Myocardial Coronary Hemangiomatous Tumors in Children*

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Persistent and severe ST segment and T wave changes associated with an apical diastolic murmur were observed in an asymptomatic five-year-old girl for ten years. Recent development of exertional dyspnea and chest pain prompted a detailed cardiovascular re-evaluation. The symptomatology and the electrocardiographic changes are attributed to an angiomatous tumor involving the anterolateral myocardium demonstrated by selective coronary arteriography. The literature regarding primary cardiac vascular tumors was reviewed and the clinical and therapeutic implications were discussed.

Primary cardiac tumors are exceedingly rare particularly in infancy and childhood. The commonest of these primary tumors is rhabdomyoma, and the most uncommon are primary hemangiomatous lesions.1-3

Anatomically, hemangiomatous tumors differ from a coronary arteriovenous cardiac fistula where one or more coronary arteries are involved,4 and hemangiomas involving the heart valves or atrial septum are now regarded as cysts of the heart valves or varicosities in the region of the foramen ovale. The greatest number of hemangiomas exclusive of cysts and varicosities have occurred among adults, have been of the cavernous variety, and have been for the most part small, subendocardial in location, and with no infiltrative features.

Uskoff+ in 1893 was the first to describe a case of hemangioma of the heart that involved the left atrium. There are few reports appearing in the literature describing various types of primary cardiac vascular tumors.5 Most of these are pericardial hemangiomas and angiosarcomas, and most of the patients reported have been adults who presented usually with cardiomegaly, congestive heart failure, and pericardial effusion.

Recently we studied a 15-year-old girl who has been followed since the age of four years because of T wave and ST segment changes, but had shown no symptoms. Selective coronary artery angiograms revealed a hemangiomatous tumor involving the left coronary artery and the anterolateral myocardium. A brief summary of the history and findings is given.

CASE REPORT

The patient is a 15-year-old girl who was seen in the cardiac clinic first in 1963 at the age of 4½ years. At that time electrocardiographic abnormalities compatible with a myocardial disease were first noted (Fig 1).

She has had a completely normal course of development, was totally asymptomatic, and her physical examination was unremarkable except for a high pitched diastolic murmur.

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Figure 1. Patient's ECG (1963) showing ST segment and T wave changes. All the precordial leads are half standard.
A left ventricular cineangiogram revealed a normal chamber in diastole. In systole, however, the cavity became abnormally small (high ejection fraction) with an abnormally thick free wall. Selective coronary angiography demonstrated a normal right coronary artery. The initial two-thirds of the left anterior descending and the oblique branch of the left coronary artery were very large and tortuous feeding into a dense vascular network that involved the apical left ventricular wall (Fig 5). No abnormal communication to any cardiac chamber was demonstrated. The distal one-third of the left anterior descending coronary artery was normal.

**DISCUSSION**

The features of this patient's vascular malformation as displayed by the coronary arteriogram are consistent with a hemangiomatous tumor of the myocardium.

She had been asymptomatic until her last admission, and the only overt abnormality she has is the ST segment and T wave changes which reflect, if anything, a certain extent of myocardial destruction and/or ischemia similar to what is seen in myocardiopathy, severe left ventricular hypertrophy, anomalous origin of the coronary artery, and coronary arteriovenous fistulas. This myocardial damage does not appear to be stationary although there have been no changes in her heart size and electrocardiogram over the years.

This patient differed from the majority of the reported cases of myocardial hemangiomas in that her coronary arteries appeared to be the origin of the vascular tumor. Coronary artery involvement by a cardiac hemangioma has been reported in only two patients before; one of those patients reported by Franciosi,² bears a striking resemblance to the patient reported here. That patient was eight years old and presented with severe cardiomegaly and ECG evidence of "LVH with strain." No murmurs could be heard. Cardiac catheterization revealed a left ventricular end-diastolic pressure of 24 mm

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**Figure 2. Aortogram (1968) demonstrating a diffuse flush over the left ventricular apex.**

That was heard over the cardiac apex.

The patient continued through 1963 and 1964 to have these electrocardiographic abnormalities in the form of ST segment depression and T wave inversion in the chest leads. In 1964 she had right and left heart catheterization and aortography which failed to demonstrate any intracardiac or coronary abnormalities. The aortogram was repeated in 1968 and, while there seemed to be no specific abnormality of the coronary arteries, this time a diffuse flush over the left ventricle compatible with a diffuse vascular lesion was visualized (Fig 2).

She continued to be totally asymptomatic and enjoying full scale activity despite the grossly abnormal electrocardiogram until October, 1973 when she started to complain of exertional dyspnea and chest pain, and she was admitted for re-evaluation.

Physical examination revealed a well-developed female in no distress. Blood pressure was 115/70 mm Hg, pulse 90/minute and respiratory rate 18/minute. The general physical examination was normal. There was a quiet precordium with no thrills. The first heart sound was normal. The second heart sound was physiologically split and not accentuated. There was a grade 1/6 soft nonspecific ejection systolic murmur over the left sternal border, and a high pitched grade 2/6 mid-diastolic murmur at the left lower sternal border and apex. The peripheral pulses were adequate and equal.

Laboratory studies included a hemogram, electrolytes, blood urea nitrogen, glucose, lipid profile, cholesterol, enzyme determinations and coagulation studies and were within normal limits. Chest x-ray films were also normal (Fig 3). The electrocardiogram exhibited the same marked ST segment and T wave changes (Fig 4).

At cardiac catheterization there was no evidence for intracardiac left-to-right shunts. The systemic cardiac index was 3.2 L/min/M². The right ventricular end-diastolic pressure was mildly elevated (10 mm Hg). After one angiogram, the left ventricular end-diastolic pressure rose to 24 mm Hg. An isoproterenol infusion failed to produce any systolic pressure gradient across the left ventricular outflow tract.

**Figure 3. Chest x-ray (1973) demonstrating no cardiomegaly or evidence of congestive heart failure.**
Hg, no shunts and no filling defects. The left anterior descending coronary artery was dilated and tortuous and the cardiac apex had an increased number of arterial branches. Histologic examinations revealed a vascular hamartoma. The second patient was reported by Timmes, but no detailed information is available.

In the past, accurate diagnosis of primary cardiac tumors was almost impossible during life and most of the literature consisted of postmortem studies and reports. At present, it is relatively easy to suspect and diagnose cardiac tumors. Hemangiomatous tumors, however, pose difficult problems since they may be present for long periods of time without producing any specific type of symptomatology, and when they do, they pose even greater problems in terms of treatment and projection of the patient into the future.

If what is known about skeletal muscle hemangiomas in general can be applied to cardiac hemangiomas, the course is indolent. However, ultimately cardiac hemangiomas are locally aggressive and are cured only by total excision if feasible. Total excision was not feasible in the patients reported by Timmes and Franciosi. Timmes’ patient was treated with radiation and was well seven years after therapy. Franciosi’s patient underwent exploration and only biopsies were obtained. May and associates reported successful resection of a vascular hamartoma, but the tumor in the patient involved only the right atrium. One of the patients reported by Van der Hauwaert died the day following an attempt to excise his tumor. Sclerosing solutions and radiation have been only temporizing measures, and the long term prognosis is poor.

REFERENCES
2 Van der Hauwaert LG: Cardiac tumors in infancy and childhood. Br Heart J 33:125, 1971
4 Reddy K, Burta M, Hamby RI: Multiple coronary arterio-systemic fistulas
5 Franciosi RA, Gay RM, Ah-Tye P: Vascular hamartoma of the heart in a child. Am Heart J 79:676, 1970