Recurrent Myxosarcoma of Left Atrium


The surgical treatment of cardiac myxosarcoma is reviewed with emphasis placed on palliative response to radiation therapy. A case of primary myxosarcoma of the left atrium in a 16-year-old girl is presented. The tumor was surgically removed in October, 1967. Recurrently myxosarcoma was removed in August, 1968, at which time involvement of the pulmonary veins and the pericardium was noted. Subsequent to postoperative radiation therapy, the patient remained symptom-free for more than three years. Death occurred 4½ years after the original operation from local and systemic recurrence of the tumor.

This is a report of a patient with left atrial myxosarcoma treated by surgical and radiation therapy with a brief summary of the literature on the subject, stressing the need to combine both types of therapy.

CASE REPORT

The patient, a 16-year-old girl, was admitted with the chief complaint of marked increase in shortness of breath during late 1967.

On physical examination, she had to remain sitting upright in bed. A blowing systolic nonradiating murmur, grade 2/6, best heard at the cardiac apex, was present. The cervical veins were distended to the angle of the jaw when the patient was sitting. The liver was percussed 3 cm below the right costal margin. Thoracic roentgenograms showed pulmonary edema and a relatively small cardiac silhouette. The electrocardiogram had ST changes compatible with ischemia, myocarditis or digitalis effect.

Selective pulmonary arteriography showed an enlarged main pulmonary artery and a large filling defect, 3 by 6 cm, within the left atrium. The left ventriculogram exhibited moderate reflux of contrast medium into the left atrium with a large filling defect outlined within the atrium which at times seemed to prolapse through the mitral valve.

The patient was taken to the operating room and via a right anterolateral thoracotomy, the left atrium was found to be enlarged by a mass that pushed down the intra-atrial sulcus. On total cardiopulmonary bypass, the left atrium was

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Please note that the image contains the following figure and caption:

**Figure 1.** a. Recurrent tumor nodules involving posterior aspect of the septal wall of the left atrium. Area of septal patch (P) is anterior to recurrent tumor. b. Closeup of tumor involving the posterior leaflet of the mitral valve.
then opened and a 5 by 4 cm smooth, somewhat indurated mass, attached by a narrow stalk to the atrial septum, close to the ostium secundum, was seen. The lower pole of this mass impinged on the mitral valve. The tumor was easily and apparently totally removed. Postoperatively, the patient did well.

The removed tumor showed both solid and cystic areas which contained mucoid material. Microscopic sections of the cellular portion disclosed spindle cells with nuclei which were large, hyperchromatic and moderately pleomorphic. Mitotic figures were identified. The histologic diagnosis was low-grade myxosarcoma.

The patient did well for seven months. Prior to the second admission, she became tired, her appetite decreased and a cough developed.

On examination, there was slight clubbing of the fingers and toes. A holosystolic nonradiating murmur, grade 2/6, was present at the left sternal border. The liver was palpable 6 cm below the right costal margin and was tender to touch. The thoracic roentgenogram showed bilateral basal infiltrates with increased pulmonary vascular markings at the apices. On the afternoon of readmission, she was taken to the operating room.

Through a right lateral thoracotomy a large amount of jelly-like fluid was found and removed from around the heart. On cardiopulmonary bypass, the left atrium was reopened; a large, firm irregularly-shaped tumor was found. It had infiltrated the septum where the stalk had been removed initially. The main tumor was removed with a major portion of the atrial septum. Biopsy proved that the tumor encroached upon the orifice of the right superior and inferior pulmonary veins and therefore could not be removed completely. The atrial septum was replaced with a prosthetic patch.

The pathologist interpreted the specimen to be an atrial myxosarcoma resembling, in general, the tumor previously removed. Myxosarcoma was present at the base of the resected specimen. After operation, a radiation course of 5,000 rads was given to the area of the heart over a period of 30 days.

Four years after the first operation, the patient noted a lump at the side of her left hip. This was excised and diagnosed as myxosarcoma. She was then given weekly vincristine and a five-day course of actinomycin D. In May, 1972, a mass over the right femur was removed and histologically shown to be myxosarcoma. The patient died on May 17, 1972.

Pathologic Findings

Autopsy was limited to examination of the heart. Two major sites of tumor were present, one in the left atrium and the other in the right ventricle.

Attached to the septal wall of the left atrium were two lobulated masses of tumor (Fig 1). A lobulated purple mass was attached to the atrial surface of the posterior mitral leaflet. The area of the atrial patch was a raised zone covered with fibrous endocardium. No tumor was identified in the immediate vicinity of the site of previous resection. However, tumor tissue which involved the posterior wall of the left atrium had penetrated through the full thickness of the atrial wall to invade the pericardial space in that vicinity. In the right ventricle, two distinct processes existed. One took the form of a large intracavity tumor within the infundibulum and subjacent part of the right ventricular sinus (Fig 2). In addition white tumor tissue was found to have invaded the walls of the right ventricular apex (Fig 3).

Histologically, the tumor tissue was composed of highly cellular but poorly differentiated stellate and spindled tumor cells lying in a myxoid stroma. The tumor was highly invasive and showed numerous mitotic figures and marked nuclear pleomorphism. The histologic picture was similar to that of the tumor tissues previously removed.

Discussion

In 1952, an intra-pericardial myxosarcoma was removed from a three-month-old boy.1 It was growing from the right ventricle and the stalk could not
FIGURE 3. Dilatation of the right ventricular infundibulum (RVI) is evident after removal of the mass seen in Figure 2. Extensive tumor invasion of the apex of the right ventricle and of the ventricular septum (VS) is evident. RV = right ventricle; LV = left ventricle.

be excised. The child died seven weeks later of pulmonary metastasis.

In 1972, there was a report of a myxosarcoma of the pulmonary valve initially removed with cardiopulmonary bypass. The patient was reoperated five months later because of recurrent symptoms and had the pulmonary valve and main pulmonary artery removed and reconstructed. There was no radiation therapy postoperation. Two years after the last operation the patient died of bronchogenic carcinoma of the left lung. There was no myxosarcoma present at autopsy (personal communication, Dr. C. J. Tatooles).

In a discussion of the role of radiation therapy in primary malignant tumors of the heart, eight cases were reviewed but no definite conclusions about the efficacy of radiation therapy could be reached. There were various cell types in these eight cases; they were not a homogeneous population of tumors.

REFERENCES


ANNOUNCEMENTS

Seminar for Intensive and Critical Care Personnel

The Department of Anesthesiology, Mount Sinai Medical Center, Miami, will present a Seminar for Intensive and Critical Care Personnel at the Americana Hotel, Miami Beach, June 27-29. A post-convention trip to the Grand Cayman Islands (BWI) is offered optionally. For information, write Critical Care Personnel, 1200 NW Tenth Avenue, Miami 33136.

Second Annual Seminar in Pulmonary Disease

Colby College/Thayer Hospital, Waterville, Maine will sponsor the Second Annual Seminar in Pulmonary Diseases at the college, August 24-27. For more information, write Mr. Robert H. Kany, Director, Division of Special Programs, Colby College, Waterville, Maine 04901.