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Primary Malignant Tumors of the Heart

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Because of their rarity, primary malignant tumors of the heart continue to evade clinical preoperative diagnosis. In five cases of malignant heart tumor that were reviewed from the diagnostic point of view, the clinical symptoms and radiographic abnormalities were found to be similar to those produced by more common heart diseases. The diagnosis of malignant cardiac tumor should be considered in a patient with heart disease who has (1) an unusual cardiomediastinal silhouette, (2) an atypical cardiac calcification, and (3) clinical or radiographic cardipulmonary findings which do not conform to the expected behavior of the common forms of heart disease.

PRIMARY MALIGNANT NEOPLASMS of the heart are rare.[1] The clinical and radiographic patterns are various depending upon the location of the lesions within the heart and their tendency to metastasize. The subject, although previously reported by others,[2-8] has received rather little emphasis in the literature[9] because of the rarity of the lesions.

The purpose of this report is to present our experience in five cases of primary malignant cardiac tumors and to review the pertinent literature.

Reports of Cases

CASE 1. A 37-year-old Caucasian man consulted his family physician because of progressive exertional dyspnea of six months' duration. Physical examination revealed plethora of the face and neck, and distended veins in the neck and both arms. Immediate admission to hospital was advised, but the patient refused and was treated with diuretics and digitalis. A few months later he was admitted to hospital because of progression of symptoms. A roentgenogram of the chest at that time showed enlargement of the right atrium with an increased density, seen in the posteroanterior projection in this area (Figure 1). The pleura on the right contained a small amount of fluid. The

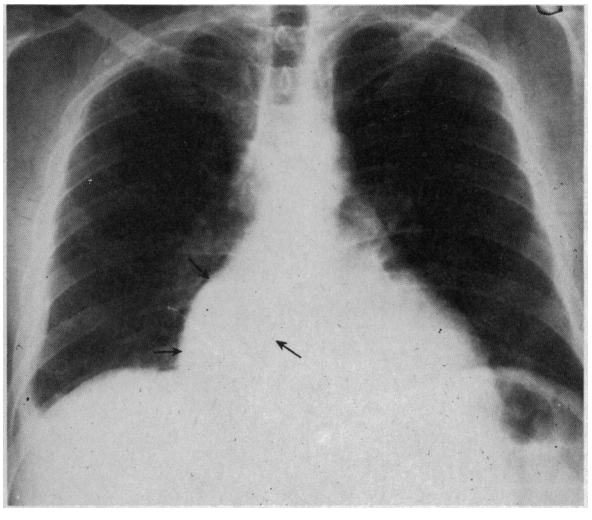

Figure 1. (Case 1.) Enlarged right atrium with increased density.

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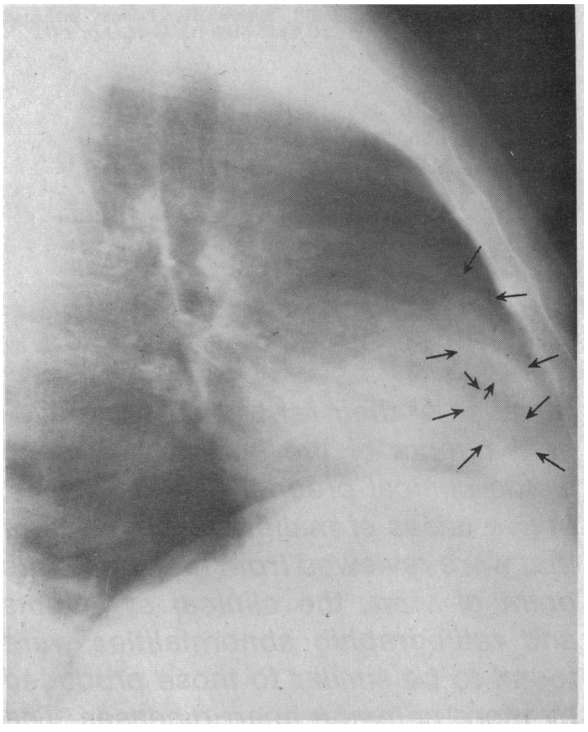

Figure 2. (Case 1.) Ovoid densities projecting over anterior heart. Small amount of fluid in the right pleura with blunting of the posterior costophrenic angle.

lateral projection revealed two irregular, oval anterior densities in the region of the right atrium with a prominence in the superior right atrial region, possibly the right atrial appendage (Figure 2). On thoracotomy, a tumor of the right atrium was found. The patient was referred to the University of California, San Francisco (UCSF) for definitive surgical operation. On admission, facial plethora and edema were present. The veins of the neck and arms were distended, but not those of the legs. Cavography revealed complete obstruction of the superior vena cava. At operation, a large intramural tumor was seen to involve the wall of the right atrium; it completely occluded the superior vena cava and partially occluded the inferior vena cava. The tumor infiltrated into the right ventricular wall and through the pericardium into the hilus of the right lung. Palliative resection was done to restore blood flow into the right atrium. Histologic sections of the tumor showed angiosarcoma. Postoperatively, signs of obstruction of the superior vena cava resolved and the patient received a course of radiation therapy to the mediastinum. Five months after operation the patient was receiving chemotherapy because of metastatic pleural lesions, but further evidence of vena caval obstruction was not apparent.

CASE 2. An 18-year-old Caucasian man consulted his school physician because of six weeks of progressive shortness of breath. After evaluation at another hospital, he was told that he had a heart murmur and pulmonary fibrosis. Two weeks later, progressive dyspnea, hemoptysis, edema of the legs, and mucoid sputum were noted. He was treated with digitalis, diuretics, and antibiotics, and then referred to UCSF for further evaluation. Previously, he had been healthy and had engaged in competitive sports.

On physical examination, the patient was pale and thin and appeared chronically ill. He was short of breath. Examination of the lungs revealed dullness and decreased breath sounds in both bases, but no rales. The pulmonic closure sound was accentuated and a moderate left parasternal dia-

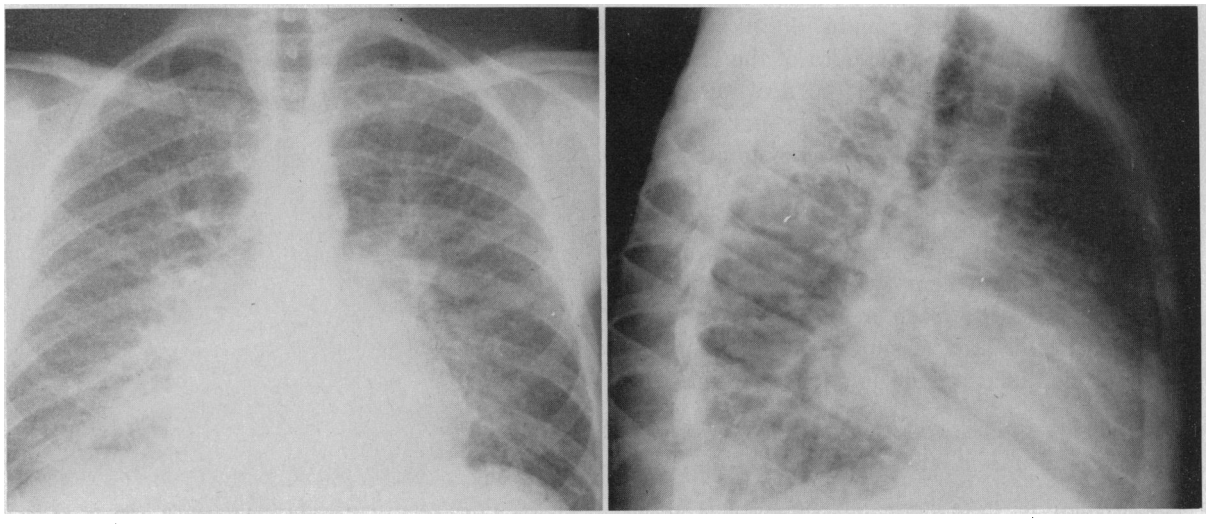

Figure 3. (Case 2.) Diffuse interstitial edema and left atrial enlargement.

stolic murmur and a holosystolic left parasternal murmur, which radiated to the apex, were heard. The sedimentation rate was 15 mm per hour. Changes in the electrocardiogram were "consistent with pulmonary heart disease." The initial chest film showed extensive interstitial pulmonary edema with left atrial enlargement and Kerley A and B lines (Figure 3). Cardiac fluoroscopy showed a mobile square calcification in the region of the mitral valve (Figure 4). It was situated in and above the valve and moved synchronously with the atrial contractions rather than with the valve excursions. A cineangiocardiogram showed that this calcification moved in association with an intracavitary left atrial mass. The catheterization revealed an increased arterial capillary wedge pressure with a 15 mm gradient over the mitral valve.

At thoracotomy a 3 × 4 cm large irregular, partly calcified tumor was found in the left atrium, which was adherent to the anterior leaflet and the annulus of the mitral valve, and invaded the left atrial myocardium. The tumor was resected partially to improve blood flow across the mitral valve. Histologic sections of the tumor showed a malignant mesenchymoma composed primarily of fibrous tissue with numerous mitoses and also of myxomatous, cartilaginous, and bone elements. An x-ray film of the chest on discharge from the hospital showed pronounced improvement of the interstitial pulmonary edema.

CASE 3. A 59-year-old Caucasian man consulted his family physician because of progressive pain in the chest, abdomen and bones, of one year's duration. The patient had no symptoms of congestive heart failure or weight loss. Physical examination showed a normal appearing man in no acute distress. Discrete firm masses were palpated in both axillae, in the inguinal areas, and in the left supraclavicular and infraclavicular fossae. The heart, lungs, and abdomen were essentially normal. The sedimentation rate was 41 mm per hour. Roentgenograms of the chest showed a bulge along the left upper border of the heart, which was first thought to be a left ventricular aneurysm (Figure 5). The bulge was higher than would be expected for a ventricular aneurysm, however, and appeared to involve the left atrium as well (Figure 6). An electrocardiogram appeared normal. On fluoroscopy of the heart the contour was seen to pulsate paradoxically. A biopsy of the right supraclavicular lymph node showed matastatic undifferentated carcinoma. The course for the next two months deteriorated pro-

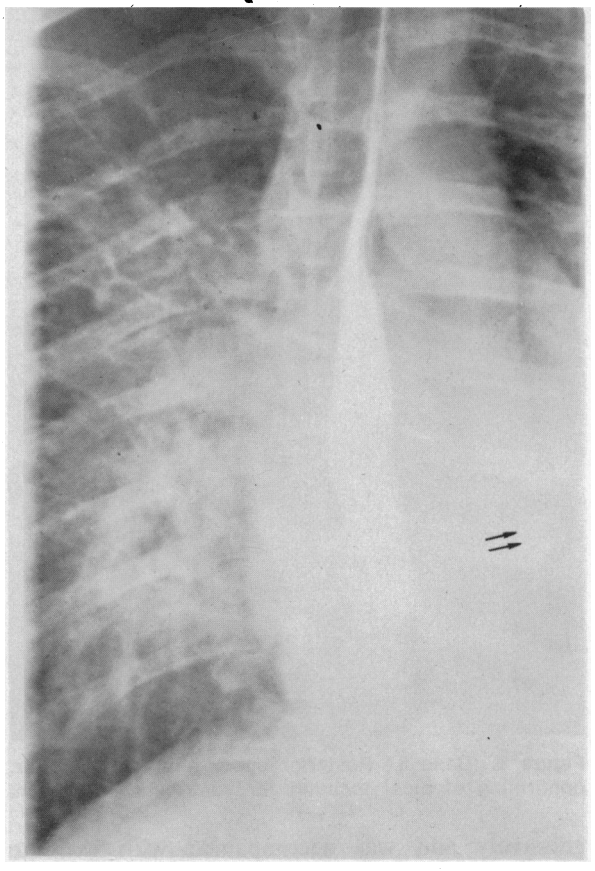

Figure 4. (Case 2.) Coarse square calcification in region of mitral valve (arrows), and horizontal left main bronchus indicating left atrial enlargement.

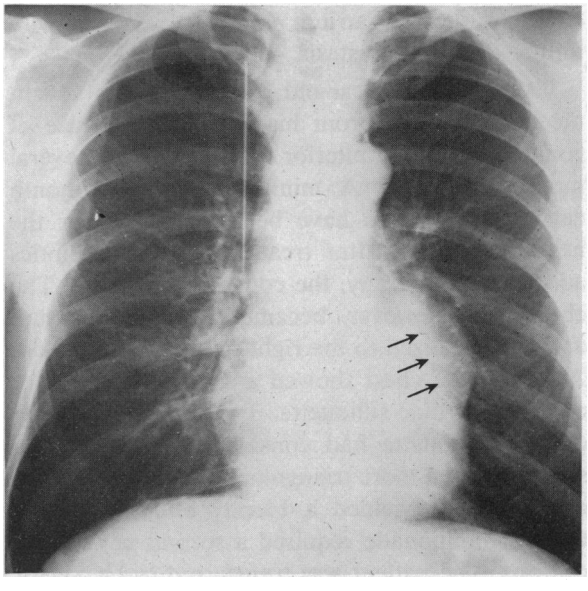

Figure 5. (Case 3.) Abnormal contour of left upper cardiac border, atypical location for left ventricular aneurysm. The linear density near the superior vena cava is an artefact.

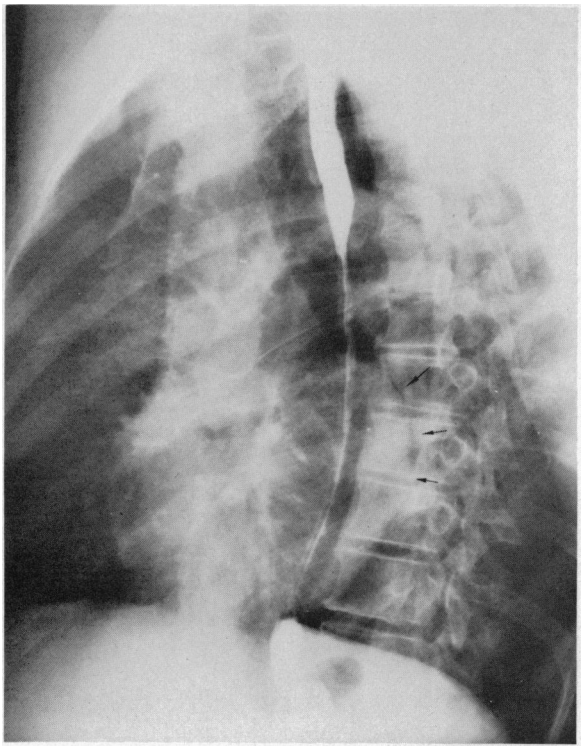

Figure 6. (Case 3.) Posterior upper abnormal contour, confirming atypical location for ventricular aneurysm.

gressively and was accompanied with extreme anterior right-side chest pain and epigastric pain, radiating to the spine. An exhaustive search for a pulmonary neoplasm was fruitless. The patient subsequently died and a postmortem examination at another institution revealed a rhabdomyosarcoma of the left atrium and left ventricle with diffuse distant metastasis.

CASE 4. A 50-year-old Caucasian man sought medical attention from his physician because of cough and right anterior chest pain of several months' duration. A miniature film one month earlier was said to have been normal. Over the next two weeks, after treatment with antibiotics and physical therapy, the cough disappeared. The chest pain, however, became pleuritic in nature and was localized to the right mid-thoracic region. A film of the chest showed a somewhat globular, enlarged cardiac silhouette. Two months later the cardiac silhouette had considerably increased in size and had a more triangular configuration. Pericardiocentesis yielded a bloody effusion. Symptoms of tamponade required a second pericardiocentesis. The patient was transferred to UCSF for operation.

On admission, the patient appeared lethargic and complained of severe back pain. The neck

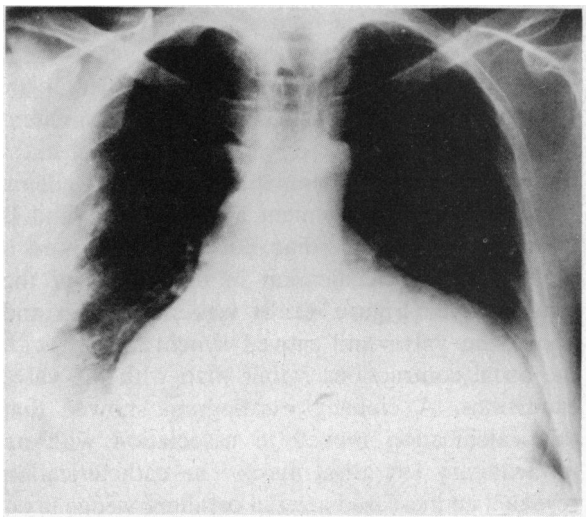

Figure 7. (Case 4.) Massive pericardial effusion suggested on admission film.

veins were flat and heart sounds distant. Sedimentation rate was 18 mm per hour. An electrocardiogram showed nonspecific ST and T wave changes consistent with pericardial effusion. A film of the chest showed right pleural effusion and massive cardiomegaly with a configuration compatible with pericardial effusion (Figure 7). A superior vena cavogram revealed a 65 mm distance from the outer aspect of the contrast medium to the outer aspect of the right cardiac border, presumably caused by a massive pericardial effusion. Pericardiocentesis and a pericardial exploration revealed 1,600 ml of blood in the pericardial sac. In the region of the right atrioventricular groove there was a soft nodular mass with blood oozing from its base. Histologic sections of the mass revealed angiosarcoma. The patient's postoperative condition became progressively worse and was marked by massive intraperitoneal bleeding and a series of cardiac arrhythmias, including final asystole. On necropsy, an angiosarcoma of the right atrium with metastatic growth in the lymph nodes, liver, spleen, and bone marrow was seen.

CASE 5. A 17-year-old Caucasian boy was admitted to another hospital because of weakness, weight loss, and cough of several months' duration. The physical examination at that time revealed distended neck veins, a regular tachycardia at 140 beats per minute, pronounced accentuation of the pulmonic closure sound and a left parasternal systolic ejection murmur. Electrocardiogram showed changes compatible with "right atrial strain and inferior ischemia but no signs of right ventricular hypertrophy." Roentgenograms showed

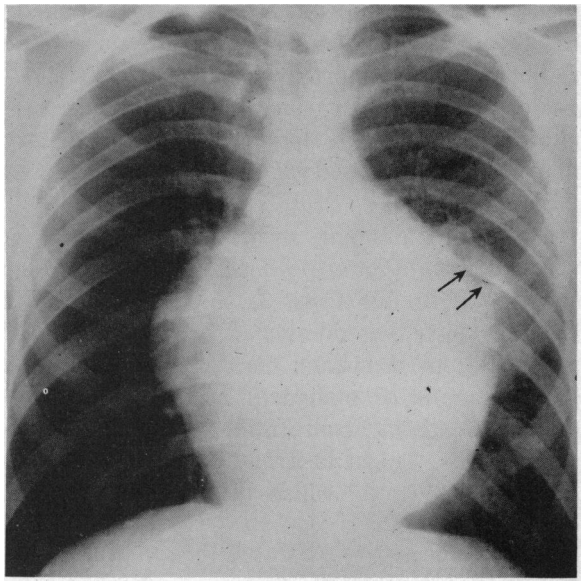

Figure 8. (Case 5.) Lobular polycylic contour to cardiac silhouette. Left pulmonary artery is displaced laterally (arrow).

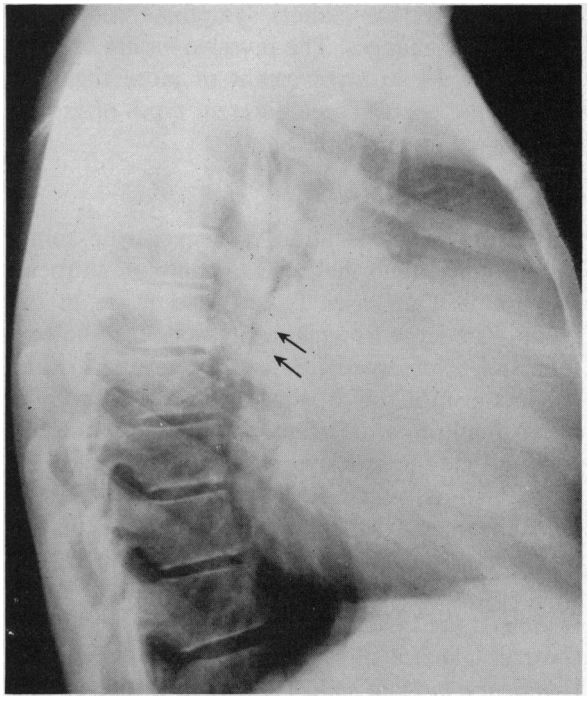

Figure 9. (Case 5.) Retrosternal space is filled in and there is associated narrowing of lower lobe bronchi (arrows). The heart appears posteriorly displaced.

irregular enlargement of the heart with polycyclic contour and filling in of the anterior mediastinum (Figures 8 and 9). The left lower bronchial lobe was compressed. The left pulmonary artery was pushed laterally and posteriorly. An angiocardiogram showed narrowing of the right ventricular outflow tract and the main pulmonary artery. The findings suggested either mediastinal tumor with cardiac involvement or primary cardiac tumor. The patient was referred to UCSF for further evaluation.

On admission the physical findings were essentially the same as those described above. Tomograms of the mediastinum revealed narrowing of both right and left main-stem bronchi. Subsequent fluoroscopy and angiocardiography revealed a large anterior mediastinal mass, which compressed the right ventricular outflow tract and the pulmonary artery. The interpretation at this time suggested mediastinal tumor with cardiac invasion. The sedimentation rate was 32 mm per hour. At operation a thick tumor beneath the epicardium enveloped the heart completely. It extended from the tip of the left ventricle around the great vessels and to the level of the right innominate vein. The tumor was 3 inches thick in the region of the right ventricular outflow tract. Sections revealed rhabdomyosarcoma or undifferentiated sarcoma. The patient was given 6,600 rad for 67 days with a 60-Co source. During the ensuing five months, progressive changes of pulmonary fibrosis developed. The patient died seven months after operation at another hospital of pulmonary insufficiency secondary to irradiation fibrosis.

Discussion

All primary malignant neoplasms of the heart are sarcomas.[10] They include angiosarcomas,[11] leiomyosarcomas,[12] rhabdomyosarcomas,[10] liposarcomas[13] and myxosarcomas. Malignant pericardial tumors have been reported.[14] It should be emphasized that secondary malignant tumors of the heart are about 20 to 40 times more common than primary neoplasms.[1] One common method of classification of primary cardiac tumors is by their location within the heart.[6] Histologic type bears no consistent relation to the site within the heart, except for angiosarcoma, which has a predilection for the right atrium.[11,15] Because of the subjective interpretation of the predominant cell-type in mixed tumors, and no universally accepted criteria for malignancy, detailed histologic classifications of malignant tumors are somewhat artificial.[16,17]

Far more important than the cell-type is the proximity of the tumor to vital intracardiac structures. Thus, a topographical classification allows a workable correlation with physiological aberra-

tions that produce clinical symptoms and roentgenographic changes. The invasive nature of some tumors results in involvement of more than one layer of the heart. Three different types of growth were found in our series:

Exophytic growth

Because of its location, any exophytic tumor may produce abnormal cardiac contours on roentgenograms. Our cases 1, 3, 4 and 5 are in this group. The large irregular, polycyclic cardiac contour and extension of mass into the anterior mediastinum in Case 5 suggests the diagnosis of mediastinal or cardiac tumor. Localized abnormal contours may be confused with more common entities, such as ventricular aneurysm, as in Case 3. On fluoroscopy the tumor may appear to pulsate paradoxically and change in contour with expiration and the Valsalva maneuver. Feist (1970) expressed the belief that cinefluorography without contrast material allows differentiation of ventricular aneurysm from solid masses. In a case of ventricular aneurysm he observed a time lag between the onset of systole and the often cyclic motion of the aneurysm. A solid tumor, in contrast, is characterized by rigidity or lack of cyclic deformity and by absence of a lag. The irregular cardiac contour in Case 1, with the ovoid densities projecting anteriorly over the region of the right atrium, suggests masses in this region. Tumors that extend to the pericardium may produce bloody effusions. Resulting tamponade, as in our Case 4, may be life-threatening and requires emergency operation.

Intramural growth

Intramural tumors may produce hemodynamic alterations by the production of conduction abnormalities and by preventing efficient myocardial contraction. Cardiac arrhythmias may result in cardiac failure. The patient in Case 4 died of progressively severe arrhythmias. An intramural tumor may be difficult to differentiate from cardiomyopathies that appear as diffuse or localized muscular masses, as for instance in hypertrophic myocardiopathy.

Intracavitary growth

Tumors with intracavitary extension may produce morbidity by obstruction to blood flow, mechanical hemolysis,[18] or embolization of thrombi or tumor cells.[4,19] In Case 2, the symptoms were clinically attributed to mitral stenosis. The evidence of severe interstitial pulmonary edema on the plain films and the data obtained on cardiac catheterization supported this diagnosis. At cineangiocardiography the unusual nature of the cardiac calcification was apparent. An intracavitary mass was demonstrated and a tumor was diagnosed preoperatively.

Sometimes a tumor, regardless of its location within the heart, never produces significant cardiac symptoms. In our Case 3, the patient's entire clinical course was dominated by bone pain that resulted from metastatic disease. Other frequent manifestations of malignant cardiac tumors include non-specific constitutional symptoms such as weakness, weight loss, and acceleration of the sedimentation rate,[2] which were present in most of our patients.

Roentgenographic Findings

Primary malignant cardiac tumors may produce abnormality detectable by chest roentgenograms, fluoroscopy, or angiocardiography. These findings depend upon alterations of the size or shape of the cardiomediastinal silhouette, presence of tumor calcifications, intracavitary masses, or secondary effects on extracardiac, intrathoracic structures. In four of our cases (1, 3, 4, and 5) there was abnormal cardiac configuration, the result of either pericardial effusion, exophytic growth from the heart, or mediastinal extension. In our second case, striking secondary pulmonary effects were related to obstruction at the level of the mitral valve.

Fluoroscopic findings included abnormal pulsations (Case 3) and the detection of calcifications (Case 2). Recording of the image on cineradiographs allowed a detailed study later of the calcified structures. The fact that location and motion patterns were inconsistent with mitral valve disease suggested the correct diagnosis of tumor. In one case (Case 5), angiocardiography showed extrinsic compression of the right ventricular outflow tract, but whether the mass was of cardiac or mediastinal origin could not be determined preoperatively.

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DECOMPRESSION SICKNESS*

WHY DO CIVILIANS, involved in sport SCUBA (self-contained underwater breathing apparatus) diving, appear to have a higher incidence of permanent damage following decompression accidents than do Navy divers? In a recent report from the University of Rhode Island (SCUBA Safety Report No. 3—"Nonfatal Pressure Related SCUBA Accidents"), statistics indicate that in a comparison of decompression accidents of civilians and U.S. Navy divers, the civilian sport divers had a 30 percent incidence of permanent damage following recompression therapy while for Navy divers the incidence was only 5.3 percent. Although the evidence is far from conclusive, it appears that the treatment of civilian divers is significantly less successful than it is for Navy divers.

The misinformation and general lack of understanding of decompression sickness by the sport diving public accounts for a large part of the delay in the diagnosis and treatment of this problem. One of the key factors in the successful treatment of decompression sickness is the time between the onset of symptoms and the initiation of treatment. Therefore, prompt diagnosis and immediate recompression are of paramount importance. However, confusion as to proper therapy, signs and symptoms, and the location of facilities capable of administering recompression therapy also has been responsible for serious time delays.

Signs, symptoms and complications of decompression sickness

Central Nervous System	*Cardiorespiratory*	*Extremities*	*Skin-Systemic*
Unconsciousness	Substernal pain	Pain	Rash
Spastic paralysis	Paroxysmal coughing	Numbness	Pruritus
Visual field defects	Tachypnea	Paresthesia	Pallor; low
Vertigo	Dyspnea (chokes)	Weakness	temperature; fever
Sensory loss	Shock	Aseptic Bone Necrosis	Fatigue
Bladder and bowel paralysis	Hemoconcentration	Joint destruction	All of the above

The accompanying chart is a brief compilation of possible manifestations of decompression sickness. Whenever cutaneous, cardiorespiratory and central nervous system symptoms are associated with a history of recent SCUBA diving activities, the possibility of a decompression accident should be suspected. Recompression is the therapy of choice and only a few specialized centers can offer you the proper facilities and personnel. Because of the expense of maintenance and operation, recompression facilities are almost exclusively run by the government. You may receive assistance and information by telephoning one of the following numbers:

Area	*Telephone Number*	*Area*	*Telephone Number*
Los Angeles/Long Beach	(213) 421-4741† or 590-2311†	Santa Barbara	(805) 966-3093†
San Diego	(714) 957-4439† or 295-3121†	Monterey	(408) 375-2278†
		San Francisco	(415) 986-5500†
		Bodega Bay	(707) 875-3596†

*Prepared by the California Medical Association Committee on Medical Aspects of Sports and Physical Fitness.

†U.S. Coast Guard Search and Rescue Operations' telephone numbers. The Coast Guard can provide transportation by helicopter or boat, supply information as to the nearest available recompression chamber, and put you in contact with a physician certified in submarine medicine to advise you in emergency situations.