Primary Hemangioendotheliosarcoma of Heart, Diagnosed by Angiocardiography

Review of the Literature and Report of a Case

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Primary malignant tumors of the heart are rare and are seldom recognized before death. This paper reports a case of primary hemangioendotheliosarcoma of the right atrium of the heart, which was diagnosed clinically by angiocardiography. A brief review of the literature is also presented. No reports are to be found in the literature of the antemortem diagnosis of a primary hemangioendotheliosarcoma of the heart by angiocardiography. Autopsy findings are included.

Whereas a considerable number of tumors of the heart have been reported in the literature, they are still a relatively rare occurrence. Primary cardiac malignancies are rarer still, their ratio to metastatic tumors being 1 to 16, and of this group the malignant hemangioendothelioma is among the least common. Recognition of cardiac tumors during life, especially in the primary form, is extremely difficult. Up to the present time the diagnosis of primary malignant tumor of the heart has been made antemortem in only six instances; in all the tumors were sarcomas. In no case was angiocardiography employed, although the latter procedure had been successfully employed in one case of myxoma and other intra- and extracardiac tumor masses.

Only seven cases of primary hemangioendotheliosarcomas of the heart have been reported; our case will be the eighth. It is of interest to note that the case we are presenting is the first case in which the correct diagnosis was suggested antemortem by angiocardiography.

Case Report

A 45 year old Negro man entered the Cook County Hospital on Feb. 11, 1954, complaining of substernal pain, cough and dyspnea of three weeks' duration, and swelling of face and feet of one week's duration. He had been well until three weeks before admission when he rather suddenly developed substernal pain that was aggravated by coughing and deep respiration, and a cough productive of a scanty amount of rusty sputum. On one occasion about half a teaspoonful of bright red blood was coughed up. At about the same time he developed dyspnea which was most pronounced on sitting up and seemed to be relieved by lying down. Progressive swelling of his face and both feet, which appeared one week before hospitalization, was most marked at night.

Physical examination on admission showed the patient to be a well developed, well nourished middle-aged Negro man. He lay comfortably in bed. Temperature was 99 F., blood pressure 100/86, respiration 28 per minute, and pulse 112 per minute and regular. There was slight edema of the lower lip but none of the face. The cervical veins were prominently distended on both sides, more so on inspiration. Examination of the chest revealed a patchy area of bronchial breathing and moist rales below the angle of left scapula. Cardiac dullness extended to the midaxillary line in the sixth intercostal space, and there was some apparent widening of the supraventricular dullness in the supine position. The heart sound were slightly muffled. No murmur or rub was heard. The liver edge was palpable 4 cm. below the right costal margin; it was soft, smooth, and moderately tender. There was no ascites or peripheral edema.

The urine showed a trace of protein and many fine granular casts. The hemoglobin content was 89 per cent, the red cell count 4,900,000, and the leukocyte count 14,900. The nonprotein nitrogen content of the blood was 50 mg. per 100 cc. Sedimentation rate (Wintrobe method) was zero. Blood Kahn test was negative. One blood culture showed no growth. An electrocardiogram revealed a right axis shift and a semivertical electrical position of the heart, with low voltages of all the complexes, inverted T waves in precordial leads V, V, and V, and isoelectric T waves in V, V, and V. The corrected Q-T interval was 0.40 second. Venous pressure...
was 320 mm. saline by the method of Moritz and Tabora. Circulation time was 8 seconds with ether and 30 seconds with magnesium sulfate. X-ray films of chest (fig. 1) revealed gross cardiac enlargement both to left and right, with haziness over left lower chest.

Patient was treated with digitalis, penicillin, tetracycline, Meralluride and Diamox. During his hospital course he was entirely afebrile. On the fourth hospital day, a paradoxic pulse was detected. More prominent venous engorgement was noted in the neck, both upper extremities and the upper chest. On the sixth hospital day pericardiocentesis was performed through the fifth left intercostal space external to the cardiac apex and also through the left xiphocostal angle. Both attempts failed to obtain any fluid. Following this a loud pleural friction rub was heard on the left side, especially around the apex of the heart.

On the eighth hospital day angiocardiography was performed. The films taken four and one-half seconds and six seconds after the dye injection (Urokonsodium, 70 per cent) showed good filling of the azygos vein, superior vena cava and the pulmonary artery. In the enlarged right atrium, a large filling defect which was constant in shape, size, density and position was seen in the films made four and one-half and six seconds after injection of the contrast substance (fig. 2). In view of the angiocardiographic findings along with the clinical course of the patient, a diagnosis of a primary cardiac neoplasm was suggested.

The patient’s condition gradually grew worse, his edema became generalized, and the patient finally...
expired on the thirteenth hospital day, after a total known duration of illness of five weeks.

Necropsy Findings: The pericardial sac contained 50 cc. of hemorrhagic fluid. The heart weighed 700 Gm. The right atrium of the heart was enlarged. Arising from the right atrium was a large cauliflower-shaped hemorrhagic tumor mass which bulged into the pericardial cavity. The tumor extended through the wall of right atrium, arose from the posterolateral aspect of the right atrium, and almost filled the entire right atrium, extending upward into the right atrium and down through the tricuspid valve orifice into the right ventricle (fig. 3). The superior vena cava was almost completely obstructed up to the level of the innominate vein. On cross section the tumor tissue was purplish and hemorrhagic, with whitish specks scattered throughout. The remaining chambers of the heart and the valves were not unusual. The coronary arteries were all patent. Except for a few scattered hemorrhagic areas in the right ventricle, the myocardium was normal. The left pleural cavity contained 1000 cc. of bloody fluid and the underlying left lung showed atelectasis. The pleural surfaces of both lungs were covered with many fine, round, purplish, hemorrhagic nodules. The liver weighed 2000 Gm., showed severe congestion, and contained one round, slightly umbilicated, purplish tumor nodule.

Several sections from the primary tumor of the heart showed a considerable pleomorphism. Most of the tumor consisted of various sized, blood-filled spaces, making up what appeared to be poorly formed vascular channels. There were large sinuses with marked proliferation of endothelial lining and small thick-walled capillaries about these cavernous vascular spaces filled with blood (fig. 4A). The endothelial lining cells varied in shape and size, and had a varying amount of pale eosinophilic cytoplasm and hyperchromatic irregular nuclei which had a fine network of chromatin with a few small and indistinct centric or eccentric nucleoli. Mitotic figures were frequently seen (fig. 4B). In other areas the capillaries had distinct thick walls composed of concentrically layered, plump, rounded pericytes (fig. 4C). There were also areas of necrosis and hemorrhage. Scattered in the myocardium of the right ventricle were many extensive hemorrhagic infarcts with leukocytic infiltration due to obstruction of the thebesian veins.

The histologic picture of the metastatic nodules in the lungs varied from capillary hemangioendothelioma with pericytomatosis to cavernous hemangioma.

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**Fig. 3.** Interior of right atrium showing the tumor growing down through the tricuspid valve opening into the right ventricle.

**Fig. 4.** Microscopic appearance of various portions of the tumor showing (A) hemangioma, (B) hemangioendothelioma, and (C) hemangiopericytoma.
The liver showed severe congestion with marked central necrosis, but no fibrosis. The tumor nodule on the surface was a cavernous hemangioma.

DISCUSSION
The case we have presented was that of a 45 year old man with symptoms of rather sudden onset of severe congestive heart failure due to an obstruction to the inflow of blood from the great veins into the right side of the heart. It was at first thought that the patient might be suffering from tuberculous pericarditis with effusion; but this diagnosis was soon abandoned because of a “dry” pericardio-centesis. Moreover, the sedimentation rate of zero was not entirely consistent with an active tuberculous process. The diagnosis of an “idiopathic” myocarditis was then considered. However, this diagnosis was also believed unlikely because of absence of fever, normal sedimentation rate, lack of tachycardia, a normal Q-T interval in the electrocardiogram, and the atypical clinical course with a predominant picture of unexplained obstruction to the venous inflow. The possibility of an intracardiac tumor, probably a neoplasm, was therefore finally considered. Support of this diagnosis was rendered by angiocardiography, and confirmation was obtained at necropsy.

Tumors of the heart are rare. Metastatic tumors are more common than those of the primary type, and benign tumors are more frequently found than malignant primary growths. Figures based on necropsy material vary considerably. According to Straus and Merliss, the incidence of primary tumor of the heart is 0.0017 per cent. This percentage is probably close to the actual incidence of primary cardiac tumors. Mahaim collected 329 published cases of primary tumors of the heart from the world literature; of these 87 were malignant. Whorton collected 99 cases of primary malignant tumors of the heart from the literature, to which he added one of his own. In 1951, Prichard in his review brought the total of primary cardiac neoplasms to 416, of which 113 were primary sarcomas. Twenty one more have since been reported, bringing the total of reported primary sarcomas of heart to 134.

Primary cardiac sarcomas are more common on the right side, especially the right atrium, in contrast to the benign myxomas which are more often found in the left atrium. According to Whorton, three-fourths of all cases of primary cardiac neoplasms occur between the ages of 20 and 60, the mean being 43 years.

There are no pathognomonic symptoms or signs of primary malignant tumors of the heart. Several authors, particularly Yater, Mahaim, Woll and Vickery, Whorton and Pfeiffer, have discussed this subject at length. In many cases the diagnosis is one of exclusion and is usually only suggested by the peculiar, atypical course of intractable heart failure in a previously healthy individual without obvious cause. Mahaim, cited by Whorton, has pointed out that, "The symptomatology of cardiac sarcoma is dominated by the frequency of its localization in the right auricle. This may give rise to obstruction of the neck veins, edema of the face, upper trunk, upper extremities, and the development of collateral circulation. Uncommonly a cardiac tumor mass in the superior mediastinum may cause a dry cough and very rarely dysphagia. Precordial or chest pain is not uncommon. Auricular sarcomas may take the form of a polyoid lesion that gives rise to a ball-valve action on the tricuspid valve." Gould mentioned respiratory difficulty as a striking feature in a patient with atrial tumor. Grewin stated that a common syndrome in malignant tumor in the heart is cough with hemoptysis, congestion of the veins of the upper part of the body and pericarditis.

Roentgenographic examination can be helpful in diagnosis. Electrocardiographic or roentgenkymographic studies, tomography, pneumopericardium, and angiocardiography may all help to clarify the diagnosis. In our case serial films made during the period of right atrial opacification showed a large, round filling defect. This presented a regular, constant contour and border in serial films in contrast to the irregular and inconstant border seen in tricuspid insufficiency, the latter being described as the "jet sign" by Dotter, Lukas and Steinberg. Although the antemortem diagnosis of primary
tumors of the heart have been made six times previously, our case is the first case of primary cardiac sarcoma which was diagnosed by means of angiography. It seems that, had angiography been available to or used by the previous authors, more cardiac tumors could have been successfully diagnosed definitely during life.

**Summary**

1. A case of primary hemangioendothelial sarcoma of the heart is reported.
2. The diagnosis was suspected ante mortem on the basis of angiographic demonstration of a filling defect in the right atrium.
3. Certain clinical manifestations are presented which were suggestive, although not diagnostic. The recognition of this rare condition during life is not entirely impossible.
4. This case of primary cardiac sarcoma brings the total recorded to 135. It is the eighth malignant hemangioendothelioma of the heart reported, the second diagnosed during life, and the first demonstrated by angiography.

**Summario in Interlingua**

1. Es reportate un caso de primari hemangioendotheliosarcoma del corde.
2. Le diagnose esseva previde ante morte super le base de un demonstration angiocardigraphic de un defecto projicite al interior del atrio dextere.
3. Et es presentate certe manifestationes clinic que esseva de valor suggestive ben que non diagnostic. Le recognition de iste rar condition morbose durante le vita del paciente non es integremente impossible.
4. Iste caso de primari sarcoma cardiac avantia le total del casos reportate in le litteratura a 135. Inter istos illo es le octave caso de maligne hemangioendothelioma del corde, le secunde diagnosticate durante le vita del paciente, e le prime demonstrate per angiocardiographia.

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