PRIMARY HEMANGIOPERICYTOMA OF THE HEART
A Case Report

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Hemangiopericytoma (HP) represents a rare cellular vascular tumor. The present report of primary HP of the heart in a 53-year-old Japanese male is the first of its kind. Most of the tumor masses were removed, but masses tightly adhering to both the pericardium and the epicardium were not excised because of profuse bleeding. The patient has remained free of complaints for 10 months post-operatively. Non-invasive methods including a chest X-ray, echocardiogram, and thallium-201 myocardial imaging were found to be useful adjuncts of the diagnosis of cardiac tumor.

HEMANGIOPERICYTOMA (HP) is a comparatively rare cellular vascular tumor. Since 1942, when Stout and Murray\(^1\) published the first description of HP, numerous accounts of this neoplasm have been reported.\(^2\)\(^-\)\(^9\) It was recognized to occur in many locations of the body.\(^2\)\(^-\)\(^9\) However, insofar as the authors are aware, this is the first report indicating the occurrence of HP in the pericardium and the myocardium.

CASE REPORT
The patient was a 53-year-old man with no history of serious illness. He consulted his home physician for an annual physical check-up on October 10, 1976, when a physical examination as well as chest X-ray (Fig. 1) revealed normal findings. He remained in good health until August 16, 1978, when he first noted slight palpitations and arrhythmia while jogging. From December 8, 1978, he began to experience exertional dyspnea and a tingling or occasionally a constricting precordial pain. In addition to the precordial pain, discomfort was noticed in the left shoulder. On December 14, 1978, he was admitted to Kyorin University Hospital for extensive evaluation. Examination on admission revealed a blood pressure of 108/78 mmHg, pulse rate of 80/min, and respiratory rate of 28/min. His temperature was 36.5\(^{\circ}\)C. The patient appeared to be a well-developed muscular man and was in no acute distress. No cyanosis, clubbing, edema or jugular distension were detected. Cardiac examinations disclosed an apical impulse 4 cm lateral to the mid-clavicular line. The first heart sound was decreased in intensity. The second heart sound was physiologically split; the pulmonary component of the second heart sound appeared slightly prominent. The third and fourth heart sounds were not audible.

A grade 2/6, early systolic ejection murmur was best heard at the apex. The liver edge descended 4 cm below the right costal margin. A chest X-ray revealed an unusual configuration of

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Key Words:
Cardiac tumor
Hemangiopericytoma

(Received on May 31, 1980; accepted on August 18, 1980)
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Fig. 1. Chest X-ray film on October 10, 1976. The chest X-ray film, when the patient was in a good health, shows a normal cardiac shadow.

Fig. 2. Chest X-ray film on admission (December 14, 1978). The chest X-ray film reveals severe cardiomegaly with irregular margins.

The cardiac silhouette (Fig. 2). This was markedly enlarged and its margin was diffusely and finely irregular. An echocardiogram showed a dense band of echoes in the region of the anterior aspect of the heart (Fig. 3). This finding was considered to be possibly indicative of the presence of a space-occupying mass in the region of the anterior portion of the heart, presumably involving, at least in part, the right ventricular cavity. As shown in Fig. 4, thallium-201 myocardial imaging also revealed a space-occupying mass, which appeared to compress the left and right ventricles from above. An electrocardiogram showed a low voltage in the limb leads and non-diagnostic ST-T wave changes. As shown in

Fig. 3. Echocardiogram. The echocardiogram shows a dense band of echoes in the region of the anterior aspect of the heart, possibly indicating the presence of a space-occupying mass in the region of the anterior portion of the heart.

Fig. 4. Thallium-201 myocardial imaging. A large mass is observed above the right and left ventricles.

Fig. 5. Angiocardiogram. There seems to be a large mass oppressing the right ventricle from above and also obstructing the outflow tract of the right ventricle.

Fig. 5, the angiocardiogram shows that there seemed to be a large mass oppressing the right ventricle from above and also obstructing the outflow tract of the right ventricle. At cardiac

*Japanese Circulation Journal*  *Vol. 45, January 1981*
catheterization, the pressures (in mmHg) were: mean right atrium 11, right ventricle 59/11, and left ventricle 95/14. The catheter could not be inserted into the pulmonary artery, probably due to outflow obstruction by the tumor. Surgery was performed on June 30, 1979, shortly after the cardiac catheterization. The pericardium was tightly distended and discolored due to coagulated blood, exudates and tumor masses beneath it. On opening the pericardium, ap-
Fig. 8. Hemangiopericytoma. The tumor cells were found varying in shape and size. The intercellular spaces were filled with light osmiophilic materials, as seen in the perivascular basement membrane. The cytoplasm was sparse. The mitochondria were rod-shaped or ovoid with moderate numbers of fine cristae (Unanyl acetate and lead citrate. x 4000).

approximately 1000 ml of sanguineous fluid and amorphous tumor masses were obtained. The tumor masses consisted of bloody cysts, and soft and whitish masses. Upon removal of these materials, profuse bleeding from tumor tissues was noted which resulted in a blood loss of about 9000 ml during the operation. Most of the tumor masses were located anterior, lateral, and posterior to the right ventricle. The tumors were also extended, in part, anterior to the left ventricle. Following meticulous hemostasis, anteriorly and laterally located masses were removed. However, other masses which adhered tightly to both the pericardium and the epicardium were not excised because of profuse bleeding. Approximately, one third of the tumor masses appeared to remain unremoved. No attempt was made to ascertain whether or not the tumor tissues invaded into the myocardium because of his critical condition. The main coronary arteries appeared to be free from the tumor masses.

On light microscopic study, the tumor cells, mostly ovoid or spindle in shape, were arranged in a nodular fashion (Fig. 6). They were packed in and around the collapsed capillaries in a random distribution. Their cytoplasm was rather scanty and the nuclei were more or less dense, often containing moderate amounts of chromatin. The nucleoli were not distinct. They were arranged like a glomus tumor near the slit-like capillaries with a layer of endothelial lining.

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A silver reticulin impregnation showed the reticulin sheaths blacked and tumor cells on the inside of the sheaths (Fig. 7). Capillary hemangioma-like or cavernous hemangioma-like patterns were encountered in places. Hemorrhage, hemosiderin deposition and necrosis were noted in some areas. On ultrastructural examination (Fig. 8), the tumor cells were found varying in shape and size. The intercellular spaces were filled with light osmiophilic materials, as seen in the perivascular basement membrane. The cytoplasm was sparse. The mitochondria were rod-shaped or ovoid with moderate numbers of fine cristae. All these findings were consistent with and characteristic of malignant hemangiopericytoma. The patient's post-operative course was excellent and without complications. A chest X-ray, taken three months after the operation, revealed a normal cardiac configuration (Fig. 9). As shown in Fig. 10, an echocardiogram, recorded three months after the operation, revealed no band of echoes in the region of the anterior aspect of the heart, which was observed before the operation as seen in Fig. 3. After normal convalescence he was discharged on July 22, 1979. Thereafter, he has been subjected to careful follow-up at the outpatient's clinic. So far, no abnormal findings have been observed in physical examination or chest X-ray.

DISCUSSION

Hemangiopericytoma (HP) has been considered to arise from pericytes as defined by Zimmerman. Most pathologists accept HP as a well-defined entity, although some have voiced considerable skepticism on the existence of HP, insisting that it represents a pattern rather than a true entity. Pericytes are found in the capillaries and venules of almost all types of tissues. Therefore, it is not surprising that HP has been reported to arise in many parts of the body, including the bone, central nervous system, viscera, and somatic soft tissues. There has been, however, no report of HP in the pericardium and the myocardium. Thus, the question arises as to whether HP of the heart could possibly have been mistakenly classified under a different diagnosis in previous literature. It is generally recognized that HP and hemangio-endothelioma (HE) have many features in common. Both consist of an increased number of thin-walled vascular channels together with tumor cells in a network of extracellular material. A distinction between the two can usually be made both by light and electron microscopy. On light microscopy, supernumerary endothelial cells in HE versus perivascular massing of cells in HP are observed. On electron microscopy, the endothelial cells in HP are found to be essentially normal in structure and distributed along the vessels, whereas those of HE occur in excessive numbers and have a voluminous cytoplasm with many processes and irregular borders. Bearing these pathological features in mind, an extensive review was made of reported cases of HE in a search for possible pathological features of HP among them. However, insofar as the English literature was concerned, no distinct features of HP were found among any reported cases of HE.

Careful evaluation of certain morphologic criteria is helpful in predicting the clinical course of HP. Prominent mitotic activity, necrosis, hemorrhage, and increased cellularity constitute ominous signs usually observed in tumors that
later show recurrence or metastasis. On the basis of the above-mentioned malignant features, the present patient was considered as a malignant rather than benign case and as requiring careful follow-up.

Finally, mention should be made of the clinical usefulness of non-invasive methods for the diagnosis of the pericardial and myocardial tumors in the present case. The cardiac shadow on the chest X-ray revealed a large, bizarre configuration which was strongly suggestive of a cardiac tumor involving the pericardium and mediastinal tumors. The echocardiogram showed a dense band of echoes in the region of the anterior aspect of the heart, suggesting the presence of a tumor in the right ventricular cavity as well as in the pericardial space. Such echocardiographic observations were in accord with those of Chandraratna and his associates, who reported 5 patients with extracardiac masses in which a dense mass was clearly identified on M-mode echocardiograms. The two-dimensional echocardiogram also revealed the presence of a pericardial mass. Myocardial images using a thallium-201 identified the existence of an extracardiac mass in the present case, although the exact location of the mass could not be determined only from such myocardial images.

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