Cardiac catheterisation was performed to obtain an anatomical diagnosis. A supravalvar aortogram (fig 3) showed a grossly dilated right coronary artery with a fistulous communication with the coronary sinus. There was a small left to right shunt with a pulmonary to systemic flow ratio (Qp/Qs) of 1:4:1.

Discussion

The presentation of this patient with congenital coronary arteriovenous fistula was unusual.

This girl probably had had infective endocarditis for over a year. Turbulence over the tricuspid valve caused by the increased flow could account for the site of the vegetations on the tricuspid valve. Though infective endocarditis is a known complication of congenital coronary arteriovenous fistula it is rare. To my knowledge congenital coronary arteriovenous fistula presenting as recurrent pulmonary embolism has not been reported before.

Cross sectional echocardiography with Doppler colour flow mapping was successful in diagnosing congenital coronary arteriovenous fistula in some patients. However, it has its limitations and cardiac angiography has often been necessary to establish the diagnosis. In my patient the mid and distal portions of the right coronary artery were not visualised and hence the provisional diagnoses included ruptured sinus of Valsalva aneurysm. This limitation of cross sectional echocardiographic diagnosis has been reported before.

The additional use of Doppler colour flow mapping was not helpful in locating the distal shunt site in this patient, perhaps because the shunt was small. Transoesophageal echocardiography might have been helpful. The diagnosis of congenital coronary arteriovenous fistula was confirmed only by cardiac angiography.

I thank Miss MN Lim and Mr JM Tan for technical assistance.

Comment

The case report above describes a patient with a coronary artery fistula that was complicated by endocarditis. Clinical presentation of this congenital anomaly resembles that of aortic regurgitation and when aortic regurgitation has been excluded by echocardiographic and Doppler studies the differential diagnosis that remains is ruptured sinus of Valsalva or a coronary artery fistula communicating with either the left or the right side of the heart. When the coronary sinus is the recipient chamber echocardiography should detect the enlarged coronary sinus, which helps to establish the diagnosis. Infective endocarditis is an important complication of coronary artery fistula. In 1975 we encountered a patient in whom a coronary artery/coronary sinus fistula had been diagnosed during life, and who subsequently died despite appropriate antibiotic therapy for endocarditis. At necropsy we found that the site of the endocarditis was on a jet lesion on the wall of the coronary sinus opposite the site of entry of the coronary artery and not as
The hugely dilated coronary sinus has been opened to display the fistula (F) into the coronary artery. There are vegetations (arrows) opposite the opening of the fistula.

might have been expected on the tricuspid valve or the coronary ostium itself (figure). It has previously been suggested that infection in these cases leads to subacute bacterial endarteritis or osteitis. The findings in the patient we studied indicate that infection may develop on the jet lesion where endothelial damage has been present and where small platelet thrombi may form.

A fatal case of endocarditis complicating a coronary artery fistula was first described in 1912. The predisposition to endocarditis associated with this anomaly can be avoided by surgical ligation of the feeding artery. This will also avoid the late sequelae of right ventricular volume overload.

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