CASE REPORT

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Primary Malignant Lymphoma of the Heart in Sudden Unexpected Death

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ABSTRACT: A case of sudden unexpected death resulting from asymptomatic primary malignant lymphoma of the heart in a previously healthy 64-year-old male is reported. Primary malignant tumor of the heart is extremely rare. The incidence of cardiac primary tumor ranges from 0.0017% to 0.06%. The signs and symptoms are usually those of congestive heart failure.

KEYWORDS: pathology and biology, death, cardiovascular system

Primary tumors of the heart, particularly malignant lymphomas, are extremely rare. A sudden unexpected death in a public place resulting from this condition was recently seen at the Westchester County Medical Examiners Office.

Case Report

A 64-year old, 190.5-cm (75-in.) tall, 86.2-kg (190-lb), moderately well-developed and well-nourished muscular black maintenance man had his usual breakfast with his wife. He went across the street to purchase cigarettes and suddenly collapsed onto the floor and died in spite of extensive cardiopulmonary resuscitation. According to his wife, he had collapsed about one month previously but had not sought medical help. He had otherwise been healthy and without any major illness or surgery. He had no children. His father had died of arteriosclerotic heart disease and his mother, at 90 years of age, was still living with no medical problems.

Postmortem examination revealed a markedly enlarged heart, weighing 650 g (Fig. 1). A large, slightly elevated, light reddish-gray, firm patch of tumor measuring 12 cm in greatest diameter was noted on the anterior surface of the right ventricle. Externally, the tumor involved the entire anterior wall of the right ventricle, sparing a portion of the apex, extending into the lower half of the right atrium and all layers of the heart. The tumor, in its thickest portion, measured 1.7 cm (Fig. 2). The cut surfaces revealed a firm reddish-grayish-white, rubbery tumor that had completely replaced the thickness of the heart in its central portion

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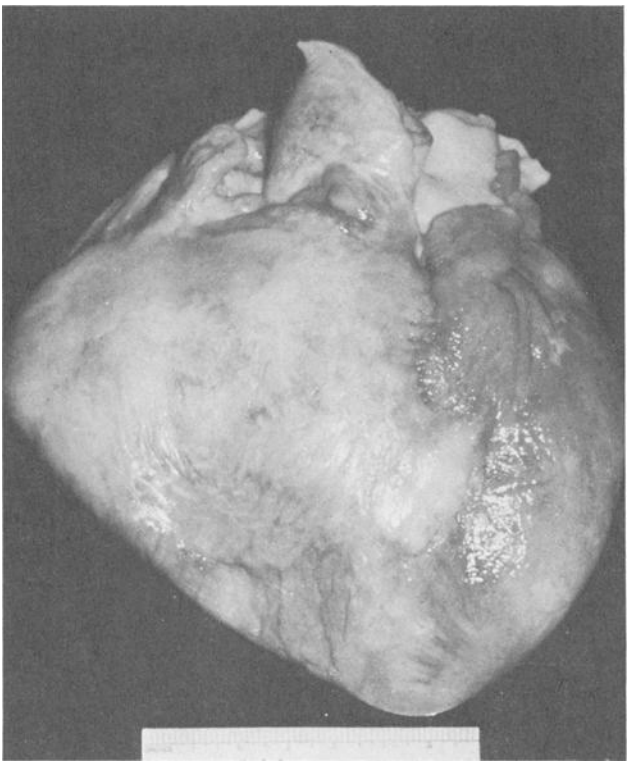

FIG. 1—*Anterior view of heart showing diffuse tumor involvement of right ventricle.*

and irregularly admixed with the myocardium peripherally. The right coronary artery was encased by tumor.

The remaining autopsy revealed no evidence of a primary tumor elsewhere or any evidence of enlarged lymph nodes. All arteries showed focal moderate to severe atherosclerosis.

The lungs weighed 1600 g in toto, and the sections revealed marked edematous change. No evidence of a space-occupying lesion was seen. The gastrointestinal system was unremarkable. The liver weighed 1800 g and the cut surfaces showed moderate congestion. The pancreas was unremarkable. The spleen weighed 110 g and sections showed normal parenchyma. Both kidneys weighed 450 g. A few small retention cysts were present on the cortical surfaces. The sections showed moderate congestion. The brain weighed 1300 g and the cut surfaces revealed no evidence of abnormalities.

Microscopically, the myocardium and epicardium of the right half of the heart were diffusely infiltrated by tumor cells with uniform, small round hyperchromatic nuclei and very scanty cytoplasm (Fig. 3). Occasionally, larger cells with vesicular nuclei with nucleoli, abundant cytoplasm, and frequent mitoses were seen. A moderate degree of desmoplasia was seen in some areas. Reticulum stain showed a moderate amount of reticulum fibers. The right coronary artery and its branches, nerves, and conduction system were also infiltrated by tumor (Fig. 4).

Discussion

The interesting aspects of this case are the rarity of primary malignant lymphoma of heart and sudden unexpected death. Historically, Boneti [*1*] and Morgagni [*2*] have both been

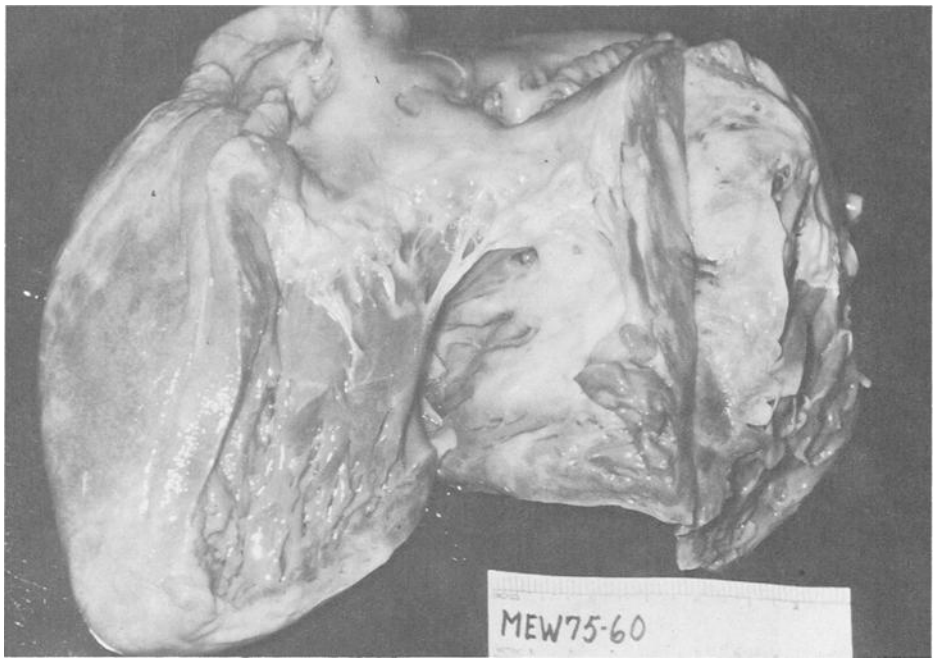

FIG. 2—*Inner aspect of right ventricle showing tumor extending to endocardium and a section through anterior wall revealing tumor infiltrating entire thickness.*

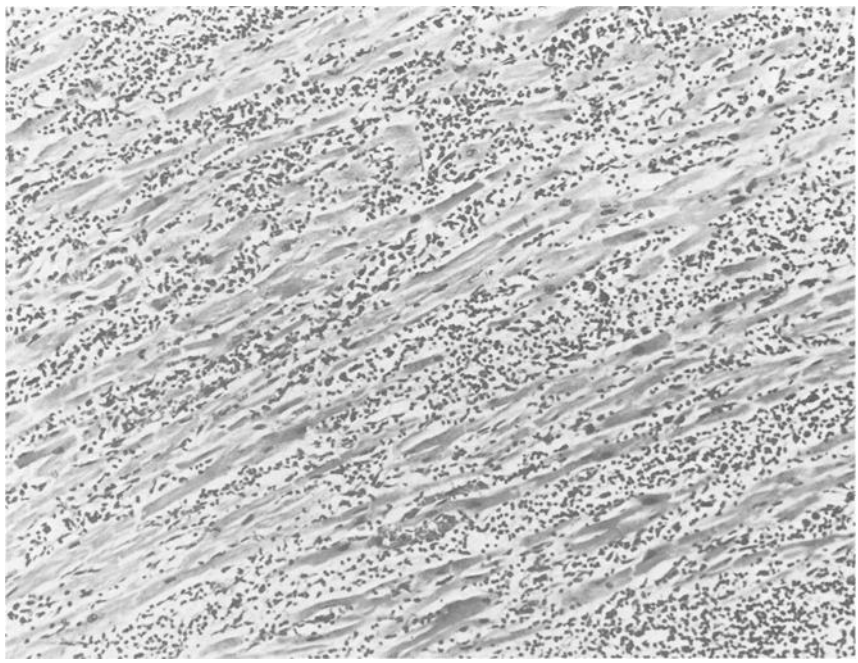

FIG. 3—*Microscopic view of tumor showing diffuse infiltration of tumor cells in myocardium.*

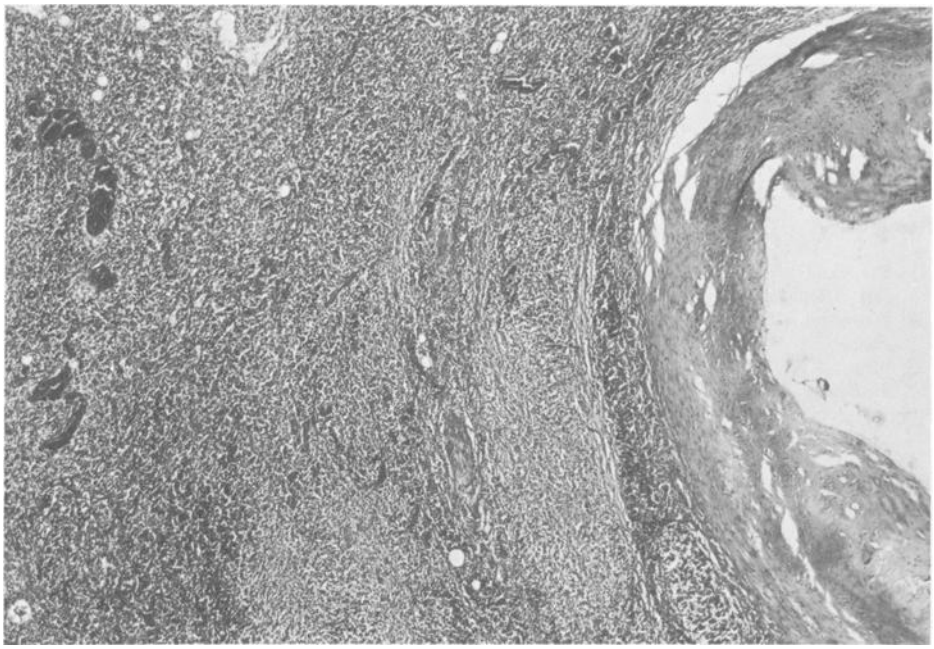

FIG. 4—*Microscopic view of right coronary sulcus showing tumor involvement of right coronary artery and nerve (in the middle of picture).*

mentioned as the first authors to describe primary tumors of the heart [*3,4*]. However, Albers [*5*] reported the first histologically proven authentic case of primary tumors of the heart (fibroma), and Bodenheimer [*6*] described the first primary sarcoma of the heart. Strauss and Merliss [*3*] observed the incidence at autopsy of benign and malignant tumors of the heart as being 0.0017%. Somers and Lothe [*7*] reported that the incidence of primary tumor of the heart was 0.06% in 6644 consecutive autopsies. Bruckner and Glassy [*8*] have seen only one primary malignant tumor of the heart in 11 131 autopsies.

General reviews of cardiac tumors have been published in the literature [*7–14*]. There have been about 400 reported cases of primary tumor of the heart, 47 of which were diagnoses as malignant lymphoma [*12–16*]. Among these were five cases of primary malignant lymphoma [*4,5,17,18*]. The most common, myxoma, accounted for 50% of the primary tumors of heart and occurred in the left atrium in 75% of cases [*16*].

In the cases of primary tumors of heart, the ages of the patients at the time of death range from 3 days to 79 years, the median age being 43 years. There was no significant sex difference [*9*]. The major clinical sign of cardiac tumors was intractable congestive heart failure [*12*]. The only other cases of reported sudden death in primary cardiac tumor were myxosarcoma [*11,19*], spindle cell sarcoma [*11,20*], and lymphosarcoma [*21*]. The lymphosarcoma case involved a 15-year-old male; death, though sudden, was not unexpected since subsequent investigation revealed he had had palpitation on exertion for 1½ years before his death.

The first case of sudden unexpected death resulting from primary malignant lymphoma of the heart is therefore reported.

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