Primary Tumors of the Heart

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RECENT advances in surgical technics make it highly probable that intracavitary cardiac tumors will be successfully removed. In view of the rarity of antemortem diagnosis of these tumors it is desirable to analyze the clinical features and to call attention to an effective method of diagnosis. In addition, primary cardiac tumors involving the myocardium and pericardium will be reviewed.

Primary tumors of the heart are rare. Autopsy incidence is only 0.0017 per cent from a study of 480,331 cases.1 Tumors of the heart have been classified as either metastatic or primary. Although many metastatic cardiac tumors are recognized clinically, therapy is limited. In a few instances x-radiation has been temporarily beneficial. Primary cardiac tumors may be classified histologically or by location. Clinically, the localization of the tumor is of paramount interest. The clinical features will depend on whether the tumor is largely within the cardiac chambers or involves the myocardium or pericardium.

INTRACAVITARY TUMORS

Intracavitary tumors originate almost exclusively in the atria. The most characteristic clinical picture is that of stenosis or regurgitation or both at the mitral or tricuspid valves with no history of antecedent rheumatic fever. The growing tumor interferes with valvular function or mechanically obstructs the orifice of the valves. Exertional dyspnea is common and when cardiac failure develops, it is usually intractable because of the mechanical interference with atrial or ventricular filling. Cardiac output is severely reduced and may result in the development of angina. Systolic blood pressure is often low. Fragmentation of tumor or dislodgement of surface thrombi may lead to clinical signs of embolization. In some patients there may be definite knowledge of the absence of murmurs until the onset of the illness. Other features include arrhythmias, syncope, unexplained episodes of dyspnea, cyanosis, orthopnea and variation in symptoms and auscultatory signs with change of position of the patient.

Cardiac catheterization data are available in seven patients in whom the diagnosis of myxoma has been proved; one with a right atrial myxoma2 and six with left atrial myxoma.3-7 In all seven cases there was evidence of obstruction to outflow from the atria. This consisted of abnormal elevation of atrial pressure. In the left atrial tumors, the pulmonary capillary pressures were elevated in five patients. Cardiac output was reduced in the patient with the right atrial tumor; normal, in one patient with a left atrial tumor; and reduced, before or after exercise, in four of the others. It is doubtful that the catheterization data can be interpreted as revealing more than interference with atrial emptying. However, in the patient reported by Bayer and coworkers8 the catheterization data suggested the presence of a mass at the tricuspid valve. This is not a proven case although the angiogram suggested a right atrial tumor.

Angiocardiography has proved to be the

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Aided by a grant from the Mallinekrodt Chemical Works.
definitive method for diagnosis of intracavitary tumor. Characteristic persistent filling defects in an enlarged atrium were noted in all cases (figs. 1 and 2). In the three patients with left atrial myxoma a point of attachment of the tumor to the atrial septal wall was clearly visualized. Mural thrombi in the atrium do not produce filling defects in the angiocardiogram. Filling defects due to the regurgitant stream in tricuspid insufficiency are transient. Angiocardiography of a patient with an atrial obstructing thrombus (ball-valve or pedunculated) has not been reported. It is probable that such a thrombus will produce similar roentgen findings. Fortunately, exact differentiation is of no consequence since surgery should be considered for both the thrombus and the tumor.
The cardiac intracavitary tumors are of two types. The myxoma is the most common and in fact comprises 50 per cent of all primary cardiac tumors. It has been reported in patients from the age of 3 months to 68 years, the majority occurring between 30 and 60 years in equal sex distribution. The myxoma generally arises by a small pedicle from the atrial septum in the region of the fossa ovalis. About 75 per cent of these tumors occur in the left atrium; extra-atrial location being rare. The tumors are variable in size and may be so small as to cause no symptoms, and sometimes are discovered incidentally at autopsy. Although there has been controversy as to whether the myxoma is a neoplasm or a thrombus, prevailing opinion holds that it is a benign tumor. Strong evidence for the neoplastic nature of the myxoma is provided by a recent study reporting growth of a myxoma fragment in the mesenteric arteries and brain. This suggests that a myxoma is potentially malignant but none of the other reported cases showed lymphatic or hematogenous spread, local invasiveness or mitotic cells. They are generally smooth, pedunculated or villous, whitish tumors of variable consistency. There may be areas of hemorrhage and occasional overlying thrombi. Microscopically, the tissue is rather poorly cellular with an endocardial covering. Multinuclear and stellate cells and focal collections of plasma cells and lymphocytes have been noted. Cysts, bone and collagen deposition have been described.

The other variety of intracavitary cardiac tumor consists of sarcomas of all histological types. Although sarcomas are predominantly mural tumors they are discussed here because the presenting symptoms are frequently due to intracavitary extension. Age and sex distribution are the same as for myxoma. The sarcomas involve the right side of the heart more frequently and are polypoid in about 20 per cent. Unlike the myxomas, they involve the ventricles as often as the atria. The clinical picture produced by the intracavitary cardiac

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2A and B. Frontal and lateral chest x-ray films illustrate slight prominence of the pulmonary segment and esophageal indentation by a large left atrium (previously published).
sarcomas may resemble that due to myxoma, except for the frequent occurrence of hemo-
pericardium, arrhythmias and the more com-
mon development of inferior and superior vena caval obstruction. These tumors grow
rapidly. Five cases have been diagnosed ante-
mortem. Approximately 25 per cent of Wharton's 100 cases\textsuperscript{11} showed no metastases at
necropsy and of these there were seven poly-
poid atrial tumors.

Mural Tumors

Tumors involving the heart wall or valves, such as angiommas, hamartomas and nodular rhabdomyomas, have for the most part been reported as incidental autopsy findings. A few
strategically placed have caused arrhythmias, interference with coronary blood flow and sudden death.

Pericardial Tumors

The benign pericardial tumors are rare; only about one-fourth as frequent as benign tumors of the heart, per se. Among the reported cases are teratomas, fibromas, lipomas, angiomas, cysts and leiomyofibromas. Although these tumors usually arise from the myocardium (fig. 5) or embryonic rests, they expand into the pericardial cavity. Many are discovered because of a chance x-ray demonstration of an unusual heart contour. In others, the sudden appearance of symptoms is due to the development of a hemorrhagic pericardial effusion. This occurs almost as frequently in benign as in malignant pericardial tumors. The malignant pericardial tumors produce a more rapid course, are associated more often with chest pain and dry cough and in one-third of the cases pulmonary metastases are seen. Electrocardiographic signs are of little help in either suggesting or confirming the presence of a primary pericardial tumor since the changes usually reflect an effusion. Diagnostic artificial pneumopericardium in the presence of an effusion has been strongly advocated by Mahaim. Examination of pericardial fluid for tumor cells has led to the diagnosis in a few cases. Angiocardiography may demonstrate a unilateral pericardial mass.

Diagnosis

The symptoms which may be produced by primary cardiac tumors are so much more frequently due to other lesions that the diagnosis will depend chiefly on a sharp awareness of the possibility. The left atrial tumor must be looked for in the patients who present symptoms and signs of obstruction of the lesser circulation. Signs of mitral stenosis not dating from birth in young children, absence of cardiac difficulties until the onset of intractable heart failure, inconstancy of mitral murmurs, and episodes of syncope or cyanosis and dyspnea without adequate explanation except for change in position are important clues. Any of these findings should lead to angiocardiography. Some patients with left atrial tumors will mimic rheumatic mitral stenosis so completely as to escape detection unless angiocardiography is used routinely in the evaluation of all patients being considered for mitral valvulotomy. At present routine preoperative angiocardiography is not advocated. The need for exact preoperative diagnosis of a left atrial tumor is more than
FIG. 5A. Hemangioma of the left ventricle. The frontal angiocardiogram of a 25 year old man who had precordial pain, dyspnea and palpitation. It illustrates a nonfilling oval mass at the left cardiac border. B. Photomicrograph of the biopsy specimen reveals a sclerosing hemangioma attached to the heart wall. (Courtesy Dr. Harold A. Lyons)

academic since successful surgery may require an open technique (figs. 1–4).

Tumors of the right side of the heart will be found in patients who present signs of inflow stasis, which include dyspnea, orthopnea, dilated neck veins, facial edema, cyanosis, hepatomegaly, ascites, dependent edema, pleural effusion and increased venous pressure. Preliminary diagnoses will include constrictive pericarditis, tricuspid stenosis and insufficiency, pericardial effusion, superior vena caval syndrome and chronic congestive heart failure. The differential diagnosis of these conditions is often difficult even with the aid of a battery of laboratory examinations. Angiocardiography, in this situation will demonstrate the presence of a cardiac tumor as well as aid in the differentiation of the other conditions.

The diagnosis of primary pericardial tumors depends on the discovery of an unusual cardiac contour which is not readily explained. There are no diagnostic clinical or laboratory criteria which will differentiate pulmonary and mediastinal masses adjacent to the heart from intrapericardial tumors. If the presence of a pericardial effusion can be established, examination of the fluid may lead to diagnosis of tumor. In general the diagnosis will await surgical exploration and biopsy (fig. 5).

However, it is possible that visualization of cardiovascular rotation, as opposed to displacement in the angiocardiogram, as demonstrated in a unique case of intrapericardial tumor may provide a significant differential sign (fig. 6).

TREATMENT

Exclusive of pericardial cysts, which have been removed on many occasions, successful surgery of benign intrapericardial tumors has been reported only four times. In 1942, Beck presented a case of an intrapericardial teratoma which he removed in two stages. Eight years
later Hochberg and Robinson\textsuperscript{19} removed an intrapericardial cavernous hemangioma which was partly adherent to the myocardium. Maurer\textsuperscript{24}, in 1952, reported the removal of a large intrapericardial lipoma which arose by a small pedicle from the left ventricular wall. In 1953, Miscall\textsuperscript{15} successfully operated on a 3 year old child who had a large bronchogenic tumor in the pericardial cavity. These four reports reflect the rarity of intrapericardial tumors and illustrate the effectiveness of surgical treatment.

In his paper Beck also had suggested the possibility of removal of intra-atrial tumors. Mahaim in 1945\textsuperscript{12} extensively reviewed the literature on cardiac myxoma and concluded that the architecture of the tumor made surgical removal feasible. He emphasized the clinical features and predicted the role of angiocardiography in arriving at the diagnosis. Six cases of intra-atrial myxoma have been diagnosed during life. One of these\textsuperscript{20} was operated on with the aid of hypothermia but the patient died of ventricular fibrillation during the procedure. Bahnsen\textsuperscript{2} succeeded in removing most of a right atrial myxoma and the patient lived for 24 days after operation. Death was due in part to obstruction of the inferior vena cava by residual tumor. In two cases,\textsuperscript{3} attempted removal of the left atrial myxomas through the auricular appendage was unsuccessful; death occurred during surgery. One patient (figs. 2, 3 and 4) was explored but the small size of the auricular appendage prevented an attempt at removal of the tumor. Surgery was not performed on Kirkeby and Lerens' patient.\textsuperscript{21} In addition, several myxomata have been found during surgery for mitral stenosis or constrictive pericarditis\textsuperscript{8}.\textsuperscript{7} but attempted removal met with failure in all.

Clowes' patient\textsuperscript{22} was found to have a left atrial tumor during surgery for presumed rheumatic mitral stenosis. Surgical removal of the tumor was considered too hazardous. Six months later, after several episodes of severe pulmonary edema, an open procedure with the use of a pump oxygenator to bypass heart and lungs, was carried out. The patient died six hours after operation.

After this review was completed the following communication was received from Crafoord.\textsuperscript{23} “I can inform you that after exact anatomical diagnosis due to selective cardioangio-angiography, on July 16, 1954, we operated on a case of pseudomyxoma of the left atrium, the size of a small orange, with the aid of
complete bypass with extracorporeal oxygenation and circulation produced by our own pump-oxygenator. The blood was shunted from the cavae to the aorta through the left subclavian artery. During the 28 minutes of exclusion of the heart from the circulation and the intracardiac manipulations the heart was in a state of ventricular fibrillation intentionally brought about by electrical stimulation in order to facilitate the extracorporeal circulation and the actual technical procedure within the heart. After closure of a maximal wide incision in the left atrium, the heart was electrically defibrillated. The postoperative period was somewhat disturbed by a slight rise in both non-protein rest urea in the blood and a moderate elevation in the bilirubin. This healed otherwise without complications and the patient is now considered completely recovered. Before the operation, the patient had the usual symptoms of paroxysmal mitral occlusion and mitral stenosis which now no longer are present.”

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Circulation. 1955;11:963-970
doi: 10.1161/01.CIR.11.6.963

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/11/6/963.citation

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