Severe subpulmonary obstruction caused by an aneurysmal tissue tag complicating an infundibular perimembranous ventricular septal defect

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SUMMARY A case of severe right ventricular obstruction caused by a huge aneurysmal tissue tag is described associated with a perimembranous ventricular septal defect with an infundibular extension.

Angiograms and surgical inspection clearly showed that the ventricular septal defect extended into the outlet septum. The distinctive feature of this case was the severity of the gradient related to the peculiar position of the aneurysmal tissue in the outlet portion of the ventricle.

We presume that such an aneurysm, however small it is, developing in association with a ventricular septal defect with an infundibular extension may be a potential cause of severe obstruction and should therefore be carefully followed in the ensuing years.

Aneurysms of the membranous septum, consisting of a fibrous tissue tag, are commonly found in patients with a ventricular septal defect and their appearance usually precedes a diminution or even spontaneous closure of the ventricular septal defect.^{1 2} Right ventricular outflow tract obstruction, usually not very severe, is one of the rare complications described in association with an aneurysm of the membranous septum.

The purpose of this communication is to present a case of a huge fibrous tissue tag associated with a perimembranous ventricular septal defect extending into the infundibular region and causing severe right ventricular outflow tract obstruction, which was observed 26 years after the first cardiac catheterisation.

Case report

A 36-year-old man known to have a heart murmur from the age of 6 months underwent cardiac catheterisation with angiocardiology at 13 years of age (1957), which showed a gradient of 30 mmHg between the right ventricle and pulmonary artery, but no left-to-right shunt at ventricular level. At right ventriculography a subpulmonary diaphragm was suspected. No left sided angiography was performed. At 31 years of age cardiac catheterisation was again performed because of increased right ventricular hyper-

trophy on the electrocardiographic examination, which showed a "transvalvular" gradient of 80 mmHg. A diagnosis of pulmonary stenosis was made, and the patient refused cardiac surgery.

Five years later he was sent to our institution because of the appearance of exertional dyspnoea. On examination the following signs were found: an "a" wave on the jugular pulse, a systolic thrill at the left sternal border, and a grade 4/6 pansystolic murmur with a crescendo/decrescendo configuration.

The electrocardiogram showed severe right ventricular hypertrophy with strain. A gradient of 105 mmHg across the outflow tract of the right ventricle without any left-to-right shunt was shown at cardiac catheterisation.

Right and left ventricular injections showed a huge septal aneurysm bulging into the right ventricular outflow tract (Fig. 1). Post-stenotic dilatation of the pulmonary infundibulum was also found. During surgery the right ventricle and the infundibular septum were seen to be hypertrophic, but the free anterior wall of the infundibulum was thin and dilated. There was a huge fibrous tissue tag anterior and superior to the anteroseptal leaflet of the tricuspid valve (Fig. 2). Two small defects were found at the apex of the aneurysm. An incision made between these two defects showed a perimembranous ventricular septal defect with an infundibular extension, 1.5 cm in diameter.

Fig. 1 Left ventricular injection in long axial projection (A) and right anterior oblique (B). A huge aneurysmal tissue tag bulges into the right ventricular outflow tract (double arrows). A prolapse of the posterior leaflet of the mitral valve (single arrow) is also present.

The ventricular septal defect was closed by suturing a "teflon" patch at the muscular base of the defect, thus reinforcing the extra aneurysmal tissue. An outflow patch was also placed. The peak systolic pressure in the right ventricle and pulmonary artery was 35 mmHg at the end of this procedure.

Discussion

It is well known that a ventricular septal defect in the membranous septal area may be reduced or even closed by a frequently associated fibrous tissue tag resulting from fibrin proliferation and/or an adhesion of tricuspid valve tissue to the edges of the defect.¹⁻⁴ Bacterial endocarditis, intracavity thrombus forma-

tion with systemic emboli, conduction and rhythm disturbances, tricuspid regurgitation, and right ventricular outflow tract obstruction are rare complications of an aneurysm of the membranous septum.¹⁻⁵

Although every perimembranous ventricular septal defect can develop an aneurysm, this event is more frequently found in the ones that extend into the inlet or trabecular septum because of their proximity to the tricuspid septal leaflet.

The diagnosis of an aneurysm of the membranous septum can be detected by using elongated angiographic projections of the left ventricle,⁶ but now it can also be diagnosed by the use of two dimensional echocardiography.⁷

A prolapsed aortic valve leaflet in a supracristal ventricular septal defect, however, has echocardiographic appearances similar to those of an aneurysm of the membranous septum and should therefore be carefully ruled out.⁸

In our case the ventricular septal defect, as recently proposed,⁶ can be classified as perimembranous, extending into the infundibulum. In these particular types of defect the interventricular portion of the membranous septum is deficient and the defect extends anteriorly towards the infundibular septum.

It is easy to understand how an aneurysmal tissue tag which occludes an infundibular perimembranous ventricular septal defect can cause right ventricular outflow obstruction, quite possibly severe, if, as we have shown, the aneurysmal dimension is large.

The patient we have described shows how the development and severity of right ventricular outflow obstruction can be the result of a progressive growth of aneurysmal tissue in this peculiar position. Having reviewed the reported cases of an aneurysm of the membranous septum associated with right ventricular outflow tract obstruction, we suspect that the perimembranous ventricular septal defect had an infundibular extension, at least in the majority of these cases.

In a case previously reported⁹ of a 12-year-old patient, the gradient was 20 mmHg, which was approximately the same as the gradient found in the first haemodynamic study of our patient at a similar age. In another observation,¹⁰ the infundibular gradient was 60 mmHg whereas in the second study of our case the gradient was 80 mmHg (the ages of both cases were again similar). This, we believe, supports our hypothesis that the obstruction not only develops from an associated infundibular hypertrophy but also from the increase in the size of the aneurysm.

In conclusion, we think that every time a transinfundibular gradient, however small it may be, is recorded in a ventricular septal defect with an aneurysm of the membranous septum, the existence of a perimembranous defect with an infundibular exten-

Fig. 2(A) The huge aneurysm (black arrow) is situated below the infundibular septum (IS). One of the small defects in the aneurysm is evident (open arrow). (B) After the incision of the aneurysm a perimembranous ventricular septal defect (D) with infundibular extension appears.

sion should be suspected. If, either with the use of elongated angiographic projections or with two dimensional echocardiography, this suspicion is confirmed, we think that these patients should be carefully followed for the possible development of right ventricular outflow tract obstruction.

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