

## PRIMARY CARDIAC KAPOSI'S SARCOMA

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Kaposi's sarcoma, or multiple idiopathic hemorrhagic sarcoma, was first described in 1872,<sup>1</sup> as a relatively rare disorder, occurring predominantly in elderly men of Eastern and Southern European descent. There are four types of Kaposi's sarcoma: sporadic classical type, endemic African type, allograft-associated type, and epidemic type associated with AIDS.<sup>2</sup>

In a large series of autopsies, more than 90% of the patients who had died of AIDS showed internal lesions of Kaposi's sarcoma, with the skin affected in only one case.<sup>3</sup> Next to the skin, the most common sites of involvement, in order of frequency, were the gastrointestinal tract, liver, lung, abdominal lymph nodes and heart.<sup>4</sup> Primary Kaposi's sarcoma of the heart is a rare vascular tumor with only a few previous reports.<sup>5-10</sup>

Primary angiosarcoma of the heart and primary Kaposi's sarcoma of the heart have undoubtedly been confused in the past.<sup>11</sup> We hope, by presenting this case, to further clarify the situation. Our patient, with four cardiac chamber tumors, is histologically a typical Kaposi's sarcoma case and fulfills the established histological criteria of the diagnosis of Kaposi's sarcoma.<sup>12-15</sup>

### Case Report

A 58-year-old man was admitted because of syncope and severe dyspnea. The chest radiography revealed a large heart with pericardial effusion, and the patient was referred to our hospital for admission. The patient had dyspnea and mild chest discomfort. On examination looked dyspneic without cyanosis. Heart rate was 120/minute and systemic blood pressure 100/80 mmHg with 20 mm paradoxical pulse. Heart sounds were distant and no murmur or rub was heard. The apex beat was impalpable. The liver was tender and palpable 5 cm below the costal margin.

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Investigation on admission revealed ESR 10 mm/hr; Hb 13.6 g/dL; white blood cell count  $8.3 \times 10^9/L$ ; urea, creatinin, electrolytes, SGOT, SSPT, CPK, LDH and bilirubin were all within normal limits. HLA typing was positive for HLA-DR5 and all investigations for HIV infection was negative. Sputum cytology was negative for malignant cells. Electrocardiographic studies revealed sinus tachycardia, abnormal Q wave in all leads except AVR and V6.

Transthoracic and transesophageal echocardiography (TTE and TEE) showed all four chambers to be enlarged, and a severely hypokinetic left ventricle with overall ejection fraction of about 30%. Large non-homogeneous masses were seen at LV and RV apices extending to septal and free walls. Large protruding masses also arose from the lateral walls of the right and left atria (Figure 1).

Chest radiography revealed a huge cardiomegaly with abnormal left contour and bilateral plural effusion. Non-contrast medium-enhanced CT scan revealed thickened pericardium with nodularity and thickening of myocardium in all four chambers, bilateral pleural effusion and dense calcification in left anterior descending and left circumflex arteries (Figure 2).

Cardiac magnetic resonance imaging (MRI) findings:

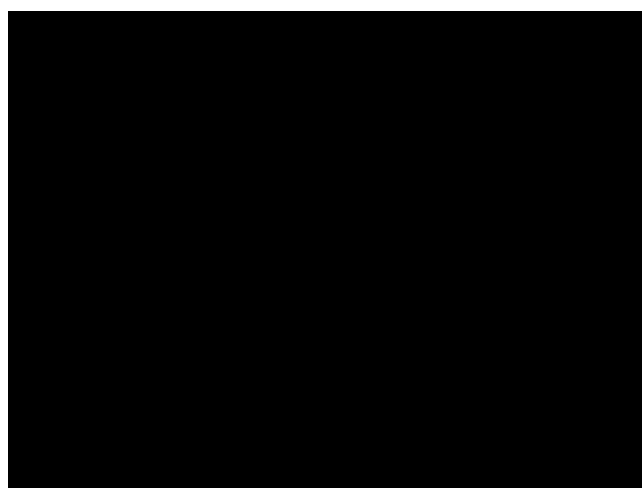


FIGURE 1. Echocardiography in four-chamber view, shows all four chambers are involved by the tumor.

TR/560, TE/16 coronal images showed enlargement of the heart, thickening of the entire heart's walls, and tumor was found in all four chambers. The largest tumor was in the left ventricle arising from the posterior wall. GE TR/600 TE/20 angle/40 revealed only a crescent bright signal indicative of blood flow in the left ventricle and bilateral pleural effusion. Cardiac tamponade was suspected, pericardiectomy and cardiac biopsy were performed.

On opening, there were more than five dark brown, nodular masses on the epicardium and pericardium and about 460 mL of bloody fluid was removed. Cytologic studies on pericardial effusion were negative for malignant cells and all microbiologic investigations were negative.

An open cardiac biopsy revealed a highly vascular tumor. Microscopic examination revealed dilated blood vessels, freely anastomosing capillaries and proliferating interstitial spindle cells. Some vascular spaces were incompletely lined by plump endothelial cells, but capillary channels were mostly well formed. Foci of siderotic pigmentation, lymphocytes, plasma cells and histiocytes were present.

In some areas, spindle cells dominated the histological picture. Within the spindle cell tissue there were slits and clefts bounded by spindle cells rather than obvious endothelium, and these clefts commonly contained erythrocytes, as Gottlieb and Ackerman<sup>16</sup> described in 1982. Spindle cells were ill-defined and had hyperchromatic nuclei. Reticulin stain showed the appearances to be like those reported by Choisser and Ramsey.<sup>5</sup> Most vascular spaces appeared to be completely surrounded by reticulin. The diagnosis of primary Kaposi's sarcoma was established.

### Discussion

Kaposi's sarcoma has recently come to the forefront because of its frequent occurrence in AIDS patients.<sup>17</sup> It is widely agreed that the tumor is multifocal, because there is no primary dermal focus that enlarges progressively, and there is spontaneous regression and some histologic examination revealing very early stages in late appearing lesions.<sup>14</sup> Primary Kaposi's sarcoma of the heart has previously been reported.<sup>5-10</sup> However, there are those who believe that some of these cases do not merit distinction from other forms of angiosarcoma on the basis of published photographs.<sup>11</sup> Hewer and Kemp<sup>18</sup> reported some cases of histologically typical Kaposi's sarcoma being reported as angiosarcoma. Perhaps the possibility of Kaposi's sarcoma was not actively considered because of the absence of typical skin lesion.<sup>10</sup> Glancy et al.<sup>11</sup> even suggested that cardiac involvement by Kaposi's sarcoma should not be diagnosed in the absence of skin lesion. This is true in our case as well. The skin lesion in our patient developed nine months later than the initial

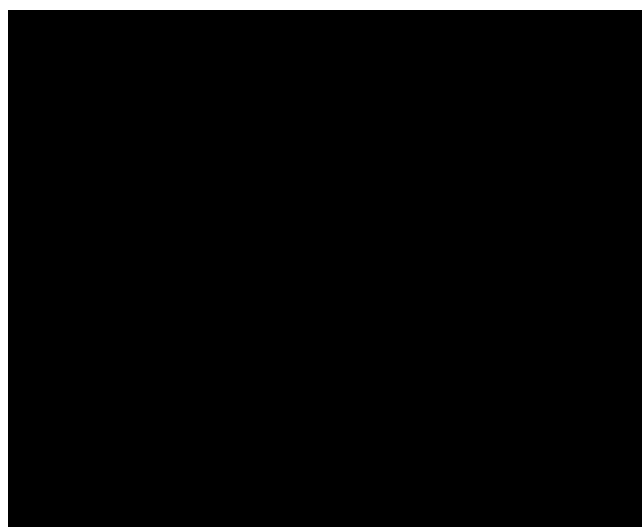


FIGURE 2. CT scan shows cardiomegaly, left ventricular mass, thickening and nodularity of pericardium plus bilateral pleural effusion.

primary cardiac tumor.

The cell of origin of Kaposi's sarcoma is the endothelial cell, as demonstrated by positive reactions with Ulex europaeus agglutination I and EN4, which reacts with both blood-vascular and lymphatic endothelial cells.<sup>19</sup>

Many cells are positive for factor VIII-related antigen.<sup>20</sup> Three distinct vascular, but not lymphatic, endothelial-cell-associated antigens are E92, OKM5 and HCl.<sup>14</sup> The spindle cells showed strong expression with E92, OKM5 and weak expression with HCl.<sup>21</sup> Use of polyclonal antibodies to type four collagen shows each tumor cell of Kaposi's sarcoma is surrounded by type four collagen.<sup>22</sup> We emphasize again that Kaposi's sarcoma occurs in the heart in the absence of, or primary to, typical skin lesion.

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