Angiosarcoma Presenting as Pleuro-pericardial Effusion

Col JJJ Falleiro, ^{SM*}, Lt Col Paramjit Singh⁺, Maj P Bhattacharya[#], Lt Col R Kaushish^{**},
Wg Cdr Nirmala Tutakne⁺⁺

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Introduction

Angiosarcoma is a highly malignant vascular tumor, which usually occurs in adults. It is a rare tumor comprising <2% of soft tissue sarcomas [1]. Common sites are skin, soft tissue, breast, bone, liver and spleen. We report a case of angiosarcoma of the pericardium.

Case Report

A 46 year old male came to medical attention in Jan 2003 with complaints of dyspnoea on exertion and pain upper abdomen and right side of chest of 2 days duration. He had anorexia, low grade fever, facial puffiness and signs of right-sided heart failure in the form of tender hepatomegaly and bilateral pleural effusion. ECG showed atrial fibrillation with low voltage complexes. Chest radiograph revealed a globular heart with cardiomegaly and bilateral pleural effusion (Fig 1). Ultrasonography confirmed pleural and pericardial effusion. CT scan of the thorax showed patchy pneumonia in the posterior segment of right lower lobe and peripheral region of left lower lobe and pericardial effusion. Mantoux was negative and ELISA for HIV was positive. Pleural fluid was exudative with predominance of lymphocytes. A clinical diagnosis of tubercular pleuro-pericardial effusion was made. He was started on anti tubercular drugs, steroids, digoxin and disprin. However, his condition continued to deteriorate. Pericardiocentesis was done and 80 ml of hemorrhagic fluid was aspirated which showed numerous RBCs, 80/cumm, WBCs 1017 specific gravity and 3.9 g/dl proteins. Injectable antibiotics were added to his treatment. ELISA for HIV repeated twice was negative. His condition deteriorated. Repeat echocardiogram showed re-accumulation of fluid along with thickened pericardium. He developed jaundice with deranged serum enzymes and impaired renal function (blood urea-106 mg/dl, serum creatinine – 2.4 mg/dl). Antitubercular treatment (ATT) was stopped. He was taken up for pericardiectomy on 11 March 2003. At surgery, there was no plane of cleavage between the pericardium and myocardium. It was not possible to remove the pericardium completely and there was profuse bleeding.

He had a stormy post-operative period developing anasarca with re-accumulation of pericardial fluid and inferior vena caval plethora. He was taken up for re-exploration. His condition continued to deteriorate and he expired three weeks after surgery.

Histopathology of the pericardial tissue revealed infiltration of pericardium by sheets of bizarre cells (Fig. 2). These cells also appeared to be lining vascular channels containing RBCs. The cells had scanty cytoplasm, hyperchromatic pleomorphic nuclei and frequent mitotic figures. There was a desmoplastic reaction. Necrosis and hemorrhage were present along with a sparse infiltrate of mononuclear cells. Immunohistochemistry was positive for CD31,

Fig. 1 : Chest radiograph (postero-anterior view) reveals a globular heart with cardiomegaly and bilateral pleural effusion

*Commanding Officer, 4033 Fd Amb, c/o 56 APO, *Classified Specialist (Pathology and Microbiology), #Graded Specialist (Pathology), Pune, **Classified Specialist (Surgery), MH (CTC), Pune, ++Medical Officer, 30 wing AF Stn.

Fig. 2 : H & E stained section shows sheets of bizarre cells showing cytologic atypia and high mitotic activity, lining the vascular channels

confirming the diagnosis of Angiosarcoma.

Discussion

Primary cardiac neoplasms are a rare entity with an autopsy incidence of 0.0017% [2]. Of the primary cardiac malignancies, angiosarcoma is the commonest. Over 80% of the cardiac angiosarcomas arise in the right atrium. Pericardial sarcoma with myocardial involvement is extremely rare. About 10% of the angiosarcomas of the heart have a primary in the pericardium [3].

Angiosarcoma of the heart usually presents in a young or middle aged individual with cough, haemoptysis, chest pain, raised central venous pressure and pericardial friction rub, similar to signs of constrictive pericarditis and cardiac tamponade. Thus antemortem diagnosis of angiosarcoma is rare [4].

Our patient presented with pleuro-pericardial effusion and signs of cardiac tamponade. The pleural fluid was an exudate with a predominance of lymphocytes. Repeated echocardiograms revealed no signs of mass in the right atrium. Since tuberculosis is the commonest cause of pleuro-pericardial effusion in our country, he was treated as such. When his condition deteriorated he was taken up for pericardiectomy for symptomatic relief. It was histopathology of his pericardiectomy specimen that revealed the diagnosis of angiosarcoma.

Trans-thoracic echocardiography has limited value in the diagnosis of intra-cardiac mass [5]. Trans-oesophageal echocardiography (TEE) and magnetic resonance imaging (MRI) have been found to be more sensitive [6,7]. However, due to the rarity of the disease, angiosarcoma was not suspected and TEE, MRI were not done.

Our patient had tested positive for HIV by ELISA initially. However, two subsequent ELISA tests for HIV were negative. False positive tests for HIV are known in malignancy.

The prognosis in a case of angiosarcoma is bleak with two-year survival of around 17% [8]. Most cases are treated with surgery along with chemo-radiotherapy with poor results. However, there is a case of angiosarcoma reported by Baay et al who was treated successfully with cardiac transplant following chemotherapy and radiation therapy [9].

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