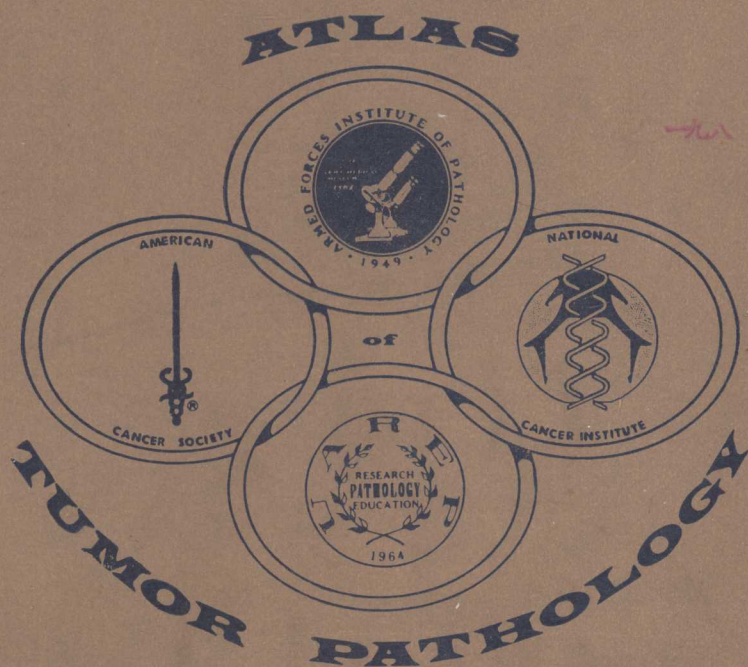


TUMORS of the CARDIOVASCULAR SYSTEM

15



一九八一年二月十二日

ATLAS OF TUMOR PATHOLOGY

Second Series

Fascicle 15

TUMORS OF THE CARDIOVASCULAR SYSTEM

by

HUGH A. McALLISTER, JR., M. D.
COLONEL, MC, USA

Chairman, Department of Cardiovascular Pathology
Armed Forces Institute of Pathology
Washington, D. C. 20306
and

JOHN J. FENOGLIO, JR., M. D.
MAJOR, MC, USAR

Staff Pathologist
Department of Cardiovascular Pathology
Armed Forces Institute of Pathology
Washington, D. C. 20306
Present Address

Assistant Professor of Pathology
College of Physicians & Surgeons of Columbia University
New York, New York 10032



Published by the
ARMED FORCES INSTITUTE OF PATHOLOGY
Washington, D. C.

Under the Auspices of
UNIVERSITIES ASSOCIATED FOR RESEARCH AND EDUCATION IN PATHOLOGY, INC.
Bethesda, Maryland

Published in the U.K.
by
Castle House Publications Ltd.,
27 London Rd., Tunbridge Wells, Kent TN1 1BX

ISBN 0 7194 0043 0

1979

ATLAS OF TUMOR PATHOLOGY

Sponsored and Supported by

AMERICAN CANCER SOCIETY*
NATIONAL CANCER INSTITUTE, NATIONAL INSTITUTES OF HEALTH*
and
ARMED FORCES INSTITUTE OF PATHOLOGY

EDITOR

WILLIAM H. HARTMANN, M. D.

Professor and Chairman, Department of Pathology
Vanderbilt University School of Medicine
Nashville, Tennessee 37232
Area Code 615 - 322-6609

COEDITOR

WILLIAM R. COWAN, COLONEL, USAF, MC

Armed Forces Institute of Pathology
Washington, D. C. 20306

EDITORIAL ADVISORY COMMITTEE

Pelayo Correa, M.D.	Louisiana State University School of Medicine New Orleans, Louisiana 70112
Harlan I. Firminger, M.D.	University of Colorado Medical Center Denver, Colorado 80220
David G. Freiman, M.D.	Harvard Medical School and Beth Israel Hospital Boston, Massachusetts 02215
Myron R. Melamed, M.D.	Memorial Sloan-Kettering Cancer Center New York, New York 10021
Henry C. Pitot, M.D.	University of Wisconsin Medical Center Madison, Wisconsin 53706
Richard J. Reed, M.D.	Tulane University School of Medicine New Orleans, Louisiana 70112
Juan Rosai, M.D.	University of Minnesota Medical School Minneapolis, Minnesota 55455
John H. Yardley, M.D.	The Johns Hopkins School of Medicine Baltimore, Maryland 21205

*(Supported by Public Health Service grant no. CA08575 from the
National Cancer Institute and American Cancer Society grant no. MG-122)

EDITORS' NOTE

The Atlas of Tumor Pathology was originated by the Committee on Pathology of the National Academy of Sciences—National Research Council in 1947. The form of the Atlas became the brainchild of the Subcommittee on Oncology and was shepherded by a succession of editors. It was supported by a long list of agencies; many of the illustrations were made by the Medical Illustration Service of the Armed Forces Institute of Pathology; the type was set by the Government Printing Office; and the final printing was made by the press at the Armed Forces Institute of Pathology. The American Registry of Pathology purchased the fascicles from the Government Printing Office and sold them at cost, plus a small handling and shipping charge. Over a period of 20 years, 15,000 copies each of 40 fascicles were produced. They provided a system of nomenclature and set standards for histologic diagnosis which received worldwide acclaim. Private contributions by almost 600 pathologists helped to finance the compilation of an index by The Williams & Wilkins Company to complete the original Atlas.

Following the preparation of the final fascicle of the first Atlas, the National Academy of Sciences—National Research Council handed over the task of further pursuit of the project to Universities Associated for Research and Education in Pathology, Inc. Grant support for a second series was generously made available by both the National Cancer Institute and the American Cancer Society. The Armed Forces Institute of Pathology has expanded and improved its press facilities to provide for a more rapid and efficient production of the new series. A new Editor and Editorial Advisory Committee were appointed, and the solicitation and preparation of manuscripts continues.

This second series of the Atlas of Tumor Pathology is not intended as a second edition of the first Atlas and, in general, there will be variation in authorship. The basic purpose remains unchanged in providing an Atlas setting standards of diagnosis and terminology. Throughout the rest of this new series, the term chosen for the World Health Organization's series "International Histological Classification of Tumours" (when available) is shown by an asterisk if it corresponds to the authors' choice, or as the first synonym in bold print if it differs from the authors' heading. Hematoxylin and eosin stained sections still represent the keystone of histologic diagnosis; therefore, most of the photomicrographs will be of sections stained by this technic, and only sections prepared by other technics will be specifically designated in the legends. It is hoped that in many of the new series a broader perspective of tumors may be offered by the inclusion of special stains, histochemical illustrations, electron micrographs, data on biologic behavior, and other pertinent information when indicated for a better understanding of the disease.



The format of the new series is changed in order to allow better correlation of the illustrations with the text, and a more substantial cover is provided. An index will be included in each fascicle.

It is the hope of the Editors, past and present, the Editorial Advisory Committees, past and present, and the Sponsors that these changes will be welcomed by the readers. Constructive criticisms and suggestions will always be appreciated.

William H. Hartmann, M. D.

William R. Cowan, M. D.

PREFACE AND ACKNOWLEDGMENTS

Since publication of the first fascicle on Tumors of the Cardiovascular System, numerous examples of primary tumors of the heart and great vessels have been carefully collected and catalogued in the Department of Cardiovascular Pathology of the Armed Forces Institute of Pathology, under the direction of the late Dr. William C. Manion and, subsequently, Dr. Hugh A. McAllister. This vast body of material has formed the basis for this volume which is not intended to replace the classic fascicle of Dr. Benjamin Landing and Dr. Sidney Farber, but rather to present new information on tumors of the heart and great vessels. The vasoformative lesions and tumors of the peripheral vascular system are not covered in this volume. These lesions are presented in detail in the original fascicle, Tumors of the Cardiovascular System, and under specific organ systems in the individual fascicles of the second series.

The number of primary tumors of the heart and pericardium available for review in the AFIP probably exceeds one half of all published cases in the world literature, and that portion of the current fascicle is based on these documented cases. Appropriate references, appearing at the conclusion of each section, primarily include major review articles but do not represent a total review of the literature on cardiac tumors. Our purpose is to present data from a large series of cases in which a definitive diagnosis has been established rather than to compile data from numerous published case reports in which diagnoses could not always be substantiated.

Unfortunately, the AFIP does not have a comparable volume of cases of tumors of the great vessels, and in this section we have had to rely on the literature. Since it is difficult in many instances to document the appropriate histologic diagnosis from the published descriptions, we have compiled the published experience on tumors of the great vessels and have not attempted to estimate the incidence of the various entities.

We wish to thank the many doctors who contributed interesting cases to the AFIP, and especially those who responded over the past two decades to Dr. Manion's personal requests for unusual cardiac tumors. Unfortunately, neither space nor circumstances permit individual acknowledgment of all contributors. Inevitable administrative changes over the years have depleted some of the files containing the names of many who undoubtedly deserve recognition. We are grateful to the following colleagues, whose correspondence remains, for their special interest in our quest for examples of cardiac neoplasms:

Dr. Giorgio Baroldi, Milan, Italy; Dr. Weldon K. Bullock, Los Angeles, California; Dr. J. N. P. Davies, Albany, New York; Dr. Israel Davidsohn, Chicago, Illinois; Dr. Mark Entman, Houston, Texas; Dr. Frank J. Glassy, Sacramento, California; Dr. Pedro Grases, Caracas, Venezuela; Dr. Antal Haraszti, Eger, Hungary; Dr. Gordon R. Hennigar, Charleston, South Carolina; Dr. Barton McSwain, Nashville, Tennessee; Dr. Erwin Uehlinger, Zurich, Switzerland; and Dr. Wallace N. Yater (deceased), Washington, D. C.



We are deeply indebted to the many individuals at the Institute who have toiled with us during the preparation of this fascicle. Most especially, we wish to thank Mrs. Marie Cunningham and Mrs. Grace Lawson who meticulously assisted in the time-consuming, often frustrating compiling, indexing, and abstracting of volumes of reference material and cases; Mrs. Anna B. Kelly and Mr. Charles E. Edwards for their expertise and assistance in photography; Mr. John I. Dexter for his technical assistance in obtaining and preparing specimens selected for illustration; and Mr. Jerry L. Badders and staff for their critical advice and artistry in the design, layout, and printing of the color plates.

Judith Lawson, our most able secretary and editorial assistant, has endured with us throughout this project. More than anyone else, she deserves credit for the encouragement and support of our efforts to bring this material to publication as an ultimate realization of Dr. Manion's aspiration to give the medical community a definitive text on cardiovascular neoplasms.

Our appreciation is extended to Dr. Harlan I. Firminger and Dr. William H. Hartmann and the editorial staff for their unending patience. The constructive comments of Dr. Jack L. Titus and Dr. Gerald Fine were most helpful. Most especially, we wish to thank Mrs. Audrey Cyr who continually saw the light at the end of the tunnel.

Finally, we wish to extend a very special, warm tribute to Mrs. Billie Manion. Her quiet devotion to her husband in all his pursuits and her enthusiastic support of his professional endeavors were in a very real sense most important contributions to this work.

Hugh A. McAllister, M. D.

John J. Fenoglio, Jr., M. D.

TUMORS OF THE CARDIOVASCULAR SYSTEM

Contents

	Page No.
Primary Tumors and Cysts of the Heart and Pericardium	1
Benign Tumors and Cysts of the Heart and Pericardium	5
Myxoma	5
Papillary Fibroelastoma	20
Papilloma of the Epicardium	25
Rhabdomyoma	25
Fibroma	32
Lipomatous Hypertrophy of the Atrial Septum	40
Lipoma	44
Hemangioma	46
Varix	51
Blood Cyst	52
Mesothelioma of the Atrioventricular Node	52
Teratoma	59
Bronchogenic Cyst	62
Pericardial Cyst	64
Tumors Simulating Pericardial Cyst	67
Other Benign Cardiac Tumors	68
Granular Cell Tumor	68
Hamartoma	68
Heterotopic Tissue	68
Leiomyoma	69
Lymphangioma	69
Neurofibroma	70

Contents-continued

	Page No.
Malignant Tumors of the Heart and Pericardium	73
Mesothelioma	73
Angiosarcoma	81
Rhabdomyosarcoma	88
Fibrosarcoma and Malignant Fibrous Histiocytoma	95
Malignant Lymphoma	99
Extraskeletal Osteosarcoma	100
Malignant Nerve Sheath Tumors	102
Malignant Teratoma	104
Thymoma	106
Other Malignant Cardiac Tumors	106
Leiomyosarcoma	106
Liposarcoma	108
Synovial Sarcoma	108
Metastatic Tumors of the Heart and Pericardium	111
Primary Tumors of Major Blood Vessels	121
Primary Tumors of Large Veins	122
Leiomyoma	123
Leiomyosarcoma	124
Primary Tumors of Large Arteries	126
Primary Malignant Tumors of the Pulmonary Artery	126
Primary Malignant Tumors of the Aorta and Large Arteries	129
Intimal Fibroplasia	130
Index	137



TUMORS OF THE CARDIOVASCULAR SYSTEM

PRIMARY TUMORS AND CYSTS OF THE HEART AND PERICARDIUM

Primary tumors of the heart and pericardium are extremely rare, with an incidence between 0.0017 percent and 0.28 percent in reported or collected autopsy series. By 1968, approximately 700 primary tumors of the heart and pericardium, including cysts, had been reported. Since 1968, approximately 100 additional cases have been added to the literature. In total, between 800 and 1000 primary tumors and cysts of the heart and pericardium have been reported.

We have reviewed 533 primary tumors and cysts of the heart and pericardium from the files of the Armed Forces Institute of Pathology. This number includes only those cases for which adequate clinical histories and representative histologic slides or gross hearts were available. Having had the opportunity to review and assess the histologic findings of each case, we have elected not to include published statistics in our series, because the histology is frequently unclear and the reviewer must depend on the authors' diagnosis. This fascicle, therefore, is based on the AFIP collection, although pertinent information from other reports and appropriate references have been included.

The 533 primary cysts and tumors of the heart and pericardium in the AFIP collection are summarized in Table 1. A number of these lesions have previously appeared in the literature, either as single reports or in a collected series.

The most common cardiac tumor is the myxoma. Twenty-five percent of all tumors and cysts of the heart and pericardium are

TABLE 1
TUMORS AND CYSTS
OF THE HEART AND PERICARDIUM

Type	Number	Percent
Benign		
Myxoma	130	24.4
Lipoma	45	8.4
Papillary fibroelastoma	42	7.9
Rhabdomyoma	36	6.8
Fibroma	17	3.2
Hemangioma	15	2.8
Teratoma	14	2.6
Mesothelioma of the A-V node	12	2.3
Granular cell tumor	3	
Neurofibroma	3	
Lymphangioma	2	
Subtotal	319	59.8
Pericardial cyst	82	15.4
Bronchogenic cyst	7	1.3
Subtotal	89	16.7
Malignant		
Angiosarcoma	39	7.3
Rhabdomyosarcoma	26	4.9
Mesothelioma	19	3.6
Fibrosarcoma	14	2.6
Malignant lymphoma	7	1.3
Extraskeletal osteosarcoma	5	
Neurogenic sarcoma	4	
Malignant teratoma	4	
Thymoma	4	
Leiomyosarcoma	1	
Liposarcoma	1	
Synovial sarcoma	1	
Subtotal	125	23.5
Total	533	100.00

myxomas, and 40 percent of benign cardiac tumors are myxomas. In adults, almost half of the benign tumors are cardiac myxomas

TABLE 2

TUMORS AND CYSTS OF THE HEART AND PERICARDIUM IN ADULTS*

Type	Number	Percent
Benign		
Myxoma	118	26.6
Lipoma	45	10.1
Papillary fibroelastoma	42	9.5
Hemangioma	11	2.5
Mesothelioma of the A-V node	9	2.0
Fibroma	5	1.1
Teratoma	3	
Granular cell tumor	3	
Neurofibroma	2	
Lymphangioma	2	
Rhabdomyoma	1	
Subtotal	241	54.3
Pericardial cyst	80	18.0
Bronchogenic cyst	6	1.4
Subtotal	86	19.4
Malignant		
Angiosarcoma	39	8.8
Rhabdomyosarcoma	24	5.4
Mesothelioma	19	4.3
Fibrosarcoma	13	2.9
Malignant lymphoma	7	1.6
Extraskeletal osteosarcoma	5	1.1
Thymoma	4	
Neurogenic sarcoma	3	
Leiomyosarcoma	1	
Liposarcoma	1	
Synovial sarcoma	1	
Subtotal	117	26.3
Total	444	100.00

*Patients 16 years of age and older.

(Table 2). Approximately one-fourth of all tumors and cysts of the heart and pericardium are malignant. Of the malignant tumors, one-third are angiosarcomas; 20 percent are rhabdomyosarcomas, 15 percent are mesotheliomas, and 10 percent are fibrosarcomas.

In children and infants, the most common cardiac tumor is the rhabdomyoma (Tables 3 and 4). Malignant tumors are rare in the pediatric age group; they comprise less than 10 percent of all tumors

TABLE 3

TUMORS AND CYSTS OF THE HEART AND PERICARDIUM IN CHILDREN*

Type	Number	Percent
Benign		
Rhabdomyoma	35	39.3
Fibroma	12	13.5
Myxoma	12	13.5
Teratoma	11	12.4
Hemangioma	4	4.5
Mesothelioma of the A-V node	3	3.4
Neurofibroma	1	1.1
Subtotal	78	87.6
Pericardial cyst	2	2.2
Bronchogenic cyst	1	1.1
Subtotal	3	3.4
Malignant		
Malignant teratoma	4	4.5
Rhabdomyosarcoma	2	2.2
Neurogenic sarcoma	1	1.1
Fibrosarcoma	1	1.1
Subtotal	8	9.0
Total	89	100.00

*Patients 15 years of age or younger.

and cysts of the heart and pericardium. In infants (less than one year of age), more than 75 percent of tumors and cysts are rhabdomyomas or teratomas. In children from one to 15 years of age, myxomas, rhabdomyomas, and fibromas are the most common cardiac tumors, in that order; these three tumors account for 80 percent of benign tumors and 60 percent of all tumors and cysts in this age group.

In this fascicle, tumors are classified on the basis of histologic type rather than by location. Generally, except for mesotheliomas of the atrioventricular (A-V) node and papillary fibroelastomas, and possibly myxomas, tumors of the heart and pericardium are similar histologically to tumors of the soft tissues.

References

- Fine, G. Neoplasms of the Pericardium and Heart, pp. 851-883. In: Pathology of the Heart and Blood Vessels, 3d ed. Gould, S. E. (Ed.). Springfield, Ill.: Charles C Thomas, 1968.
- Primary tumors of the pericardium and heart. Cardiovasc. Clin. 5:207-238, 1973.
- and Horn, R. C., Jr. Soft tissue tumors. Aids in differential diagnosis. Pathol. Annu. 4:211-229, 1969.
- Mahaim, I. Les Tumeurs et les Polypes du Coeur. Paris: Masson & Cie, 1945.
- Nadas, A. S. and Ellison, R. C. Cardiac tumors in infancy. Am. J. Cardiol. 21:363-366, 1968.
- Prichard, R. W. Tumors of the heart. Review of the subject and report of one hundred and fifty cases. Arch. Pathol. 51:98-128, 1951.

TABLE 4

TUMORS AND CYSTS OF THE HEART AND PERICARDIUM IN INFANTS*

Type	Number	Percent
Benign		
Rhabdomyoma	28†	58.3
Teratoma	9	18.8
Fibroma	6	12.5
Hemangioma	1	2.1
Mesothelioma of the A-V node	1	2.1
Subtotal	45	93.7
Bronchogenic cyst	1	2.1
Subtotal	1	2.1
Malignant		
Fibrosarcoma	1	2.1
Rhabdomyosarcoma	1	2.1
Subtotal	2	4.2
Total	48	100.00

*Patients one year of age or younger.

†Includes 3 stillborn infants.

Stout, A. P. and Lattes, R. Tumors of the Soft Tissues. Fascicle 1, Second Series, Atlas of Tumor Pathology. Washington: Armed Forces Institute of Pathology, 1967.

Van der Hauwaert, L. G. Cardiac tumors in infancy and childhood. Br. Heart J. 33:125-132, 1971.

Whorton, C. M. Primary malignant tumors of the heart. Report of a case. Cancer 2:245-260, 1949.



TABLE 4
TUMORS AND CYSTS OF THE HEART
AND PERICARDIUM IN INFANTS*

Type	Number	Percent
Benign		
Rhabdomyoma	28	58.3
Teratoma	9	18.8
Fibroma	6	12.5
Hemangioma	1	2.1
Mesothelioma of the A-V node	1	2.1
Subtotal	45	93.7
Bronchogenic cyst	1	2.1
Subtotal	1	2.1
Malignant		
Fibrosarcoma	1	2.1
Rhabdomyosarcoma	1	2.1
Subtotal	2	4.2
Total	48	100.00

*Patients one year of age or younger.
†Includes 3 stillborn infants.

Stout, A. P., and Lister, R. Tumors of the Soft Tissues.
Fascia II. Second Series. Atlas of Tumor Pathology.
Washington: Armed Forces Institute of Pathology,
1957.
Van der Houwer, L. G. Cardiac tumors in infancy and
childhood. Br. Heart J. 33:128-133, 1971.
Wharton, G. M. Primary malignant tumors of the heart.
Report of a case. Cancer 2:243-250, 1949.

and cysts of the heart and pericardium. In
infants (less than one year of age), more
than 75 percent of tumors and cysts are
rhabdomyomas or teratomas. In children
from one to 15 years of age, myxomas,
rhabdomyomas, and fibromas are the most
common cardiac tumors; in that order,
these three tumors account for 80 percent
of benign tumors and 60 percent of all
tumors and cysts in this age group.

In this fascicle, tumors are classified on
the basis of histologic type rather than by
location. Generally, except for mesotheli-
omas of the atrioventricular (A-V) node
and papillary fibroelastomas, and possibly
myxomas, tumors of the heart and pericar-
dium are similar histologically to tumors of
the soft tissues.

References

1. Stout, A. P., and Lister, R. Tumors of the Soft Tissues. Fascia II. Second Series. Atlas of Tumor Pathology. Washington: Armed Forces Institute of Pathology, 1957.
2. Van der Houwer, L. G. Cardiac tumors in infancy and childhood. Br. Heart J. 33:128-133, 1971.
3. Wharton, G. M. Primary malignant tumors of the heart. Report of a case. Cancer 2:243-250, 1949.
4. Pridmore, R. W. Tumors of the heart: Review of the sub-
ject and report of one hundred and fifty cases. Arch.
Pathol. 57:85-128, 1951.
5. Nadas, A. S., and Ellison, R. C. Cardiac tumors in infancy.
Am. J. Cardiol. 21:323-336, 1968.
6. Mason, B. G. 1968.
7. Mahaim, J. Les Tumeurs et les Polypes du Coeur. Patho-
logique. Paris: Masson, 1968.
8. Pridmore, R. W., and Hume, R. C. The soft tissue tumors. A review
of the literature. Pathol. Annu. 4:211-239, 1969.
9. Cardiac tumor. Clin. 5:120-128, 1971.
10. Primary tumor of the pericardium and heart.
C. Thomas, 1968.
11. Gold, S. E. 1961. Springfield, Ill: Charles
C. Thomas, 1968.
12. 88-893. In: Pathology of the Heart and Blood Vessels.
3rd ed. Gold, S. E. 1961. Springfield, Ill: Charles
C. Thomas, 1968.

BENIGN TUMORS AND CYSTS OF THE HEART AND PERICARDIUM

MYXOMA

SYNONYMS AND RELATED TERMS: Angioreticuloma; right atrial myxoma; left atrial myxoma; endothelioma; intracardiac endodermal heterotopia.

Definition. Myxoma is the most frequent primary tumor of the heart. It arises from the endocardium as a polypoid, often pedunculated tumor mass, extending into a cardiac chamber. Cardiac myxomas are derived from mesenchymal cells of the sub-endocardial layer and imitate primitive mesenchyme.

Clinical Aspects. The clinical presentation of the cardiac myxoma is most dependent upon the cardiac chamber involved by the tumor. The presentations of the 130 patients in the AFIP series are summarized in Table 5. For convenience, all patients with signs and symptoms of mitral valve disease are tabulated together.

Before 1960, cardiac myxomas were generally diagnosed as, and the patient treated for chronic rheumatic heart disease. Since that time, as new diagnostic methods of cardiology (cardiac catheterization and, most recently, echocardiography) have become available, and as clinicians have become increasingly aware of the ability of cardiac myxomas to imitate mitral valve disease, these tumors have been correctly diagnosed and surgically excised. Of the 57 patients with clinical signs and symptoms of mitral valve disease in this series, 56 had a myxoma in the left atrium; the other had a myxoma in the right atrium. Clinical symptoms included dyspnea, progressive or refractory congestive heart failure, systolic and diastolic murmurs, and atrial arrhythmias. There have been extensive reviews in the literature of the symptoms of patients with myxomas in the left atrium and the

TABLE 5

CARDIAC MYXOMA CLINICAL PRESENTATIONS IN 130 PATIENTS*

Signs and symptoms of mitral valve disease	57
Embolic phenomena	36
No cardiac symptoms — incidental finding	16
Signs and symptoms of tricuspid valve disease . .	6
Sudden unexpected death	5
Pericarditis	4
Myocardial infarction	3
Signs and symptoms of pulmonary valve disease .	2
Fever of undetermined origin	2

*One patient with multiple myxomas had signs and symptoms of mitral and tricuspid valve disease.

clinical basis by which these patients are distinguished from patients with intrinsic mitral valve disease. Although this distinction may be difficult by physical examination, murmurs of changing intensity and systemic findings, such as an elevated sedimentation rate or increased gamma globulin levels, may be of diagnostic aid. Myxoma should be suspected in any patient with signs and symptoms of mitral valve disease, especially of recent onset.

The second most common clinical presentation of patients with cardiac myxomas is embolic phenomena. In 36 patients in this series, the initial event was embolic and was present in patients with myxomas arising both in the left and right side of the heart. The six patients with pulmonary emboli from a right atrial myxoma created diagnostic problems. They each had persistent fever, changing murmurs, and progressive congestive heart failure—all are symptoms of recurrent pulmonary emboli. Of the patients with myxomas in the left atrium and ventricle, slightly more than one-third (12 patients) died of a cerebrovascular accident, and an embolic myxoma was found at necropsy. Symptoms in the remaining 18 patients were sudden hemiparesis, sudden diplopia, or sudden loss of blood supply in an extremity. The common denominator was sudden onset of symptoms of arterial occlusion, and, in retrospect, these patients had either cardiac murmurs, often of changing intensity, or atrial arrhythmias. Many of these tumors were diagnosed following an arterial embolotomy. Occasionally, the myxoma was not recognized microscopically, and some patients had repeated episodes of peripheral embolization before a cardiac myxoma was suspected. All but one of the patients with a myxoma in the left ventricle pre-

sented with signs and symptoms of embolic phenomena. The association of arterial occlusion of sudden onset with cardiac murmurs, especially of varying intensity, should alert the clinician to the possibility of a cardiac myxoma.

An additional 18 patients had episodes of arterial embolic occlusion during the course of their disease or evidence of myxoma emboli at autopsy. Three patients who presented with signs and symptoms of myocardial infarction had a myxoma embolus in at least one coronary artery at necropsy.

Patients with a myxoma in the left side of the heart most frequently have symptoms of mitral valve disease or of embolic phenomena. These were the presenting symptoms in 86 of 108 patients with myxomas of the left atrium or ventricle in our series. In the remaining patients, multiple cardiac myxomas were present or the myxoma was an incidental finding at the time of necropsy.

In contrast to myxomas in the left side of the heart, those arising in the right side of the heart may mimic any number of clinical entities. In addition to pulmonary embolic phenomena, patients with myxomas in the right atrium or ventricle may have signs and symptoms of tricuspid valve disease (frequently diagnosed as Ebstein's deformity), of pulmonary stenosis, or of pericarditis and may even present with fever of undetermined origin. A myxoma should be suspected when signs and symptoms of tricuspid or pulmonary valvular disease develop, especially when the cardiac murmurs are variable, as they so frequently are in patients with myxomas in any cardiac chamber. Rhythm disturbances, especially right bundle branch block and atrial flutter or fibrillation, and ab-

normal peaked P waves on electrocardiogram are frequently seen in patients with a myxoma in the right side of the heart. They should alert the clinician to the possibility of a cardiac myxoma, especially in a patient with signs and symptoms of pericarditis or with a fever of undetermined origin. As with myxomas on the left side of the heart, newer advances in diagnostic cardiology have greatly aided in the diagnosis of right atrial and right ventricular myxomas.

Nine percent of the patients in our study were in the pediatric age group (15 years or under). The youngest patient was four years old. We have never seen a myxoma in a newborn or young infant. More than 50 percent of the patients with cardiac myxoma were in their fourth, fifth, and sixth decades; however, myxomas occur in all age brackets, and 12 percent of our patients were over age 70. Most series indicate a nearly equal sex distribution.

Description. Although cardiac myxomas differ in clinical presentation, depending on the chamber in which they are located, they are similar in gross and microscopic appearance, irrespective of their location. Sites of myxomas in the AFIP series are summarized in Table 6. The myxomas were multiple in 5 percent of the patients. In all these patients, one tumor arose in the left atrium, and symptoms were most frequently due to this myxoma. In three patients, however, a second myxoma was situated on the right side of the heart, and in one patient, a myxoma was present in each of the four cardiac chambers. Most myxomas arising in the atria, either left or right, are attached to the atrial septum, usually in the region of the limbus of the fossa ovalis. Only 1 of the 10 ventricular myxomas was attached to the ventricular septum. Atrial

myxomas do not invariably originate from the atrial septum; 10 percent of the atrial myxomas in our series originated from sites in the atria other than the septum. After the atrial septum, the most common site was the posterior atrial wall, followed by the anterior atrial wall and the atrial appendage. We have never seen a myxoma arising from a cardiac valve.

Grossly, myxomas are usually polypoid, friable, and pedunculated, although some may be smooth-surfaced and rounded. Many myxomas have a short, broad-based attachment. True sessile myxomas are a rarity, if they exist at all. The majority of so-called sessile myxomas have previously embolized, leaving only the broad base of the polypoid tumor attached to the endocardium. Myxomas are soft and gelatinous, almost mucoid in appearance, and gray to gray-white, often with areas of hemorrhage or thrombus. Their size varies from 1 cm. up to 15 cm. in diameter, although the majority of myxomas are 5 to 6 cm. in diameter (figs. 1-7).

TABLE 6
CARDIAC MYXOMA
LOCATION OF
138 MYXOMAS IN 130 PATIENTS*

Site	Number	Percent
Left atrium	103	74.5
Right atrium	25	18.1
Right ventricle	5	3.7
Left ventricle	5	3.7

*Multiple myxomas were present in 6 patients.

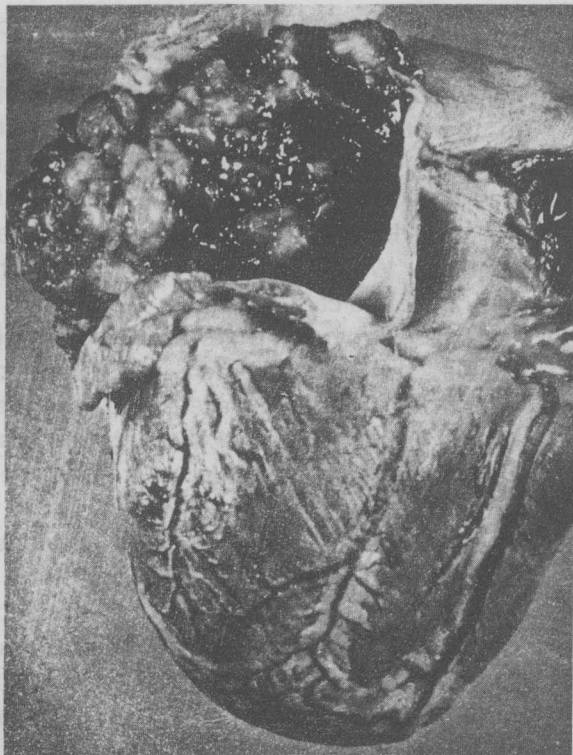


Figure 1

MYXOMA

(Figures 1 and 2 from same case)

A cardiac myxoma fills the left atrium. The majority of myxomas consist of multiple, friable, polypoid fronds and have a distinctive mucoid or gelatinous appearance. Approximately 75 percent of cardiac myxomas arise in the left atrium, as illustrated here, and 18 percent arise in the right atrium.



Figure 2

MYXOMA

The most common site of attachment of myxomas in the left atrium (fig. 1) is the atrial septum adjacent to the fossa ovalis. The fronds of the myxomas are extremely friable and embolization is a common occurrence. Extreme care must be taken during surgical removal of the myxoma to avoid operative embolization.