Undifferentiated sarcoma of the mitral valve with secondaries in brain in a girl of 22 years

ABSTRACT

During a mitral valve replacement surgery in a girl of 22 years of age, it was accidentally discovered that the valve was destroyed due to a tumor and the histopathology and immunohistochemistry findings have proved it to be undifferentiated sarcoma. She was advised by the surgeon to go for chemotherapy. There was a delay of three months from the side of the patient to reach us and during that interval she has developed secondaries in the brain. This case is being presented here for its rarity.

KEY WORDS: Malignant tumors of the heart, brain secondaries from malignant tumors of heart

INTRODUCTION

Before the era of cardio-thoracic surgery, tumors of the heart were mostly diagnosed during postmortem. In contrast, cardiac tumors are detected during open-heart surgeries now.[1] The combined incidence of primary cardiac tumors is 0.02%[2] and that of secondary tumors is 1.23%. [3] The primary tumors of heart are mostly benign and among the benign tumors, the frequency is as follows: myxoma 29%, papillary fibroelastoma 8%, rhabdomyoma 5%, fibroma 5% and haemangioma 4% and other benign tumors account for 3% or less. Out of the malignant tumors that account for 37% of primary cardiac tumors, 2% are lymphomas and the remaining are sarcomas and the most common sarcomas are angiosarcoma, malignant fibrous histiocytoma, osteosarcoma and leiomyosarcoma. Some sarcomas are pathologically unclassifiable (undifferentiated) and they account for 0 - 24% of all cardiac sarcomas.[4] It is also a known fact that secondary tumors to the heart are more frequent than primary tumors approximately to the ratio of 100:1 to 1000:1.[4] The pediatric tumors of heart are less common when compared to the tumors of heart in adults. Our patient is 22 years old and has undifferentiated sarcoma arising from the mitral valve.

CASE REPORT

A patient of 22 years of age is a known case of rheumatic heart disease, was seen by cardiologist in November 2004 with the symptoms of shortness of breath on exertion. There was no history of paroxysmal nocturnal dyspnea orthopnea, hemoptysis, congestive heart failure, angina or syncope. The 2-D echocardiogram had shown LVIDD of 4.9 cm; LVIDS of 3.1 cm; EF 65%. The final report was: Chronic rheumatic heart disease with severe mitral regurgitation. Echocardiogram was repeated in November 2005 and the findings were almost same and in addition to those findings, they reported to have observed a “clot” under the mitral valve and the presence of this clot was also seen during trans esophageal echocardiogram. She was operated in January 2006. A mitral valve replacement with prosthetic was carried out. At the time of valve removal, the surgeon could see a tumor attached to the undersurface of the valve that was being reported as a clot in previous occasions. The patient recovered from the surgery as per their expectations. The tumor part was sent for histopathology study and the report was: high grade sarcoma. The report of immunohistochemistry was as follows: CD 31 positive in vessels and negative in neoplastic cells; CD 34 positive in vessels and negative in neoplastic cells; H and E stained slides show neoplastic cells [Figure 1] and these cells were positive for Desmin [Figure 2]; these cells were Vimentin positive and Pancytokeratin negative. Thus, our pathologist has confirmed the diagnosis as undifferentiated sarcoma. Immediately after surgery, a positron emission tomography scan was performed and this did not show any active disease in the body. She came to us again in May 2006 with right hemiparesis. Computerized tomography (CT) scan of the brain showed metastases in left frontal and parietal lobes of brain [Figures 3, 4]. Due to the size of these lesions and the surrounding edema, intracranial bleed was ruled out. She was given 45 Gy of external radiation to the whole brain and then one course of chemotherapy with actinomycin-D, cyclophosphamide and vincristine. When she came back for second dose
of chemotherapy in July, she could walk and clinically her CNS status was normal. The CT scan of the brain showed total regression of metastatic lesions, but only edema could be seen [Figures 5, 6].

**DISCUSSION**

Primary tumors from the heart are not that common when compared to secondaries. Lam *et al.* have mentioned that in
males, the most common tumor causing metastases in the heart was carcinoma of lung (31.7%), followed by esophageal carcinoma (28.7%) and lymphoma (11.9%). In females, carcinoma of the lung was most common at 35.9%, followed by lymphoma (17%) and breast cancer (7.5%). In children, the most common primary tumor arising from heart was rhabdomyoma (32%) followed by fibroma (21%) and both were benign tumors. The incidence of sarcoma from heart in children was approximately 20%. Symptoms due to cardiac tumors depend on its intracardiac location. Intracavitary lesions are associated with constitutional symptoms, obstructive symptoms, valvular symptoms and arterial embolization; myocardial lesions are associated with atrial or ventricular arrhythmias, symptoms and electrocardiographic signs of coronary artery involvement and cardiomegaly on X-ray. Sarcomas that are most common primary cardiac tumors cause shortness of breath, but they can also manifest chest pain, cardiac tamponade, palpitations and syncope. The regular investigations carried out in suspected case of tumor of heart are electrocardiogram, radiograph of chest, echocardiography, CT scan and/or magnetic resonance imaging (MRI), EKG usually shows sinus rhythm in cardiac myxoma where as mitral stenosis might cause atrial fibrillation and the chest radiograph of cardiac myxoma might reveal left atrial enlargement and signs of pulmonary hypertension. Cardiac fibroma infiltrating into myocardium might show changes of myocardial infarction on EKG and chest radiograph might show cardiomegaly and calcifications some times. Cardiac lipoma sometimes presents with ventricular tachycardia. Trans-esophageal echocardiography helps in the visualization of right and left atria, especially the appendages. MRI is preferred to CT scan as soft tissue delineation is better with MRI. Complete surgical resection of malignant cardiac tumors is only possible in less than 50% of patients and the mean survival time from diagnosis ranges from seven months to two years. Extra cardiac spread could be seen in 30% of sarcomas at the time of initial diagnosis. Secondaries were observed in bones, liver, jejunum, spleen and adrenal. Our patient had secondaries in the brain. As the tumors were not completely resectable, some surgeons have done heart transplants and could achieve increased survivals. Role of preoperative or postoperative chemotherapy / radiation is still debatable.

REFERENCES


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