

# Heart

**Protocol applies to primary malignant cardiac tumors.**

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*Protocol revision date: January 2004  
No AJCC/UICC staging system*

## **Procedures**

- **Cytology** (No Accompanying Checklist)
- **Incisional Biopsy**
- **Excisional Biopsy**

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## Surgical Pathology Cancer Case Summary (Checklist)

*Protocol revision date: January 2004  
Applies to malignant cardiac tumors only  
No AJCC/UICC staging system*

### HEART: Resection

Patient name:

Surgical pathology number:

**Note: Check 1 response unless otherwise indicated.**

### MACROSCOPIC

#### Specimen Type

Excisional biopsy

Other (specify): \_\_\_\_\_

Not specified

#### Tumor Site (check all that apply)

Pericardium

Right ventricle

Left ventricle

Right atrium

Left atrium

Other (specify): \_\_\_\_\_

Not specified

#### Tumor Size

Not applicable

Greatest dimension: \_\_\_ cm

\*Additional dimensions: \_\_\_ x \_\_\_ cm

Cannot be determined (see Comment)

**MICROSCOPIC****Histologic Type**

- Angiosarcoma  
 Malignant fibrous histiocytoma  
 Myxosarcoma  
 Fibrosarcoma  
 Leiomyosarcoma  
 Rhabdomyosarcoma  
 Osteosarcoma  
 Synovial sarcoma  
 Malignant schwannoma (malignant peripheral nerve sheath tumor)  
 Malignant mesenchymoma  
 Other (specify): \_\_\_\_\_  
 Sarcoma, type cannot be determined

**Histologic Grade**

- Not applicable  
 Cannot be determined  
 Low-grade  
 High-grade  
 Other (specify): \_\_\_\_\_

**Extent of Invasion (as appropriate)**

- Cannot be determined  
 No involvement of adjacent tissue(s)  
 Involvement of adjacent tissue(s)  
 Other organ involvement (specify): \_\_\_\_\_

**Margins (as appropriate)**

- Not applicable  
 Cannot be assessed  
 Uninvolved by tumor  
 Involved by tumor  
     Specify site(s), if known: \_\_\_\_\_

**\*Additional Pathologic Findings (check all that apply)**

- None identified  
 Benign tumor (specify): \_\_\_\_\_  
 Therapy-related changes (specify): \_\_\_\_\_  
 Inflammation  
 Other (specify): \_\_\_\_\_

**\*Comment(s)**

\* Data elements **with asterisks** are **not required** for accreditation purposes for the Commission on Cancer. These elements may be clinically important, but are not yet validated or regularly used in patient management. Alternatively, the necessary data may not be available to the pathologist at the time of pathologic assessment of this specimen.

## Background Documentation

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*Protocol revision date: January 2004*

### **I. Cytologic Material (Pericardial Fluid)**

#### **A. Clinical Information**

1. Patient identification
  - a. Name
  - b. Identification number
  - c. Age (birth date)
  - d. Sex
2. Responsible physician(s)
3. Date of procedure
4. Other clinical information
  - a. Relevant history
    - (1) primary cardiovascular disease
    - (2) myocarditis
    - (3) congenital heart disease
    - (4) history of tumor elsewhere
    - (5) immunosuppression
    - (6) tuberous sclerosis
    - (7) previous irradiation
  - b. Relevant findings (eg, echocardiographic [ECHO] findings, evidence of tumor elsewhere in body)
  - c. Clinical diagnosis
  - d. Procedure (eg, fine-needle aspiration [FNA] of pericardial fluid)
  - e. Anatomic site(s) of specimen (eg, anterior pericardial sac)

#### **B. Macroscopic Examination**

1. Specimen
  - a. Description
  - b. Unfixed/fixed (specify fixative)
  - c. Number of slides received, if appropriate
  - d. Quantity, appearance of fluid specimen, if appropriate
  - e. Results of intra-procedural consultation
2. Material submitted for microscopic evaluation
3. Results of rapid smear review
4. Special studies (specify)

#### **C. Microscopic Evaluation**

1. Adequacy of specimen (if unsatisfactory for evaluation, specify reason)
2. Tumor (Note **A**)
  - a. Histologic type, if possible
  - b. Histologic grade, if possible
3. Additional pathologic findings, if present
  - a. Therapy-related changes
  - b. Degenerative changes
  - c. Atypical cellular reaction
  - d. Inflammation
  - e. Other

4. Status/results of special studies (specify)
5. Comments
  - a. Correlation with intraoperative consultation, as appropriate
  - b. Correlation with other specimens, as appropriate
  - c. Correlation with clinical information, as appropriate

## II. Incisional or Excisional Biopsy

### A. Clinical Information

1. Patient identification
  - a. Name
  - b. Identification number
  - c. Age (birth date)
  - d. Sex
2. Responsible physician(s)
3. Date of procedure
4. Other clinical information
  - a. Relevant history
    - (1) primary cardiovascular disease
    - (2) myocarditis
    - (3) congenital heart disease
    - (4) history of tumor elsewhere
    - (5) immunosuppression
    - (6) tuberous sclerosis
    - (7) previous irradiation
  - b. Relevant findings (eg, ECHO findings, evidence of tumor elsewhere in body)
  - c. Clinical diagnosis
  - d. Procedure
  - e. Operative findings
  - f. Anatomic site(s) of specimen (eg, pericardium, left/right ventricle, atrium)

### B. Macroscopic Examination

1. Specimen
  - a. Tissue(s) received
  - b. Unfixed/fixed (specify fixative)
  - c. Number of fragments
  - d. Dimensions
  - e. Descriptive features (color/consistency)
  - f. Orientation, if designated by surgeon
  - g. Results of intraoperative consultation
2. Tumor
  - a. Size (Note **B**)
  - b. Descriptive features (eg, consistency, color, hemorrhage, necrosis)
  - c. Extension
3. Margins, if appropriate
  - a. Vascular
  - b. Pericardial
  - c. Other
4. Tissue submitted for microscopic evaluation
  - a. Tumor (Note **C**)
  - b. Designated areas including those marked adherent to other structures

- c. Margin(s)
- d. Frozen section tissue fragment(s) (unless saved for special studies)
- e. Other (specify)
- 5. Special studies (specify) (eg, histochemistry, immunohistochemistry, electron microscopy, morphometry, DNA analysis [specify type])

### C. Microscopic Evaluation

- 1. Tissue(s) present
- 2. Tumor
  - a. Histologic type(s) (Note **D**)
  - b. Histologic grade (Note **E**)
  - c. Status of designated areas
  - d. Extent of invasion (adjacent tissues)
- 3. Margins, as appropriate
- 4. Additional pathologic findings, if present
  - a. Benign tumor
  - b. Therapy-related changes
  - c. Degenerative changes
  - d. Atypical cellular reaction
  - e. Inflammation
  - f. Other
- 5. Results/status of special studies (specify) (Note **F**)
- 6. Comments
  - a. Correlation with intraoperative consultation, as appropriate
  - b. Correlation with other specimens, as appropriate
  - c. Correlation with clinical information, as appropriate

## Explanatory Notes

### A. Cytologic Findings

Pericardial effusions are rarely caused by primary cardiac tumors. The most common causes of malignant pericardial effusions are metastatic adenocarcinoma from lung or breast, malignant melanoma, or extension of malignant mesothelioma into the pericardium. The pathologist should evaluate the nature and clinical significance of a malignant pericardial effusion by discussing the findings with the clinician, reviewing the patient's medical record, or both. Cellular changes considered to be infective, reactive, or degenerative (eg, viral infection, immunotherapy, chemotherapy, or radiation effect) should be clearly distinguished from malignant or atypical (potentially malignant) cytologic findings. Additional patient history and pertinent clinical findings may be helpful in arriving at a definitive diagnosis.

### B. Staging

The greatest diameter of the tumor in centimeters should be recorded. There is no published staging system for primary cardiac tumors.

### C. Number of Sections

The number of sections varies with the size of the specimen and the nature of the neoplasm. The pathologist should sample areas with diverse gross appearances. In addition to tumor evaluation, routine sampling of the non-neoplastic components of the specimen should be performed.

#### D. Histologic Type

The classification of malignant cardiac tumors as recommended by the Armed Forces Institute of Pathology (AFIP) fascicle on tumors of the heart and great vessels follows.<sup>1</sup> This protocol, however, does not preclude the use of other histologic classifications.

#### AFIP Classification of Malignant Cardiac Tumors

Angiosarcoma  
Malignant fibrous histiocytoma  
Myxosarcoma  
Fibrosarcoma  
Leiomyosarcoma  
Rhabdomyosarcoma  
Osteosarcoma  
Synovial sarcoma  
Malignant schwannoma (malignant peripheral nerve sheath tumor)  
Malignant mesenchymoma  
Malignant mesothelioma  
Other

As with sarcomas in other sites, a variety of histologic patterns may be found. Although not included in the classification, lymphomas also are found in the heart.

#### E. Histologic Evaluation

Pathologists should grade the tumor and indicate the grading system used. Most malignant tumors of the heart are sarcomas. Necrosis of groups of cells and mitotic rates of greater than 5 mitoses per 10 high-power fields have been associated with reduced survival.<sup>1</sup>

#### F. Special Studies

Immunohistochemistry can be used to ascertain the histogenesis of a sarcoma or substantiate the diagnosis of mesothelioma. Generally speaking, mesotheliomas contain cytokeratins, which are usually lacking from sarcomas (see Thoracic Mesothelioma protocol). Transmission electron microscopy is also very helpful in the distinction of these tumor types. Myxoma, the most common benign tumor, has no distinctive immunohistochemical features.

#### Reference

1. Burke AP, Renu V. *Atlas of Tumor Pathology. Tumors of the Heart and Great Vessels*. 3rd series. Fascicle 16. Washington, DC: Armed Forces Institute of Pathology; 1996.

#### Bibliography

- Blondeau P. Primary cardiac tumors: French studies of 533 cases. *Thorac Cardiovasc Surg*. 1990;38(suppl 2):192-195.
- Burke AP, Cowan D, Virmani R. Primary sarcomas of the heart. *Cancer*. 1992;69:387-395.

- Burke AP, Rosado-de-Christenson M, Templeton PA, Virmani R. Cardiac fibroma: clinico-pathologic correlates and surgical treatment. *J Cardiovasc Surg.* 1994;108:862-870.
- Burke AP, Virmani R. Osteosarcomas of the heart. *Am J Surg Pathol.* 1991;15:289-295.
- Dein JR, Frist WH, Stinson EB, et al. Primary cardiac neoplasms: early and late studies of surgical treatment in 42 patients. *J Thorac Cardiovasc Surg.* 1987;93:502-511.
- Melo J, Ahmad A, Chapman R, Wood J, Starr A. Primary tumors of the heart: a rewarding challenge. *Am Surg.* 1979;45:681-683.
- Miralles A, Bracamonte L, Soncul H, et al. Cardiac tumors: clinical experience and surgical results in 74 patients. *Ann Thorac Surg.* 1991;52:886-895.
- Murphy MC, Sweeny MS, Putnam JB Jr, et al. Surgical treatment of cardiac tumors: a 25-year experience. *Ann Thorac Surg.* 1990;49:612-617.
- Reece IJ, Cooley DA, Frazier OH, Hallman GL, Powers PL, Montero CG. Cardiac tumors: clinical spectrum and prognosis of lesions other than classical benign myxoma in 20 patients. *J Thorac Cardiovasc Surg.* 1984;88:439-446.
- Ryan RE Jr, Obeid AI, Parker FB Jr. Primary cardiac valve tumors. *J Heart Valve Dis.* 1995;4:222-226.
- Tazelaar HD, Locke TJ, McGregor CG. Pathology of surgically excised primary cardiac tumors. *Mayo Clin Proc.* 1992;67:957-965.
- Turner A, Batrick N. Primary cardiac sarcomas: a report of three cases and a review of the current literature. *Int J Cardiol.* 1993;40:115-119.
- Verkkala K, Kupari M, Maamies T, et al. Primary cardiac tumors: operative treatment of 20 patients. *J Thorac Cardiovasc Surg.* 1989;37:361-364.