-up is necessary to assess the long-term results of xenograft or homograft implantation in this group of patients but we feel that decisions on aortic valve replacements in such patients should consider both avoidance of anticoagulation as much as prosthetic durability.

References