Constrictive endocarditis

Report of a case with successful surgery

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SUMMARY An 18-year-old white youth presented with severe right heart failure and was found to have an obliterated and funnel-shaped right ventricle, massive tricuspid regurgitation, and mitral regurgitation. The haemodynamic findings were those of cardiomyopathy with obliteration on the right side. He underwent surgery consisting of decortication and peeling off of a thickened right ventricular endocardium, tricuspid valve replacement, and mitral valve repair. After surgery there was pronounced haemodynamic as well as clinical improvement. The pathological picture was that of constrictive endocarditis. We recommend this method of treatment for patients with oblitative cardiomyopathy on either side of the heart.

A variety of cardiac disorders characterised by thickening of the endocardium and involvement of myocardium have been described. Despite the relatively unique modes for their clinical presentation, because of minor differences in pathology they have been classified differently, namely: endocardial fibroelastosis (Van Buchem *et al.*, 1959; Moller *et al.*, 1964; Sellers *et al.*, 1964), endomyocardial fibrosis (Shillingford and Somers, 1961; Moller *et al.*, 1964; Somers *et al.*, 1968a, b; Falase *et al.*, 1976), Loeffler's eosinophilic endocarditis (Brockington and Olsen, 1973; Bell *et al.*, 1976).

The purpose of this paper is to describe a patient with clinical and pathological findings characteristic of constrictive endocarditis, who underwent successful surgery of the lesion. The pre- and postoperative haemodynamic data are presented in detail.

Case report

An 18-year-old white youth was admitted in May 1976 with palpitation, dyspnoea on exertion, chest pain, dizziness, oedema, and swelling of the abdomen.

He had been in fairly good health until 6 years before admission when he developed palpitation and dyspnoea on exertion. During the ensuing 6 years he became progressively incapacitated, developing chest pain, oedema, and ascites. The past and family history were essentially unremarkable, he

gave no history of rheumatic fever and no other member of the family had a similar illness.

On physical examination he looked ill and pale, but was well developed, weighing 66 kg; heart rate was 76 to 84/min, and irregular, respiration 24/min, and blood pressure was 115/70 mmHg. The peripheral pulses were of good quality. The jugular veins were distended in the sitting position with equal a and v waves. The apical impulse was hyperactive and first and second heart sounds seemed normal. A grade 3-4/6 systolic murmur was heard at the apex with radiation to the left sternal border. The lungs were clear. The liver was palpable 4 to 5 cm below the right costal margin. Ascites was present. All laboratory investigations were within normal limits and no abnormal eosinophilia was noted. The patient underwent a right and left heart catheterisation, with findings characteristic of restrictive endocardial fibrosis (Fig. 1a).

Cineangiography showed that the left ventricle was large and that it contracted poorly; there was severe mitral regurgitation. The right ventricle was almost obliterated, being small and funnel-shaped, with no trabeculation and poor contraction (Fig. 2a); there was massive tricuspid regurgitation. The right atrium was dilated. Despite vigorous medical treatment the clinical course deteriorated, and he agreed to undergo surgery. During operation a huge right atrium and a small right ventricle were seen. The right ventricular endocardium was

Fig. 1 (a) Right atrial, right ventricular, and pulmonary arterial pressures with intracardiac electrocardiogram before operation showing diastolic plateau and early diastolic dip. Note similar pressure contours in all the chambers and vessels. (Pressure standards 30 mmHg.) (b) Normal right atrial, right ventricular, and pulmonary arterial pressures after operation.

extremely thickened and glossy and there were firm adhesions between the right ventricular endocardium, the myocardial fibrotic tissue, and the

tricuspid valve. The thickened right ventricular endocardium was extensively peeled off together with the deformed tricuspid leaflets. A number 29

Fig. 2 (a) Frame of cineangiogram obtained from the right ventricle showing a smooth-walled small ventricular chamber with massive tricuspid regurgitation. (b) Frame of cineangiogram obtained from the right ventricle. Note enlarged right ventricular cavity and absence of tricuspid regurgitation.

Carpentier valve was inserted in the tricuspid position. The mitral valve was repaired.

The postoperative course was uneventful and the patient had no audible murmur. The congestive heart failure receded and the patient's general condition improved significantly. Chest x-ray film showed a conspicuous reduction in heart size. Repeat cardiac catheterisation 6 weeks after operation showed that intracardiac pressures had returned to normal (Fig. 1b), and a right ventricular cineangiogram showed a normal sized chamber with good contraction and no tricuspid regurgitation (Fig. 2b). A left ventricular cineangiogram showed no mitral regurgitation. The patient was maintained on digitalis and at present is leading a fairly normal life.

The endocardial specimen removed during operation consisted of several thick membranous rigid sheets of endocardium with thickness varying from 1.5 to 3 mm. The inner surface of the endocardium was glistening and no mural thrombi were noted.

The parietal surface of the endocardium was partially covered by ragged pieces of myocardial tissue. The dense white endocardial fibrous tissue anchored the papillary muscle and enmeshed the roots of the chordae tendineae. Microscopical

sections (Fig. 3) of various parts of the specimen showed distinct thickening of the endocardium with collagen. With Verhoeff van Gieson elastic stain the appearances were unlike those in endocardial fibroelastosis and sections showed irregular masses of broken elastic fibres instead of proliferating tissue. The thickened areas showed destruction of the original endocardium and adjacent superficial myocardium with replacement by vascular fibrous tissue. There were foci of sparse perivascular lymphocytic cell infiltration in the deeper layer of the endocardium.

The pathological diagnosis was constrictive endocardial sclerosis.

Comment

The clinical picture of patients with endocardial fibrosis is similar to that in endomyocardial fibrosis or Loeffler's endocarditis (Shillingford and Somers, 1961; Somers *et al.*, 1968a, b; Brockington and Olsen, 1973; Bell *et al.*, 1976) and classification is principally based on the pathological findings.

Our patient presented with clinical findings of right heart failure and the haemodynamic studies showed the characteristic findings described for endocardial fibrosis of the right ventricle with

Fig. 3 Microscopical section of the resected tissue, showing extension of fibrous tissue fascicles from the thickened endocardial layer in between the subendocardial muscle bundles. (Masson's trichrome. $\times 100$.)

tricuspid regurgitation (Shillingford and Somers, 1961; Somers *et al.*, 1968a, b). The right ventricular angiogram which demonstrated a funnel-shaped and constricted right ventricle led us to diagnose endomyocardial fibrosis as described previously (Somers *et al.*, 1968). The reduced compliance of the right ventricle in our patient resulted in high end-diastolic pressure in this chamber and an increase in right atrial pressure. In the absence of left ventricular endocardial fibrosis, the pulmonary arterial pressure is reported to be normal (Somers *et al.*, 1968).

Although in our case the left ventricular involvement was not as extensive as the right ventricular lesion, poor contraction of the left ventricle was present. Nevertheless during operation no fibrosis was found in the left ventricular endocardium. The overall haemodynamic picture in this case is compatible with generalised myocardial disease with predominant right ventricular involvement.

The pathological findings were interpreted as constrictive endocarditis. It seems that the above entity is a variation of endomyocardial fibrosis. The latter lesion is often described in Africans (Shillingford and Somers, 1961; Somers *et al.*, 1968 a, b), but to our knowledge no cases have been observed in this area. Though Loeffler's endocarditis presents with similar findings (Friedberg, 1966; Brockington and Olsen, 1973; Bell *et al.*, 1976), the reported cases have pronounced eosinophilia during phases of the disease, which was not seen in our case. The pathological findings have differed from those classically described for endocardial fibroelastosis (Van Buchem *et al.*, 1959; Moller *et al.*, 1964; Sellers *et al.*, 1964). In a recent review of endomyocardial fibrosis it was described as a distinct entity different from Loeffler's endocarditis (Patel *et al.*, 1977). Use of the term restrictive cardiomyopathy for the spectrum of the disease has been recently recommended (Chew *et al.*, 1977).

The aetiology of the disease in our case as in most of the published cases remains unknown.

An important point of emphasis in this case is the successful surgery. In an extensive review on the treatment of cardiomyopathies (Goodwin, 1973), the results in obliterative cardiomyopathies were cited as unsatisfactory. Our success with decortication and peeling of the right ventricular endocardium as described by Dubost *et al.* (1976) and Cachera *et al.* (1976) sheds hope on the management of such patients. Dubost's recent article (Dubost *et al.*, 1976) reporting the results of this operation on 5 cases is further support that this method can successfully palliate patients. Clinically our case showed dramatic improvement with resolution of hepato-

megaly and ascites. The heart size was reduced and the patient's functional classification improved significantly.

Haemodynamic studies after operation showed distinct improvement and nearly normal intracardiac pressures; angiograms have shown reduction of obliteration and improved ventricular function.

Although the long-term results and future course of the patient are unknown, in view of the extremely poor results of pure medical management in such patients we recommend the above-mentioned method of operation for all patients whose course is one of deterioration with right or left sided obliterative cardiomyopathy. These efforts may perhaps give hope in the management of a potentially incurable heart disease.

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