Massive Pericardial Coelomic Cyst*
Diagnostic Features and Unusual Presentation

Stanley N. Snyder, M.D.**

Pericardial coelomic cysts are uncommon tumors that are usually detected on routine roentgenograms of the chest in an asymptomatic individual. Findings from physical examination are usually unremarkable. Needle aspiration, after echocardiographic diagnosis, may be diagnostic and therapeutic. Surgery may be indicated to exclude neoplasm or vascular abnormality. This case report presents the largest coelomic cyst yet described. Ultrasonic studies were useful in the correct preoperative diagnosis, and surgical excision relieved the patient's symptoms.

This report demonstrates the giant proportions that a coelomic cyst may reach and the symptoms that it may produce. Ultrasonic studies provided information suggesting the appropriate diagnosis.

CASE REPORT

A 49-year-old man was observed and treated for five years (June 1968 until September 1973) for atrial fibrillation and for elevation of the right hemidiaphragm diagnosed by chest roentgenogram. When the patient's symptoms of shortness of breath with mild to moderate exercise and difficulty in sleeping on his left side due to dyspnea had progressed significantly, he agreed to be hospitalized for further diagnostic and therapeutic evaluation. At that time, examination revealed absence of breath sounds in the right lower two-thirds of his lungs, controlled atrial fibrillation, and a liver which was palpable six fingerbreadths beneath the right costal margin. Chest roentgenograms (Fig 1) depicted progressive elevation of the right hemidiaphragm. Fluoroscopic examination of the chest revealed some motion of the elevated diafragm on full inspiration and a slight shift in the cardio-mediastinal silhouette toward the left. Ultrasonic studies (Fig 2) of the right side of the chest revealed a large volume of fluid almost totally filling the right chest cavity. Lung perfusion imaging (Fig 3A) revealed a large anterior filling defect occupying all but the apex of the right lung. Pulmonary function studies indicated a marked decrease in the patient's vital capacity. Findings from a liver scan (Fig 3B), upper gastrointestinal series, barium enema, and intravenous pyelogram were intrinsically normal but showed marked displacement of intraabdominal structures from their normal positions by the gigantic right intrathoracic cystic mass.

The preoperative diagnosis was probable pericardial coelomic cyst. At surgery a large cyst in the right lower portion of the chest, attached to the right side of the pericardium and without communication with the pericardial cavity, was removed. The cystic structure was multilocular, weighed 1,780 gm, measured 28 × 24 × 5.5 cm, and contained about 6,000 ml of a pale, yellow, clear, serous fluid. It was necessary to aspirate approximately 4,000 ml of fluid from the cyst to facilitate its removal. Microscopic examination showed "benign pericardial coelomic cyst."

The patient's postoperative course was uneventful. The electrocardiogram was unchanged, but the chest roentgenogram revealed reexpansion of the right lung and return toward the midline of the cardiome diastinal structures. The patient was discharged on the tenth postoperative day and has remained asymptomatic.

DISCUSSION

Over 200 cases of pericardial coelomic cysts have been described in the medical literature. They comprise about 7 percent of all mediastinal tumors. Several good reviews have described the clinical and diagnostic features, possible causes, differential diagnosis, pathogenesis, and surgery. In 1940, Lambert proposed the name of pericardial coelomic cysts, attributing their de-

*From the Presbyterian Intercommunity Hospital, Whittier, Calif.
**Cardiologist and Associate Clinical Professor of Medicine, Los Angeles County Hospital, University of Southern California Medical Center, Los Angeles.
Reprint requests: Dr. Snyder, 9209 Colima Road, Whittier, California 90605

Figure 1. Preoperative roentgenograms of chest. A (left), Five years before surgery (June 19, 1968). B (right), Immediately before surgery (Sept. 10, 1973).
development to failure of one or more primitive lacunae to join the other lacunae and to persist as a cyst lined with endothelium or mesothelium. Kindred (personal communication to Drash and Hyer) described the "pleural fold theory," and Lillie et al described the "ventral parietal recess theory."

The cysts usually do not become apparent until middle adult life. None has been malignant. Somewhat less than half the patients have been asymptomatic, with the abnormality detected on a routine roentgenogram of the chest. In the series reported by Wychulis and associates, 54 of 72 patients undergoing surgery for this condition at Mayo Clinic through 1969 were asymptomatic. Only a few patients have been considerably symptomatic, because the cysts are usually relatively small and do not increase in size or increase very slowly. Surgery is performed mainly for diagnosis and has only rarely been indicated for relief of incapacitating symptoms. Diagnosis may now be facilitated by the echocardiographic finding of fluid and by needle aspiration to reveal the characteristic appearance of the fluid. Large cysts can be aspirated to relieve symptoms or for identification of malignant cells; however, care must be taken, because some apparently fluid-filled cysts by ultrasonic B scan may actually be solid tumors. Only two patients had cysts containing up to 1,000 ml of fluid. Other cysts as large (up to 16.5 × 11 × 4 cm) have been described, but specific detail as to clinical presentation was not given. Atrial fibrillation has not been described previously, although palpitations and supraventricular tachycardia have been mentioned. The atrial fibrillation in our patient was believed to be coincidental. Seventy-five percent of these cysts have been located in the right anterior cardiophrenic

![Figure 2. Ultrasonogram of right side of chest obtained with transducer positioned 8 cm to right of midline at level of xiphoid process (X at top of dotted line) reveals large volume of fluid occupying most of right chest cavity.](image)

![Figure 3. A (left), Lung perfusion imaging with macroaggregated albumin labelled with technetium shows large anterior filling defect occupying all but apex of right lung. B (right), Liver scans with 2 millicuries of technetium sulphide. Upper right, During first six to eight seconds following injection, there is cardiac displacement toward left. Lower right, Later phase showing inferior displacement of liver.](image)
region. Studies by Cooper et al., by Jansson, and by Lam have described certain radiographic characteristics of the cyst.

Definitive diagnosis depends on exploratory thoracotomy, since the fluid removed by needle aspiration does not distinguish pericardial cysts from other benign cysts which may be located in the thoracic cavity. Usually noted is a unilocular cyst not continuous with the pericardial cavity and containing clear fluid. The wall of the cyst is usually made up of endothelium or mesothelium. Surgery has always been uneventful, and recovery has been satisfactory in all previously described cases. In nearly all cases, preoperative symptoms have cleared following removal of the cyst.

ACKNOWLEDGMENTS: I would like to thank Aziz A. Khan, M.D., for performing the surgery on the patient and Francis Y. K. Lau, M.D., for reviewing the manuscript.

REFERENCES
9 Klatte EC, Yune HY: Diagnosis and treatment of pericardial cysts. Radiology 104:541-544, 1972
11 Friday RO: Paracardiac cyst: Diagnosis by ultrasound and puncture. JAMA 226:82, 1973

Cardiac Rhabdomyoma Simulating Mitral Atresia*

Douglas D. Mair, M.D.; Jack L. Titus, M.D.;** George D. Davis, M.D.; and Donald G. Ritter, M.D., F.C.C.P.

Clinical, catheterization, and pathologic findings were recorded in a newborn infant with tuberous sclerosis and multiple cardiac rhabdomyomas that produced a clinical picture simulating mitral atresia and the hypoplastic left-heart syndrome. The clinical picture was due to a left atrial tumor that completely obstructed the mitral valvular orifice. Even if the diagnosis of left atrial tumor had been made, successful surgical correction was unlikely because of left ventricular rhabdomyomas, which produced severe subvalvular aortic stenosis and did not appear to be resectable. This case demonstrates the possibility that a hamartoma, such as a rhabdomyoma, occasionally can mimic the hypoplastic left-heart syndrome.

Although intracardiac tumors are rare during infancy and childhood, about 50 percent of the reported cardiac rhabdomyomas occur in children who die at less than six months of age. More than 50 percent of the patients with cardiac rhabdomyomas have pathologic evidence of tuberous sclerosis. Occasionally, a cardiac rhabdomyoma causes obstruction to blood flow, as in the report by Kuehl and associates of an infant with tuberous sclerosis who died during the fourth day of life from severe subvalvular aortic stenosis secondary to a rhabdomyoma situated in the left ventricular outflow tract and the report of a 14-month-old child with tuberous sclerosis and a rhabdomyoma obstructing the right ventricular outflow tract. Our report describes the clinical, catheterization, and pathologic features of a newborn infant with cardiac rhabdomyomas, one of which obstructed the mitral valve and simulated the findings of mitral valvular atresia. This child also had evidence of tuberous sclerosis at autopsy.

CASE REPORT

A male infant weighed 3.6 kg (8 lb 1 oz) at birth. There was no family history of tuberous sclerosis, seizure disorders, mental retardation, or congenital heart disease.

The patient was cyanotic and in respiratory distress immediately after birth. A chest roentgenogram taken at four hours of age showed a large heart. Cyanosis gradually increased in severity during the first day of life, and the infant also developed increasing tachypnea and tachycardia. At 22 hours of age, the child was transferred to this institution via air ambulance.

Examination showed a deeply cyanotic infant in severe distress. The heart rate was 180 beats per minute, and the respiratory rate was 90/min with subcostal retractions. Flush

*From the Mayo Clinic and Mayo Foundation, Rochester, Minn.
**Presently with the Department of Pathology, Baylor College of Medicine, Texas Medical Center, Houston.
Reprint requests: Section of Publications, Mayo Clinic, Rochester, Minnesota 55901