Interventricular fibroma

Echocardiographic diagnosis and successful surgical removal in infancy

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SUMMARY A 7 month old infant underwent successful surgical removal of a huge interventricular septal fibroma. We believe this to be the youngest child to have undergone successful removal of such a lesion.

Case report

Shortly after birth a baby boy was found to have a 2/6 systolic murmur, which was maximal in the pulmonary area. The murmur was thought to be insignificant and no further investigations were undertaken.

At the age of 5 months he was examined by a cardiologist, and the only positive clinical finding was a soft (2/6) mid-systolic murmur at the lower left sternal edge. Chest x ray examination showed moderate cardiomegaly with some elevation of the apex and normal pulmonary vascularity. An electrocardiogram showed sinus rhythm, a QRS axis of 105° , and an rSR pattern in V4R and V1. On M mode echocardiography chamber sizes appeared normal but the ventricular septum was thickened. Cross sectional echocardiography showed a mass in the ventricular septum extending from near the crux of the heart just inferior to the tricuspid valve to the apex but not impinging on the right ventricular outflow (Fig.1). Cardiac catheterisation showed normal saturations and pressures, and angiocardiography suggested the presence of a large mass in the right ventricular cavity, arising from the ventricular septum but not obstructing the right ventricular inflow or outflow. Serial echocardiography over the next two months showed no change in the size of the mass. There was no evidence of tuberosus sclerosis on clinical or Wood's light examination.¹

Operation was undertaken when the child was 7 months old and weighed 8.0 kg. With the patient on cardiopulmonary bypass and cooled to 25°C , the aorta was cross clamped and 300 ml of cold St Thomas's cardioplegia administered. A vertical right ventriculotomy was made and the septum visualised. The tumour occupied the septum from the posterior leaflet of the tricuspid valve to the apex. The septal endocar-

Fig. 1 Cross sectional echocardiogram showing four chambers before resection of tumour. RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium.

dium was incised between the posterior and septal papillary muscles. At a depth of 2 mm a well defined, pale firm mass measuring $5 \times 2.5 \times 2.5$ cm and weighing 16.5 g was exposed. It was removed easily from the septum leaving a 1.5×2 cm defect in the posterior wall of the right ventricle and adjacent septum. Although muscle was adherent to the tumour in places, the whole mass was excised leaving no residual macroscopic tumour. The defect was closed with a Teflon patch using pledgeted sutures. The distal third of the posterior descending coronary artery was excised with the tumour, but the mitral and tricuspid mechanisms were not disturbed and the patient was weaned from bypass without difficulty on a small dose of isoprenaline. Aortic and right ventricular PO_2 measurements confirmed adequate closure of the ventricular septal defect.

Fig. 2 Cross sectional echocardiogram after tumour resection. VS, ventricular septum. For other abbreviations see Fig. 1.

The postoperative course was complicated by respiratory difficulties which necessitated ventilatory support for seven days. Intermittent nodal bradycardia occurred, but this was not associated with low cardiac output and heart block did not occur. Histological examination of the tumour showed it to be a benign cardiac fibroma.

Chest x ray examination 15 days after operation showed a substantial reduction in heart size and a further echocardiogram (Fig. 2) showed the considerable change in septal and right ventricular sizes after excision of the tumour.

When seen eight months after operation the patient was well and maintaining growth about the 25th centile. A soft mid-systolic murmur was audible at the lower left sternal edge. Further echocardiography showed slight enlargement of the right atrium and ventricle, but no recurrent tumour, and an electrocardiogram showed complete right bundle branch block.

Discussion

Cardiac fibroma is a rare lesion, with only about 75 cases reported. We believe that this child could be the youngest to have undergone successful surgical removal of an interventricular fibroma, the previous youngest to survive having surgery at 13 months.²

Cardiac fibroma occurs most often in infants and typically affects the ventricular free wall or septum. Only seven cases have been reported in which the right ventricular free wall was affected. Cardiomegaly is a constant feature of these lesions in childhood, and it is not unusual for the child to be asymptomatic until

sudden death,³ arrhythmia,⁴ or heart failure ensues.

Although there were few physical signs in this patient, the presence of cardiomegaly on a chest x ray film led to cross sectional echocardiography, which established the diagnosis early and with anatomical accuracy. Cardiac catheterisation was undertaken to exclude other lesions but did not add further diagnostic information. Recently,⁵ contrast enhanced computed tomography has been used to delineate the extent of a large right ventricular fibroma, and this may constitute a further non-invasive diagnostic technique which, combined with serial echocardiography, may be used to exclude local recurrence.

Although cardiac fibroma is a benign condition, slow and continual growth may occur and cause conduction defects or obstruction or may spread to the ventricular free walls. Early diagnosis and surgical treatment in this patient allowed complete excision, and a recent study by Williams *et al.*⁶ suggests that cure may therefore be expected.

Although direct suture of the septum has been performed after tumour excision,⁶ a patch repair was used in this case because the remaining septal muscle was thin and the apical portion of the right ventricular free wall with 2 cm of posterior descending coronary artery had to be resected. When last seen nine months after operation, the child was in good health and should have an excellent chance of remaining disease free after complete excision.

References

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