An Autopsy Study of Rheumatic Heart Disease

Part II. Associated Findings

Veena MALHOTRA, M.D., P. C. BEOHAR, M.Sc., Ph.D.,
R. GONDAL, M.D., U. A. KAUL, M.D., D.M.,
and S. K. KHANNA, M.Ch.

SUMMARY

Histo-morphological changes of the coronary vessels in cases of rheumatic heart disease (RHD) have been studied in 60 cases.

Involvement of intramyocardial branches of coronary vessels in the form of active rheumatic vasculitis or inactive lesions characterized by medial hypertrophy and replacement fibrosis was seen in 15 of 60 cases. These lesions may affect myocardial function. Atherosclerosis of the pulmonary trunk and its branches was frequently seen in these hearts, indicating that this may be an important index of pulmonary hypertension. An unusual association of bicuspid aortic stenosis and RHD was seen in one case. Another case showed acute myocardial infarction due to coronary embolism from bacterial vegetation of bacterial endocarditis.

Additional Indexing Words:

RHD Associated lesions Coronary vessel involvement Pulmonary atherosclerosis

CORONARY vessel involvement in rheumatic heart disease (RHD) is well known. The involvement can be due to the presence of paravascular bodies, which while healing, may extend into the adventitia of the vessel, or due to rheumatic vasculitis involving the vessel wall with formation of Aschoff bodies in the wall of the vessel. Myocardial changes seen in the form of myovacuolation and replacement fibrosis in the myocardium can be due to coronary vascular involvement or due to a hemodynamic effect. In this communication, the nature and the extent of coronary vessel involvement in RHD as seen on autopsy is described. The finding of rare abnormalities such

From the Departments of Pathology, Cardiology, and Cardiac Surgery, G. B. Pant Hospital, New Delhi, India.

Address for reprint: Dr. Veena Malhotra, Associate Professor in Pathology, G. B. Pant Hospital, New Delhi-110002, India.

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as pulmonary atherosclerosis, bacterial endocarditis and congenital heart disease during an autopsy study of RHD is also presented.

MATERIALS AND METHODS

Autopsy material from 60 cases of RHD was reviewed to demonstrate the incidence of involvement of coronary vessels and other unusual associated lesions. Special stains including Van Gieson's, Verhoeff's elastic stain and Masson's trichrome were used.

RESULTS

Of the 60 cases of RHD, 15 showed involvement of myocardial vessels on autopsy.

Involvement of the coronary sinus: Two cases showed presence of Aschoff cells in the wall of the coronary sinus. These cells had an irregular shape and a prominent nuclear central nucleolus. The cells were arranged all along the wall in a circumferential manner (Fig. 1). At places, these cells had a smudgy appearance. No localized nodules were seen.

Involvement of the coronary arteries and their branches: Major coronary arteries did not show involvement. Intramyocardial arteries to the level of the arterioles were involved. Involvement was in the form of active vasculitis or inactive lesions characterized by medial hypertrophy and replacement fibrosis. Active vasculitis was seen in the hearts where active or healing rheumatic lesions were present, whereas inactive lesions were present in hearts with healed rheumatic lesions.

Active vasculitis in medium sized vessels and arterioles was seen in 5 cases. There was evidence of narrowing of the lumen with the media showing irregular hypertrophy, replacement fibrosis and inflammatory cells infiltrating into it (Fig. 2). The adventitia showed inflammation and fibrosis. A few medium sized vessels and arterioles showed intimal injury with evidence of fibrinous material being incorporated into the intima (Fig. 3). Active vasculitis was accompanied by the presence of Aschoff bodies in the heart.

Medial hypertrophy with replacement fibrosis of the vessel in the left ventricular free wall was seen in 1 case (Fig. 4). No active inflammation was present in the wall.

Vessels of the papillary muscles of the left ventricle showed medial hypertrophy and replacement fibrosis. Smooth muscle cells showed edema and vacuolar degeneration. Adventitial and periadventitial fibrosis were seen as well (Fig. 5). Involvement of the vessels of the papillary muscles was much

Fig. 1. The figure shows coronary sinus with smudge cells and inflammatory cells in its wall. H & E $\times 250$.

Fig. 2. This shows intramyocardial vessel with media showing irregular hypertrophy, replacement fibrosis and inflammation. H & E $\times 250$.

more frequent and was seen in 12 cases.

Association with other abnormalities:

1) Mural thrombi: Mural thrombi were seen in the left atrium, left atrial appendage, left ventricle and the pulmonary trunk (Table I).

2) Pulmonary atherosclerosis: 25% of the cases showed evidence of

Fig. 3. This shows a vessel with fibrinous material being incorporated into intima. A healing Aschoff lesion is seen as well. H & E $\times 250$.

Fig. 4. This shows vessels with medial hypertrophy and replacement fibrosis. H & E $\times 250$.

atherosclerosis of varying degree in the pulmonary trunk and its branches. In a few cases atherosclerotic plaques showed a large number of cholesterol clefts (Fig. 6).

3) Bacterial and fungal endocarditis: Three patients showed evidence of valvular endocarditis. Two patients had bacterial endocarditis and 1 had

Fig. 5. Papillary muscle vessel showing adventitial fibrosis, medial thickening, interstitial edema in the media and mild replacement fibrosis. H & E $\times 250$.

Table I. Association with Thrombi

Atrial appendages	10
Left ventricle	2
Left atrium	3
Pulmonary trunk	3

Fig. 6. This shows right pulmonary artery showing an atherosclerotic plaque with prominent cholesterol clefts. H & E $\times 160$.

postoperative fungal endocarditis. The fungal infection in this case had disseminated to other organs. One of the patients with bacterial endocarditis had a coronary embolism resulting in an acute myocardial infarction. In both patients with bacterial endocarditis, the aortic valve showed vegetations.

4) Association with congenital heart disease: Congenital heart disease was seen in association with RHD in 4 patients. Three of them showed ASD and 1 had congenital bicuspid aortic stenosis.

DISCUSSION

Coronary sinus involvement seen in 2 of our cases was in the form of the presence of Aschoff cells in the wall of the sinus. Involvement of the coronary sinus has been described previously,¹⁾ as has rheumatic phlebitis of the coronary veins.²⁾

Lesions seen in the intramyocardial coronary vessels were in the form of active vasculitis or inactive lesions characterized by medial hypertrophy and replacement fibrosis. Active vasculitis involved all coats of the vessel wall with evidence of intimal injury, medial hypertrophy, adventitial and periadventitial fibrosis and inflammation in the adventitia and media. These lesions were considered to be due to vasculitis in view of the presence of inflammatory cells in the wall. The presence of healing Aschoff lesions elsewhere in these hearts supports the inflammatory nature of these lesions. Lesions in the intramyocardial branches of the coronary vessels in the form of medial hypertrophy, glassy medial hypertrophy, net like fibrinous thrombi affecting small arteries and arteritis have been reported previously in rheumatic heart disease.^{3),4)} Formation of Aschoff bodies in the medial coat of these vessels has been described by Murphy¹⁾ who considered the smooth muscle cell of the vessel wall as a possible cell of origin for Aschoff cells. Befler et al⁵⁾ have shown involvement of coronary vessels in 13 out of 26 cases of RHD on cardiac catheterization and cineventriculography. Five patients showed severe involvement and 8 showed minor arterial changes.

Lesions seen in the intramyocardial vessels of the left ventricular free wall and papillary muscle in the form of medial hypertrophy and replacement fibrosis can be due to healed vasculitis or due to stress on the vessel wall in response to an anoxic stimulus, or in demand to work hypertrophy stimulus to maintain supply to the failing heart. Although both of the above explanations for these morphological changes can be considered, the presence of replacement fibrosis in the wall of vessels favors a healed vasculitis. Vessels of the papillary muscles were much more frequently involved than those of the free wall. Lesions in the intramyocardial vessels can be considered as one of

the factors responsible for poor myocardial function in these patients. Well defined fibrotic scars in the left ventricular myocardium were seen in 3 cases where intramyocardial vessels showed involvement, but a complete correlation between vessel involvement and changes in the myocardium could not be observed. The changes in the myocardium are partly due to hemodynamic effects. A positive correlation between changes in the intramyocardial coronary artery branches in rheumatic mitral valvular disease and left ventricular function has been shown by Subramanyam et al.⁶⁾

Atherosclerosis of the pulmonary trunk and its branches was seen in 25% of the cases. Pulmonary atherosclerosis was considered to be due to prolonged severe pulmonary hypertension. Pulmonary atherosclerosis in RHD has been described earlier.⁷⁾ In our material, significance of pulmonary atherosclerosis could not be established, as its correlation with pulmonary pressures could not be studied due to the nonavailability of angiocardiographic data in the few cases with marked pulmonary atherosclerosis.

An association with bacterial endocarditis was seen in 4% of the cases. An incidence of 5.7% has been reported earlier.⁸⁾ One case showed evidence of myocardial infarction which occurred as a result of a coronary embolism from bacterial vegetation. This rare complication has been reported previously as well.⁹⁾ Coexistent congenital heart disease was present in 6.6% of cases, which is comparable to earlier reports.^{8),10)} Association with ASD is well known and association with bicuspid aortic stenosis has been described earlier by McReynolds et al¹¹⁾ and Borman et al.¹²⁾

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