Successful Excision of a Left Ventricular Hamartoma

Report of a Case

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PRIMARY cardiac tumors are rare. Myxomas comprise over half the total and sarcomas about one third. Among the remainder are the benign ventricular tumors, which are less than 5 per cent of the total. Surgery for cardiac tumors began with Beck's excision of a pericardial teratoma in 1942. Crafoord excised the first endocardial tumor successfully in 1951. There has been one case report to date of a partial removal of a benign ventricular tumor. We have recently excised a large hamartoma of the left ventricle from a 2-year-old boy and have observed him thereafter for 27 months.

Case Report

A 26-month-old white boy was referred to the University of California Medical Center, Los Angeles, for evaluation of a heart tumor in June 1959. He had been entirely well until 14 months of age, when he was hospitalized in Salt Lake City for 3 days with the diagnosis of bronchopneumonia. There were no signs of cardiac disease. The past history was significant in that a sibling had died at 5 days of age with aortic atresia. A lateral and superior enlargement was noted along the border of the left ventricle on chest x-ray (fig. 1). Since the heart contour suggested endocardiofibroelastosis, the patient was rehospitalized in August 1958, and an angiocardiogram showed a filling defect in the left ventricle (fig. 2). Calcification was also noted (fig. 3). The electrocardiogram showed left ventricular hypertrophy and strain. Digoxin was prescribed for the patient in November 1958, as a precaution against arrhythmias.

On October 12, 1959, the patient was admitted to the UCLA Medical Center. There were still no signs of a cardiovascular disorder. The pulse was 120, the blood pressure 94/60 in the arms and 100/70 in the legs, and the lungs were clear. The cardiac findings were normal. No murmurs were heard, and the sounds were normal. Hematologic examinations were within normal limits. A vectorcardiogram was normal, but the electrocardiogram showed left ventricular hypertrophy and strain (fig. 4).

On October 15, 1959, the patient was operated on through a median sternotomy. A large, firm, nodular tumor was noted along the lateral and posterior aspects of the left ventricle. In spite of the large size of the tumor, there seemed ample left ventricular wall for reconstruction. Extracorporeal circulation was instituted, the aorta was cross-clamped, and the heart was rotated anteriorly. The left ventricle was entered adjacent to the tumor, and the tumor mass was excised with a narrow rim of ventricular wall. No papillary muscles were damaged or detached during the excision, and adequate left ventricular wall remained for reconstruction of the ventricular cavity. The ventricular wall was closed with three layers of continuous 3-0 chronic catgut. The cardiac beat was excellent, and no difficulty was encountered in discontinuing the extracorporeal circuit. The operation was completed without incident, and the patient's postoperative course was completely benign.

In the 2 years since operation the patient has continued to have no symptoms. X-rays have shown a changing contour of the heart, suggesting a possible aneurysm at the site of surgical closure (fig. 1). Electrocardiograms also have changed, and suggest extensive fibrosis of the left ventricle in the region of the operative site (fig. 4).

The final diagnosis was fibrous hamartoma of the left ventricle. The firm tumor was irregularly nodular and measured 7.2 by 4.5 by 3.5 cm. and weighed 61.5 Gm. The tumor cut with great resistance, and calcified areas were noted along the cut surface. Microscopically the tumor was composed largely of dense fibrous tissue, but with striated cardiac muscle fibers interlaced throughout it in small numbers (fig. 5).

This microscopic description is quite similar to case reports of rhabdomyoma, fibroma, and hamartoma. All these benign left ventricular tumors

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contain fibrous and cardiac muscle elements in varying amounts. These benign hamartomatous tumors have a relatively sharp border with the ventricular muscle and a relatively small margin of excision should be curative.

Discussion

Cardiac tumors cause symptoms and signs that depend upon their location in and about the heart.

Pericardial Tumors

Large pericardial tumors cause symptoms because of either inflow blockage or pericardial effusion. The superior or inferior vena cava may be partially or totally occluded. Symptoms of constrictive pericarditis can occur with extensive epicardial involvement. Recovery of bloody pericardial fluid from a patient without evidence of bronchogenic carcinoma is diagnostic of pericardial sarcoma. Chest x-rays may show an irregular outline of the heart or the globular heart associated with pericardial effusion.

Myocardial Tumors

The benign myocardial tumors occur from birth to 8 years of age. They usually manifest themselves by paroxysmal tachycardia and unexplained heart failure. Many of the reported cases were of infants who died suddenly following moderate to severe exertion. The chest x-ray may show an abnormal bulge on the cardiac border. If this bulge contains calcification, the diagnosis of a benign hamartomatous tumor of myocardial origin is fairly certain (fig. 3). Angiocardiography can prove the diagnosis and should be done in all young children with unexplained paroxysmal tachycardia or heart failure. Murmurs may or may not be present. If present, they are systolic in timing and are caused by partial obstruction of flow within the ventricle.

Endocardial Tumors

Endocardial myxomas are the most common type of primary heart tumor. Commonly the

Figure 1

Chest x-rays from August 1958 through January 1962. Note lack of relative growth of the tumor between discovery and operation. The left lateral contour of the heart changed from 3 to 18 months after operation but not subsequently. This may represent an aneurysm at the site of left ventricular repair.
Venous angiocardiogram shows dye in the left ventricle and ascending and descending aorta. The crescent-shaped shadow in the left ventricular outflow tract outlines the hamartoma.

Right anterior oblique x-ray showing calcification within the left ventricular hamartoma.

diagnosis is made during exploration for mitral stenosis through a left thoracotomy. Over 75 per cent of myxomas occur in the left atrium and their bulk interferes with the blood flow through the mitral valve, thus giving rise to the signs and symptoms of mitral stenosis. The symptoms may vary promptly with change of position. When tricuspid stenosis is suspected, the absence of other valvular disease almost always ensures the diagnosis of a right atrial myxoma. Again, angiocardiography should be done in all suspected cases so that the surgeon may be prepared for the tumor. In adults selective angiocardiography with use of a large catheter and a pressure injector for the dye overcomes difficulties of interpretation due to faint opacification.
Endocardial tumors also give rise to arterial emboli of blood clot or tumor fragment.

Operative Treatment

The only hope of cure for cardiac tumors lies in extirpative surgery. The excision of each of the three types of tumors requires special consideration.

In the excision of pericardial tumors, which are commonly found over the right atrium and may extend to the ventricles, cavae, and aorta, particular attention must be paid to preservation of the coronary arteries. An attempt at removal of an extensive tumor must be tempered by the possibility of irreparable injury to the coronary artery.

The excision of a portion of the ventricular wall in a case of myocardial tumor can be carried out with the use of the pump oxygenator as demonstrated in our case. Technical problems evolve with concern for the preservation of adequate ventricular chamber size and adequate myocardium to pump blood from the ventricle. Fortunately, the benign hamartomatous tumors seem to enlarge by displacing the normal ventricular muscle. Thus, in our case even though a 7 by 5 cm. tumor was removed from a 2-year-old child, there still was sufficient remaining left ventricular wall to permit construction of an adequate left ventricle. The atrioventricular valves must remain competent, and no papillary muscles may be sacrificed unless they are replaced by a prosthesis.4

Endocardial tumors also should be removed with use of the pump oxygenator, to give adequate time for the removal. Parts of the tumor or clot may break off during the operation and must be removed from the left ventricle after removal of the main tumor in order to prevent later embolization. Any injury to cardiac valves must be meticulously repaired in order to prevent lethal insufficiency.

If angiocardiography is performed in all cases suspected of cardiac tumor, then the number of diagnostic explorations will be few. A definite preoperative diagnosis will make it possible to have a pump oxygenator ready in all cases of myocardial and endocardial tumors. Thus a second operation can be avoided, along with the problems of bleeding adhesions.

Frozen sections are of doubtful value in the borderline case with the problem of differentiation between a benign or sarcomatous primary tumor. Pathologists can make a diagnosis by frozen section in the case of sarcoma with implants and bloody pericardial fluid. The solitary tumor without bloody pericardial fluid or implants requires detailed, prolonged cytologic examination, often with special stains. For this reason, it would be well to consider all single tumors as locally recurrent sarcomas and remove them with as wide a margin as practical.

Summary

A case of a 2-year-old child with a large hamartoma of the left ventricular wall is presented. The excision of the tumor is described, and a 2-year follow-up is given. Primary cardiac tumors should be suspected in patients with unexplained cardiac failure, unexplained cardiac arrhythmias, intracardiac calcifications, irregular shadows on x-ray, or symptoms and murmurs that change with position. Angiocardiography is suggested for diagnosis. Technical problems at operation involve the maintenance of satisfactory cardiac function and valve competence.
Acknowledgment

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References

Osler's Personal Ideals

I have had three personal ideals. One, to do the day's work well and not to bother about tomorrow. The second ideal has been to act the Golden Rule, as far as in me lay, toward my professional brethren and towards the patients committed to my care. And the third has been to cultivate such a measure of equanimity as would enable me to bear success with humility, the affection of my friends without pride, and to be ready when the day of sorrow and grief come to meet it with courage befitting a man.—OSLER. The Quiet Art: A Doctor's Anthology. Compiled by Dr. Robert Coope. Edinburgh & London, E. & S. Livingstone Ltd., 1952, p. 94.
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