

3-1985

Leiomyosarcoma of the Heart: A Twenty-Year Cure

Gerald Fine

B. Usha Raju

Follow this and additional works at: <https://scholarlycommons.henryford.com/hfhmedjournal>



Part of the [Life Sciences Commons](#), [Medical Specialties Commons](#), and the [Public Health Commons](#)

Recommended Citation

Fine, Gerald and Raju, B. Usha (1985) "Leiomyosarcoma of the Heart: A Twenty-Year Cure," *Henry Ford Hospital Medical Journal* : Vol. 33 : No. 1 , 41-44.

Available at: <https://scholarlycommons.henryford.com/hfhmedjournal/vol33/iss1/11>

This Article is brought to you for free and open access by Henry Ford Health System Scholarly Commons. It has been accepted for inclusion in Henry Ford Hospital Medical Journal by an authorized editor of Henry Ford Health System Scholarly Commons.

Leiomyosarcoma of the Heart: A Twenty-Year Cure

Gerald Fine, MD* and B. Usha Raju, MD*

Excision of a predominantly intracavitary right atrial tumor, which mimicked a number of other clinical disorders, effected a 20-year cure. Microscopic, ultrastructural, and immunocytochemical characteristics of the tumor were those of smooth muscle; cellular an-

aplasia, mitotic activity and tumor infiltration of the auricular myocardium indicated malignant neoplasia, a leiomyosarcoma. Distribution of the tumor was consistent with its origin from the auricular endocardium.

PPrimary malignant cardiac neoplasms, encountered infrequently, may cause a variety of symptoms but occasionally lead to sudden death without previous clinical manifestations. Total extirpation of such cancers by procedures short of cardiac transplantation is virtually impossible because of the extensive myocardial and (or) pericardial infiltration. Only those tumors that are principally exophytic into a cardiac chamber or the pericardial cavity and have minimal myocardial involvement are likely to be amenable to surgical removal. This report documents a 20-year cure following excision of a primary cardiac leiomyosarcoma.

Case Report

A 14-year-old boy was hospitalized in February, 1965 with dyspnea, weakness, and two episodes of syncope. All were attributed to an earlier upper respiratory infection and sore throat. Blood pressure was 94/80 and 100/82, respectively, in the right and left arms, pulse 126/min. A Grade IV-VI harsh blowing systolic murmur was heard best at the left sternal border radiating to the left axilla, and a thrill was felt over the right supraclavicular area. At times a protodiastolic gallop rhythm with tachycardia was observed. P2 was greater than A2. The liver was enlarged three fingerbreadths below the right costal margin and the jugular veins were markedly distended. Femoral, radial, and dorsalis pedis arterial pulsations were good.

Laboratory studies revealed: hemoglobin of 9 g/dL with hematocrit of 33%, reticulocyte count 7% to 9%, platelets 33,500 to 79,000 per cubic mm, WBC 9,350 to 12,400 per cubic mm with 53% polymorphonuclear neutrophil leukocytes and 47% lymphocytes, clotting time 6 to 7 min, prothrombin time 17 sec, partial thromboplastin time 42 sec, prothrombin consumption 13 sec, absence of clot lysis in 24 hours, negative serum bilirubin and Coomb's test, and normal total serum protein and fractions. Adequate megakaryocytes, normal myeloid series, and an increase in normoblastic activity were observed in the bone marrow. ECG revealed a sinus rhythm, PR interval of 0.16 sec, an incomplete right bundle branch block, low voltage upright T waves over the left precordium, and peaked P waves.

Chest radiography revealed enlargement of the right heart identified at fluoroscopy as right atrial enlargement. Cardiac

angiograms were interpreted as showing a right atrial tumor with an attachment to the atrial septum by a broad pedicle. The right ventricular and right atrial pressure were 12 and 20 mm Hg, respectively. Oxygen saturation was 56%, 61%, 48% and 45%, respectively, in the superior vena cava, inferior vena cava, right atrium, and right ventricle.

Thoracotomy disclosed a pedunculated endocardial tumor attached to and distorting the right atrial appendage, filling most of the right atrial cavity and distending the tricuspid valve. It was removed with a segment of attached atrial appendage. Postoperative recovery was uneventful. The liver became nonpalpable and the heart rhythm, ECG, hemoglobin, red blood cells, and platelets returned to normal within several days after the operation.

There has been no evidence of local recurrence or metastasis in the 20 years since the tumor was removed.

Pathology

A circumscribed 6.5 x 6 x 4.8-cm smooth-surfaced, firm tumor with an attached segment of right atrial appendage was submitted (Fig 1). Its cut surface was variegated gray and red with large soft areas covered with a mucinous fluid.

Microscopically, a loose, fine, collagenous and reticular tissue containing areas of fibrin, acid mucopolysaccharide and coagulation necrosis with ghosts of tumor cells made up 40% to 60% of the many sections studied. Tumor cells were sparsely and irregularly distributed in various areas of the stroma and were concentrated particularly at the base of the tumor (Fig 2). They were elongated with a variable amount of finely vacuolated cytoplasm that often extended from one or both ends of the cell and stained moderately with the fuchsin of the Masson trichrome stain. Only in the more cellular areas was there parallel alignment of the tumor

Submitted for publication: December 6, 1984

Accepted for publication: February 25, 1985

*Anatomic Pathology, Henry Ford Hospital

Address reprint requests to Dr. Raju, Division of Anatomic Pathology, Henry Ford Hospital, 2799 W Grand Blvd, Detroit, MI 48202.

cells resembling muscle (Fig 3). No cytoplasmic striations were found to indicate striated muscle differentiation. Nuclear pleomorphism was moderate, and some tumor cells contained large, irregular, chromatin-rich nuclei with prominent nucleoli. Mitoses were present infrequently, five in 50 high power fields (10x ocular and 40 objective). Scattered lymphocytes and rare plasma cells were intermixed with the tumor cells. Immediately adjacent to the tumor, the endothelial cells lining the endocardium were prominent and covered a small portion of the tumor's base. The auricular myocardium immediately underlying the mass contained tumor interdigitating with cardiac muscle, but tumor did not extend to the surgical margin. Features of smooth muscle were found in many of the tumor cells at the ultrastructural level. These consisted of parallel-arranged 8 to 10-nm cytoplasmic filaments, pinocytotic vesicles, basement membranes at the periphery of the tumor cells, and sparsely distributed dense bodies and peripheral plaques (Fig 4). A number of tumor cells contained prominent, sometimes dilated, rough endoplasmic reticulum with finely granular material in the cisterns.

Immunocytochemical examination of formalin-fixed, paraffin-embedded tumor using the Avidin-Biotin peroxidase complex technique (1) and employing rab-

bit antiserum to actin (1:1000) and myosin (1:600) (Miles Scientific Laboratory, 30 W 475 North Aurora Rd, Naperville, IL 60566) for 18 hours at room temperature revealed a positive reaction in many of the tumor cells that was less intense than that in the cardiac muscle. Reaction with 1:800 antimyoglobin (Dako Corporation, 22 North Mipap, Santa Barbara, CA 93103) for 18 hours was negative in the tumor cells but strongly positive in the cardiac muscle.

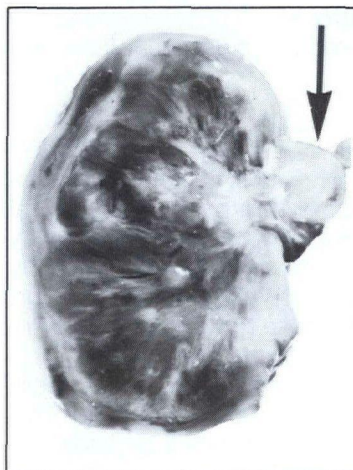


Fig 1
One half of the smooth-surfaced tumor with a portion of underlying atrial appendage (arrow). Reprinted from Fine G. Neoplasms of the pericardium and heart. In: Gould SE, ed. Pathology of the heart and blood vessels. 3rd ed. Fig XIX-17, p 870. Copyright 1968, Charles C Thomas, Springfield, IL.

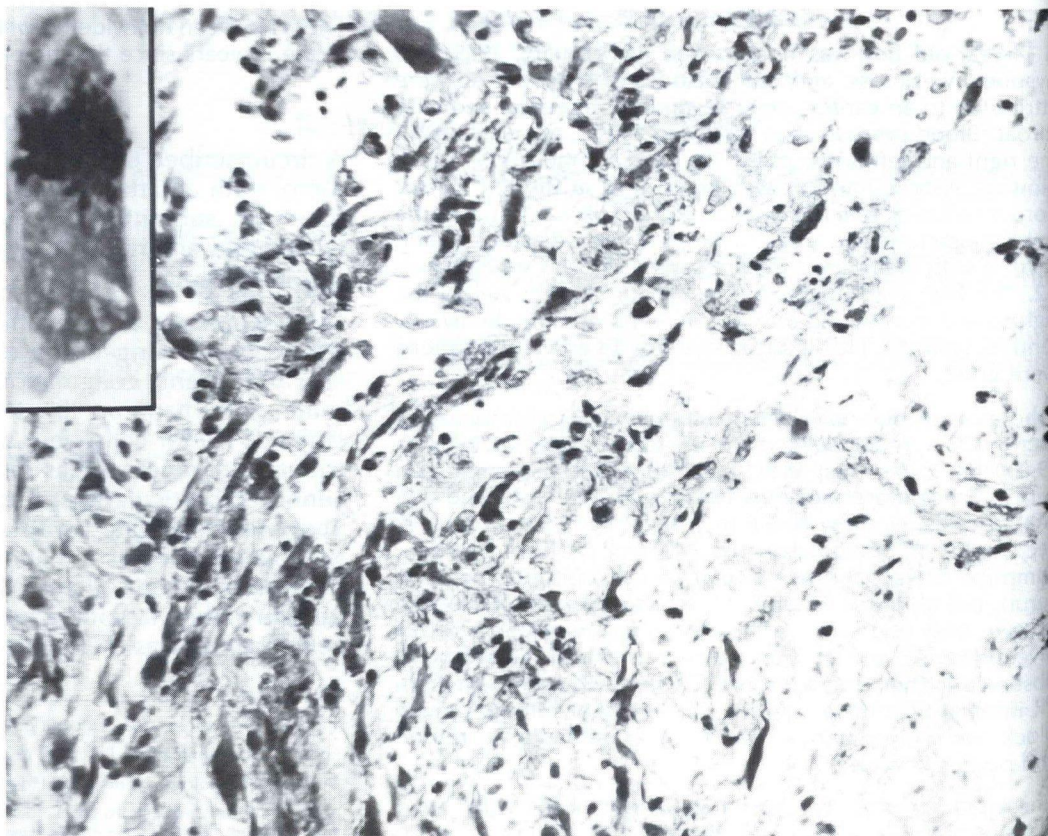


Fig 2
Loosely and haphazardly arranged tumor cells in a loose mucinous stroma. Detail of a dividing cell in inset. H&E 375x. Inset H&E 1200x.

Sarcoma: mately c tumors. second in coma ha were rec pared to sarcoma: ocardium
The freq rence in a in this ca However not confi atrial atta microsc served microgra tumor ce lack of a r thelial ce the micro diagnosis (6). Subs tions for did not muscle.

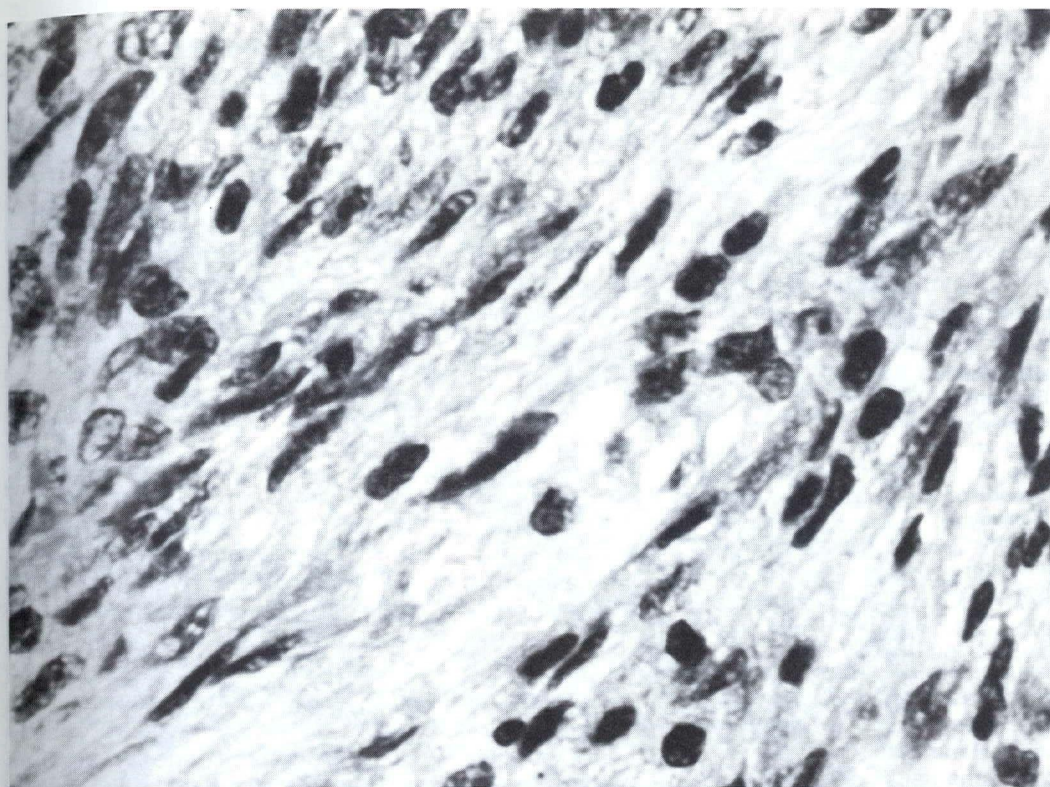


Fig 3
A more heavily cellular portion of the tumor in which an appearance of muscle is suggested. H&E 1200x.

Discussion

Sarcomas of the heart are encountered with approximately one third the frequency of benign cardiac tumors. Among the myogenic sarcomas, which rank second in frequency to the angiosarcoma, leiomyosarcoma has been reported rarely. Seven such tumors were recorded among 303 cardiac sarcomas (2-5) compared to 26 rhabdomyosarcomas among 125 cardiac sarcomas (5). No report of leiomyoma of the myocardium has been found in the literature.

The frequency of the cardiac myxoma and its occurrence in an angiographic setting similar to that observed in this case lend credence to the diagnosis of myxoma. However, surgical and pathological observations did not confirm this impression. The auricular rather than atrial attachment of the tumor as well as its gross and microscopic characteristics were unlike personally observed cardiac myxomas and published photomicrographs of such tumors. The arrangement of the tumor cells, often with a nonmucinous stroma, and the lack of a micropapillary tumor surface covered by endothelial cells were not like the cardiac myxoma. Instead, the microscopic features were those of muscle, and the diagnosis of rhabdomyosarcoma was favored initially (6). Subsequently, positive immunocytochemical reactions for actin and myosin indicated a muscle tumor but did not distinguish between smooth and striated muscle. The negative myoglobin reaction supported

the diagnosis of smooth muscle tumor rather than rhabdomyosarcoma. Ultrastructure established conclusively the smooth muscle histogenesis.

Malignancy of the neoplasm was questioned by some observers because of the scant cellularity and mitotic activity and abundant stroma. However, the nuclear pleomorphism, the mitotic activity, even though infrequent, and the infiltration of the auricular myocardium fulfill the criteria established for a low grade extracardiac leiomyosarcoma (7,8).

Plausible sites of origin for cardiac leiomyosarcoma are vascular or endocardial intimal smooth muscle. In the present case, the bulky polypoid and microscopic plaque-like intimal involvement with limited infiltration by tumor support an endocardial origin.

The varied symptoms and clinical findings manifested by this patient have been associated with other polypoid intracavitary cardiac tumors, and alternative diagnoses are often considered—rheumatic heart disease, carcinoid syndrome, congenital heart disease (Ebstein's anomaly), atrial myxoma and viral myocarditis. Cardiac angiography established the presence of intracardiac tumor, but microscopic examination was necessary to define its histogenesis. The mechanism for some of the abnormal laboratory findings is not clear, but their return to normal after tumor removal indicates a causal relationship.

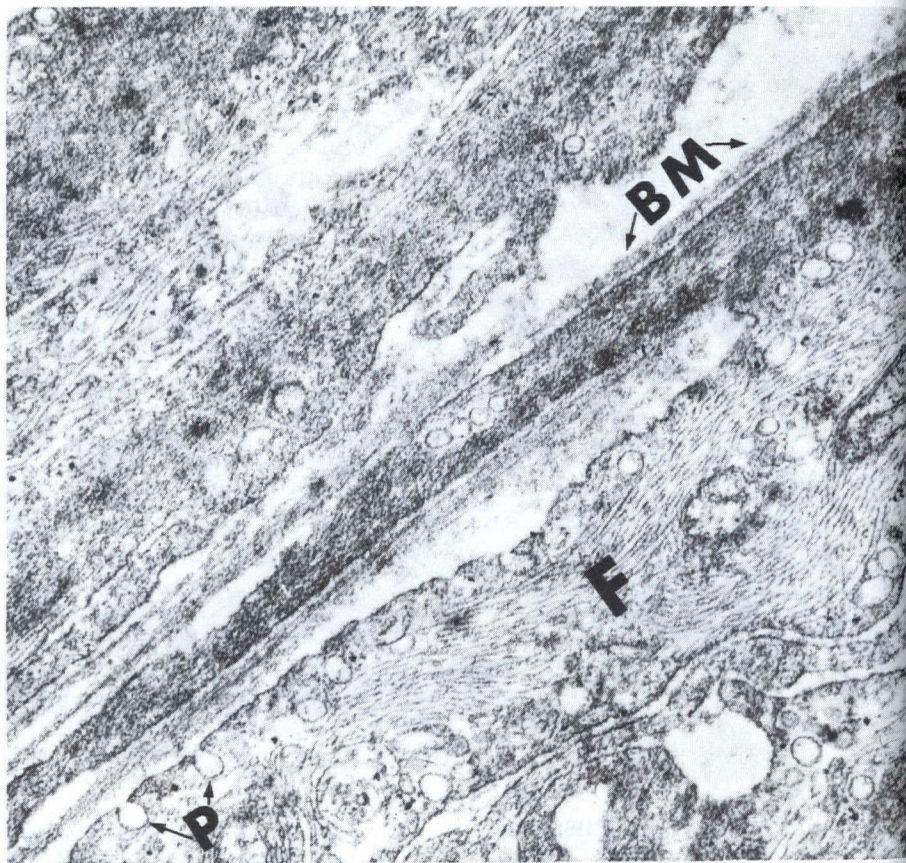


Fig 4

Portions of a number of tumor cells illustrate basement membrane (BM), filaments (F), and pinocytotic (P) vessels. The sparsely distributed dense bodies and plaques are not present in these cells. The above cell components are diagnostic of smooth muscle. The arrangement of the filaments and absence of cytoplasmic bands further negate striated muscle differentiation. Uranyl Acetate-Lead Citrate 12000x.

References

1. Hsu SM, Raine L, Fanger H. The use of antiavidin antibody and Avidin-Biotin-Peroxidase Complex in immunoperoxidase technics. *Am J Clin Pathol* 1981;75:816-21.
2. Whorton CM. Primary malignant tumors of the heart. *Cancer* 1949;2:245-60.
3. Brucker EA, Glassy FJ. Primary reticulum cell sarcoma of the heart with review of the literature. *Cancer* 1955;8:921-31.
4. Somers K, Lothe F. Primary lymphosarcoma of the heart. *Cancer* 1960;13:449-57.
5. McAllister HA, Fenoglio JJ Jr. Tumors of the cardiovascular system. *Atlas of tumor pathology*. Armed Forces Institute of Pathology, 1978: fascicle 15.
6. Fine G. Neoplasms of the pericardium and heart, In: Gould SE, ed. *Pathology of the heart and blood vessels*. Springfield, IL: Charles C Thomas, 1968:869-75.
7. Stout AP, Hill WT. Leiomyosarcoma of the superficial soft tissue. *Cancer* 1958;11:844-54.
8. Ranchod M, Kempson RL. Smooth muscle tumors of the gastrointestinal tract and retroperitoneum. A pathologic analysis of 100 cases. *Cancer* 1977;39:255-62.

Henry Ford
Vol 33, No

Centra

John E. Br

Central por
that has occ
hyponatrem
who had a
signs of cen
tion of se

Central p
neurologic
axons. Who
call (1), CP
deficiency
However,
occurs clin
(2,3). Exp
healthy do
induced h
characteriz
sparing of
vessels, a
central po
pontine m
thalamus,
ebellum, a

CPM shou
condition
natremia.
progress
plegia, an
within ho
natremia

Patients g
disease, a
at autopsy
have sur
(7,8,12,13)

This repor
after corre
had prima

A 52-year-
from cough
Three days
weakness
received 5