CASE REPORTS

ENDOCARDIAL FIBRO-ELASTOSIS IN A YOUNG ADULT

BY

RISTEÁRD MULCAHY AND ROBERT P. TOWERS

From St. Vincent's Hospital, Dublin

Endocardial fibro-elastosis has been frequently reported in recent years, and the subject has been reviewed by Gray (1951), Davies and Ball (1955) and Lynch and Watt (1957), among others. The largest series of cases have been described in the native peoples of East Africa (Bedford and Konstam, 1946; O'Brien, 1954; Davies and Ball, 1955), although the white inhabitants of Africa are apparently not immune. Relatively few cases have been reported in the indigenous populations of the British Isles and America.

The purpose of this case-report is to describe the clinical and pathological findings in a girl of 18 years. Unusual features were the localization of the lesion to the right side of the heart and obvious retardation of sexual development.

Case History

Miss M., aged 18 years, was first admitted to St. Vincent's Hospital in May 1956, with gross cardiac enlargement and congestive heart failure. Despite appropriate treatment, the heart failure showed little improvement. From this time until her death in January 1957, she had a raised venous pressure with systolic expansion, widespread oedema, ascites, and hepatic enlargement. Extensive investigations failed to reveal the cause of the heart lesion. The electrocardiogram showed controlled auricular fibrillation and low voltage in all leads. There was no eosinophilia.

There was a history of jaundice as a young child. Otherwise the patient had been well until March 1955 when she had an appendicectomy. It was noted that her abdomen became distended after operation, and her symptoms of fatigue and dyspnoea on exertion dated from this time. Previous to this, her effort tolerance had been normal. There was also a history of primary amenorrhoea, and on examination, there was retarded breast development and absence of axillary and pubic hair.

Necropsy Findings. There was severe generalized oedema. Internally, the right pleural cavity was almost filled by opalescent, straw-coloured fluid, with a smaller amount in the left pleural cavity. The right lung was mainly collapsed, with apical emphysema; the left lung showed congestive changes with early bronchopneumonia.

The pericardium contained a small excess of fluid. The heart and thoracic aorta weighed 360 g. after fixation. The right atrium was greatly dilated, with a large mass of adherent ante-mortem thrombus in the auricular appendage. The right ventricle was dilated. The endocardium of the right side of the heart was thickened, white, and opaque, with occasional patches of greater depth; that on the left side was more or less normal. The valves appeared normal and no congenital malformations were noted. The coronary veins and coronary sinus were dilated. The aorta was rather small; otherwise the great vessels and coronary arteries were normal.

There was a large quantity of straw-coloured fluid in the peritoneal cavity. The liver was about normal in size, but showed marked "nutmeg" change and a fine cirrhosis. The other abdominal viscera were normal except for congestion of the kidney and the brain. The ovaries were normal in size.

All other organs were examined histologically except the uterus and ovaries, which were unfortunately mislaid.

Heart. Sections were stained by hæmatoxylin and eosin, by the Verhoeff-van Gieson method for elastic tissue, and by Masson's trichrome method. They showed much endocardial thickening due to fibro-elastosis in the right-sided chambers, and to a much lesser extent in the left atrium and ventricle. This process was beginning to extend into the muscle of the right ventricular wall, which contained numerous dilated vessels (see Fig. 1 and 2). Several branches of the coronary arteries showed a little intimal thickening, apparently

resembling that of the endocardium. Some of the sub-epicardial fat cells in the region of the base of the heart showed the peculiar multilocular appearances of so-called brown fat. The significance, if any, of this finding is not clear.

FIG. 1.—A low-power view, showing the grossly thickened endocardium of the right ventricle, with extension into the underlying muscle. Many dilated vessels can be seen. Masson's trichrome. $\times 35$.

FIG. 2.—Showing the numerous elastic fibres in the thickened endocardium of the right ventricle. Verhoeff-van Gieson. $\times 100$.

Liver. There was a rather unusual type of cirrhosis present, with bands of fibrous tissue running through the liver in a haphazard manner, unlike the typical centrilobular fibrosis of cardiac cirrhosis. This was undoubtedly the effect of long-standing, severe congestive failure (Sherlock, 1951).

Comment

This case appears to fall into the adult group described by Thomas *et al.* (1954). As in the other reported cases, the origin of this lesion is obscure and, until a satisfactory explanation is forthcoming, all examples should be recorded. This particular case was a sporadic one in a relatively enclosed population in rural Ireland, the age of the patient and the lack of sexual development being unusual features.

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