PRIMARY HEMANGIO-ENDOTHELIOMA OF THE HEART

REPORT OF A CASE

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Among the sixty-odd cases of primary malignant tumors of the heart and pericardium reported in the literature, four lymphangio-endotheliomas of the heart (1–4) and one angiosarcoma of the pericardium (5) are listed. Three of the four cases of lymphangio-endothelioma caused heart-block. The angiosarcoma of the pericardium gave the symptoms of pericarditis.

Only one case of hemangio-endothelioma of the heart was found in the literature. This case was reported by Penneman (6) in 1908. The tumor, 1.5 × 2.5 × 3.0 cm., was an incidental, unsuspected autopsy finding in a seventy-year-old woman whose heart was otherwise normal.

The case here presented is, to our knowledge, only the second case of hemangio-endothelioma of the heart to be recorded.
Case Report

A forty-five-year-old white woman was admitted to the Western Pennsylvania Hospital complaining of spasms in the left arm. She had had rheumatic fever at the age of twenty-five, and for the past two years had been treated at the cardiac clinic for rheumatic heart disease. During this time fibrillation was continuous except for a period of twenty-seven days, four months previous to admission, when the rhythm was normal as a result of quinidine therapy. Several days prior to admission hemiplegia developed on the left side.

The symptoms and physical findings suggested cerebral embolism, the probable origin of which was a mural thrombus in the left auricle. The pulse was irregular in force and rhythm; the rate between 68 and 70 per minute. The heart was slightly enlarged to the left. Auscultation revealed sounds of fair quality and a systolic murmur at the apex which was transmitted into the axilla. The blood pressure was 110 to 120/60. No pulse deficit was present.

During hospitalization, the patient’s paralysis improved. Twenty days before death symptoms of splenic infarction developed. Death occurred on the fifty-fourth day after admission, due to congestive heart failure and terminal bronchopneumonia. An autopsy was performed three hours post mortem.

The heart weighed 600 gm., including a large thrombus in the left auricle. The enlargement was but moderate, involving predominantly both auricles and the right ventricle. The ventricular epicardium was normal except for numerous petechiae. The epicardium of the right auricle was gray and opaque; that of the left auricle showed adherent fibrous tags upon the posterior aspect, where large numbers of intercoursing fine vessels gave the surface a violaceous color.

The left ventricle was well contracted and of average consistency; the myocardium, 15 mm. thick, appeared normal. Although the right ventricle was large, the wall was firm, 5 to 7 mm. thick, with robust trabeculae carneae. The right auricular wall was 3 to 6 mm. thick and had prominent, rotund pectinate muscles. The cavity was empty and the mural endocardium normal.

The left auricle had the consistency of a solid mass, due to the presence of a large thrombus which almost completely filled this chamber. The thrombotic mass was nodular and polypoid, yellowish-white, mottled with hemorrhagic areas, soft, and very friable. This mass almost completely blocked the orifices of the pulmonary veins, but did not extend

Fig. 2. Irregular intercommunicating tumor vessels, containing here and there a few red blood cells

The lining tumor cells are large and many project like little polypi into the lumen. × 450
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FIG. 3. LARGE BUT SIMPLE TUBULAR NEOPLASTIC VESSELS, CONTAINING DESQUAMATED TUMOR CELLS

There is a diffuse infiltration of this tissue by leukocytes. A few cardiac muscle fibers are seen in the lower right portion of the illustration. \( \times 580 \)

FIG. 4. THE ADVANCING EDGE OF TUMOR

Large tumor vessels, one containing red blood cells and tumor cells, are seen; also cardiac muscle fibers at the top of the illustration. \( \times 450 \)

into them. The thickness of the thrombus varied from 1.0 to 2.5 cm. in different regions. It covered almost the entire auricular wall, including that of the auricular appendage, and extended downward from the orifices of the pulmonary veins to within 1.0 cm. of the mitral ring anteriorly. Posteriorly the lowest point of the mass was 2.0 cm. above the mitral ring.

The left auricular wall was markedly but irregularly thickened (Fig. 1). Posteriorly the thickness exceeded 13 mm. and was associated with a greatly increased consistency. The endocardium beneath the thrombus presented numerous, irregularly scattered, slightly raised, flattened nodules, the surfaces of which were roughened and hemorrhagic. They varied in size from 3 to 6 mm. in diameter. A number of these nodules, 1 to 2 mm. in diameter, were present on the auricular aspect of the mitral valve. Hanging from the valve
The cells are spindle-shaped and form only occasional slender vessels. × 450

FIG. 6. A NODULE OF TUMOR TISSUE IN THE EPICARDIUM. × 450

surface was a cluster of globular, friable vegetations measuring $0.5 \times 1.0 \times 1.0$ cm. The mitral orifice resembled a buttonhole, $1 \times 2$ cm. The leaflets were irregularly thickened and rigid. Associated with the fusion of the mitral leaflets were thickening and retraction of the chordae tendineae.

The cut surface of the left auricular wall presented a dry, firm tissue, mottled brown and yellowish-white, with numerous coarse interlacing bundles of gray fibrous tissue. The thrombus was delimited from the auricular wall by thickened subendocardial fibrous tissue.

Minute filiform vegetations were present near the line of closure of the aortic cusps. The tricuspid valve showed diffuse thickening of the leaflets associated with thickening and shortening of the chordae tendineae.

Microscopically the wall of the left auricle showed partial and, in some regions, extensive replacement of the myocardium by tumor tissue. The tumor, as a whole, was composed of closely crowded vessels which varied from complex cavernous structures to simple
capillary tubes. The cells lining these vessels were often epithelioid in character and varied markedly in size and shape. Many were large and spherical, with abundant, deeply staining cytoplasm, and projected like polypi into the lumen. The nuclei were often gigantic and hyperchromatic. Some were round or lobulated; others were of irregular shape. Mitotic figures were infrequent. The vessels contained variable proportions of blood and desquamated tumor cells.

The neoplastic vessels replaced the muscle fibers, which were widely separated from one another by interstitial edema. There was considerable infiltration of the tumor by polymorphonuclear leukocytes and of the myocardial tissues by lymphocytes and large mononucleated cells, and, in foci, by plasma cells. Fresh hemorrhages and hemosiderin deposits were frequently encountered.

Tumor cells were found free in the lumen of capillaries of normal appearance some distance from the tumor. Associated with the tumor embolism was an apparent increase in the number and size of capillaries, which were markedly engorged with blood. An occasional small tumor nodule was found beneath the epicardium.

There was considerable invasion of the thickened and hyalinized subendocardial connective tissue by the neoplastic vascular structures, which extended into the adjacent thrombotic mass. The latter was indistinguishable from the endocardium except for the delimiting elastic tissue. The thrombus had three zones: an outer, organized zone, which was hyalinized and replete with tumor tissue; a middle zone consisting of granular débris of erythrocytes and fibrin, devoid of tumor cells; and an inner zone, where the erythrocytes and fibrin were better preserved. The inner surface of the thrombus was covered by an irregular layer of tumor cells, sometimes four to six cells deep, although more commonly only one cell in thickness. These cells were more often smaller and stained more deeply than those elsewhere and were even more variable in size and shape. They were, as a rule, not interconnected. Occasional foci of loosely dispersed, spindle-shaped tumor cells, which often resembled fibroblasts, were found in this zone of the thrombus and near the lumen.

Thrombi covered by tumor cells were also found upon the mitral valve, on both the auricular and the ventricular aspects, and in one instance in the left ventricular cavity, near the apex.

Aside from the histologic changes incident to the presence of the tumor, the sections showed inflammatory changes consistent with the diagnosis of rheumatic pancarditis.

Other findings included: old and recent splenic infarcts, old hemorrhage in the cortex of the right cerebral hemisphere in the rolandic area, bilateral hydrothorax, and bilateral

Fig. 7. Portion of Thrombus Covered by Tumor Cells. × 450
bronchopneumonia. No tumor cells were found in the splenic infarcts or in the region of the cerebral hemorrhage, nor were there any metastases.

**Comment**

In contrast to the circumscribed hemangioendothelioma reported by Penneman (6), the tumor here described was a diffusely infiltrating one, involving not only all portions of the auricular wall, but also thrombi in the left auricular cavity, and those attached to the mitral valve, as well as to the left ventricular wall. Another point of difference is the presence, in our case, of severe rheumatic pancarditis with an associated high-grade mitral stenosis, whereas in Penneman's case the heart was apparently normal. Both tumors originated in the left auricle, and the histologic appearance of the two neoplasms is strikingly similar.

Very little can be said regarding the etiology. The frequency of rheumatic heart disease and the rarity of primary malignant tumors of the heart in general, and of this type of tumor in particular, preclude any attempt to associate the two diseases.

**Summary**

A case of primary hemangioendothelioma of the heart is described. The tumor diffusely infiltrated the wall of the left auricle and involved thrombi which were adherent to the left auricular wall, the mitral valve, and the left ventricular wall. Severe rheumatic pancarditis with mitral stenosis was also present. There was no evidence of tumor embolism or of metastases outside of the heart.

**Bibliography**