PRIMARY PERICARDIAL MALIGNANT MESOTHELIOMA: CASE REPORT AND LITERATURE REVIEW

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PERICARDIAL MESOTHELIOMA is a very rare disease. We present a Filipino male patient with this rare disease who had worked as a panel beater in a car repair body shop for 10 years in Saudi Arabia. The sudden onset of symptoms of cardiac tamponade in this patient, initially successfully treated with pericardiocentesis, progressing to a rapidly deteriorating stage of shock necessitated an emergency left thoracotomy and partial pericardiectomy which resulted in interval improvement of the patient's condition. The patient succumbed within 6 weeks of diagnosis.

Case Report

A 43-year-old Filipino man was admitted to Assir Central Hospital, Abha, Saudi Arabia, as a transfer from a district hospital with a complaint of dyspnea and chest pain of 3 days duration. He had been previously healthy and had worked as a panel beater in a car body repair shop in Saudi Arabia for 10 years. Physical examination revealed a very sick, orthopneic man, restless as well as tachypneic with a raised jugular venous pressure of 10 cm of water above the sternal angle. He had positive Kussmaul's sign. The apex beat was not palpable. The heart sounds were faint and there was summation gurgle rhythm. Brachial blood pressure was 100/60 mm Hg. Pulse rate was 120/min and paradoxical in nature. No pericardial rub or cardiac murmurs were heard. The liver was palpably enlarged, about 10 cm below the right costal margin.

Chest radiograph revealed gross enlargement of the cardiac silhouette with clear lung fields (Figure 1). Electrocardiogram showed sinus tachycardia, isolated premature ventricular beats with occasional ventricular couplets, and generalized low voltage. Electrical alternans were not recorded (Figure 2). Two-dimensional echocardiogram showed massive pericardial effusion compressing the right ventricle and interfering with its diastolic filling. The pericardium was thickened anteriorly. There was no intra- or extra-cardiac masses (Figure 3). All other laboratory investigations results, including complete blood count, random blood sugar, urea, creatinine, and liver function tests, were within normal limits except for elevated serum aspartate aminotransferase and serum alanine aminotransferase (SGOT and SGPT). Hepatitis B surface antigen (HBsAg) was positive. Blood group was 0 positive.

The patient was admitted into the coronary care unit. His central venous pressure was 24 cm of water, and it was kept above 30 cm of water with plasma and normal saline infusion to maintain adequate cardiac output. An echocardiography-guided pericardiocentesis yielded hemorrhagic fluid which was positive for malignant cells. Pericardiocentesis was done a couple more times with immediate temporary hemodynamic improvement. The patient later deteriorated with a systolic blood pressure of 60 mm Hg and a heart rate of 130/min. An emergency thoracotomy was performed on the fourth post-admission day,

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through a left anterolateral incision; during this period peripheral pulses were not palpable. A limited pericardectomy was then performed, anterior and posterior to the left phrenic nerve.

Histopathology of excised pericardium showed papillary structures arising from mesothelial surface. In addition, mesothelial changes in situ were also present, confined to a single layer of mesothelial cells. The cancerous cells were arranged in palisade, were larger than mesothelial cells, appeared frequently elongated or clubshaped, and were attached by the narrow end to the mesothelium. The nuclei were large and contained distinct nucleoli. Transitions between the in situ changes and the papillary structures were frequent (Figure 4). The malignant' cells were stained with Alcian Blue. A periodic acid Schiff reaction stain was also positive but was removed by prior diastase digestion. Immunoperoxidase stains for carcinoembryonic antigen and vimentin were negative.

Postoperative improvement allowed for a computed tomographic scanning of the chest which revealed no abnormal chest findings except for a thickened pericardium. Ultrasonography of the abdomen was negative. The patient was advised to return to his homeland for further management with a maintenance dose of 20 mg of prednisolone four times a day. He was reported dead in the Philippines about one month following discharge from our hospital.
Pericardial mesothelioma is a very rare neoplasm. In the rodent, it has been found to be extremely rare; one pericardial mesothelioma, having been found among 96 primary cardiac neoplasms was identified from 79,971 Fischer 344 (F344) rats used in chronic toxicity and carcinogenicity studies by the National Toxicology Program and National Cancer Institute for an overall incidence of 0.001%.1 It may mimic a left atrial myxoma when an intraatrial pendulous extension occurs,2 and it may present with a hemorrhagic pericardial exudate2 as in our patient. The patient may not have been exposed to asbestos.3 Asbestos exposure, however, is a predisposing factor in the development of the disease.4,5 Our patient denied any history of asbestos exposure, and asbestos fibers were not recognized in the histopathological specimen. Although viruses have been known to induce mesotheliomas,6 the only virus-related disease that our patient had was hepatitis-B virus (positive HBsAg).

The disease is usually found in the adult, although it has been reported in an infant.7 It usually presents insidiously. It may masquerade as a benign pericardial effusion8 or present as a recurrent cardiac tamponade7 as in our patient. It has been known to cause pericardial constriction,9,10 right-sided heart failure,10 or even present as systemic lupus erythematosus.11 It has been found in association with rheumatic heart disease12 and tuberous sclerosis.13 Hyaluronic acid has been found in the pericardial fluid.14

The diagnosis can be made by noninvasive methods (two-dimensional echocardiography or computed tomographic scan)15-17,8 or detected by gallium-67 scintigraphy.18 It can also be diagnosed by magnetic resonance imaging.19 The diagnosis in our patient was confirmed by histopathological examination which showed characteristic papillary clusters of malignant cells as well as in situ changes, characteristic of carcinomatous mesothelioma. One such case has been illustrated by Koss.20

The treatment of this disease is uncertain since there are very few cases reported in the literature and that cardiac complications occur very early in the disease. 15 Distant metastases are rare,21 perhaps due to the fact that the patients succumb to cardiac complications before the disease spreads to distant organs. 15 The disease has been known to cause a perforation into the atrial wall creating an intracavitary mass.10

The incidence of this disease in Saudi Arabia is unknown. Mesotheliomas have been reported in Saudi Arabia before, the most recent being the report of Khan et al.22; but these reports pertain to pleural and peritoneal mesothelioma, not pericardial mesothelioma. We do not know of any previous reports of pericardial mesothelioma from Saudi Arabia, hence the uniqueness of this case report.

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References