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General Medicine

PRIMARY CARDIAC ANGIOSARCOMA:CASE REPORT OF A FATAL DISEASE

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ABSTRACT Angiosarcoma is a rapidly proliferating malignant neoplasm derived from anaplastic endothelial cells. It arises most commonly from soft tissue of head, neck, breast, liver, skin and deep tissue and is clinically unpredictable. Cardiac angiosarcoma is a rare and clinically challenging pathology, and this is an aggressive neoplasm with high rate of recurrence irrespective of treatment modality. Herein, we report a case of primary cardiac angiosarcoma with pulmonary metastasis who was asymptomatic at the time of presentation.

KEYWORDS : Cardiac Angiosarcoma, Cardiac Tumours , Lung Metastasis, Echocardiography.

CASE REPORT

A 59year old gentleman, presented to the general medicine out-patient department with the complaints of oral ulcers, blood tinged sputum and easy fatiguability of 2 months duration. The ulcers in the deeper part of oral cavity were painful causing difficulty in swallowing for both solids and liquids (more for solids than liquids). He started to cough out sputum mixed with blood since 2 months which was not associated with fever, chest pain, weight loss and bleeding elsewhere. He did not suffer from any significant past medical illness. He consumes non- vegetarian diet with a adequate calorie intake. He has never consumed any form of tobacco and alcohol.

On examination, he was conscious, alert, well built and well nourished with a body mass index (BMI) of 21kg/m2. He was normothermic and normotensive with regular rhythmic pulse of 80 beats per minute. He was comfortably breathing with a respiratory rate of 18 cycles per minute. There were no physical signs of bleeding diathesis. He did not have clinical evidence of anemia or nutritional deficiency.

There was no evidence of icterus, cyanosis, clubbing, lymphadenopathy, edema or markers of tuberculosis. Oral examination revealed healed scars more in the posterior surface of hard palate and there was no signs of bleeding from the healed ulcers at the time of examination.

His cardiac and respiratory examinations were unremarkable.

Laboratory evaluation revealed a mild normocytic, normochromic anemia with hemoglobin levels of 11gm/dl and with normal leucocyte count of 8980 cells/cu.mm. He had adequate number of platelets with a count of 3.19 lakhs/dl. Erythrocyte sedimentation rate (ESR) which is inflammatory marker was normal at 12mm at the end of first hour. Liver and renal function tests were normal. The plain chest x-ray (CXR) taken in PA view was remarkably abnormal with a ill-defined non homogenous radio-opaque shadows in the lower zone of right lung field obscuring right heart border which was compelling to consider possibility of right atrial enlargement. There were also well defined nodular opacities in bilateral lung fields [fig-1]. The sputum tested for acid fast bacilli (AFB) test and cartridge-based nucleic acid amplification (CBNAAT) test was negative.

Two-dimensional Transthoracic echocardiography (2D-Echo) showed focal thickening of right atrial wall measuring 2.5cm with extension into right atrial cavity. There was no evidence of extension in to inferior vena cava (IVC). Otherwise 2D-Echo was unremarkable [fig-2].

CT thorax (plain and contrast) was done, which revealed a enlarged right atrium with irregular frond like thickening of its lateral wall with maximum thickness 2.5cm. There was extension of this thickening into the right epicardial fat, which includes the differentials like myxomas, lipomas, fibromas, angiosarcoma and rhabdomyosarcoma. But there was no extension into right ventricular wall. Left ventricle and left atrium, pulmonary arteries, ascending aorta and superior vena cava (SVC) were normal. Infra-diaphragmatic inferior vena cava (IVC) was dilated (31mm). There were multiple bilateral well defined peripheral enhancing nodular lesions in the lung parenchyma ranging from 5mm to 15mm with peripheral ground glass halo in all the segments suggestive of hemorrhagic metastasis [fig-3]. Considering the cardiac and pulmonary lesions, possibility of right atrial angiosarcoma with bilateral pulmonary hemorrhagic metastasis was considered.

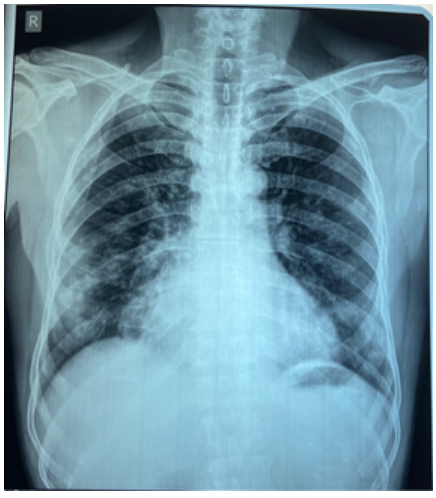

Fig 1: Chest X-ray PA view- ILL defined non homogenous radio-opaque shadows in the lower zone of right lung field

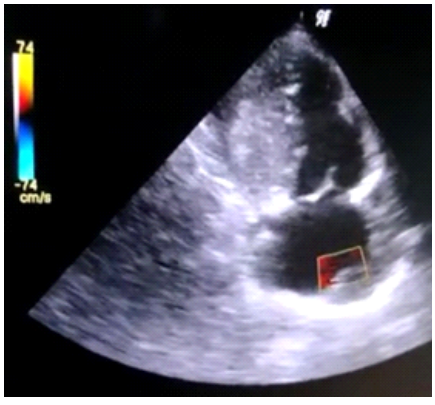

Fig 2: 2D ECHO-focal thickening of right atrial wall

Fig 3a & 3b: CT thorax (plain and contrast)-Irregular and frond like thickening of right atrium

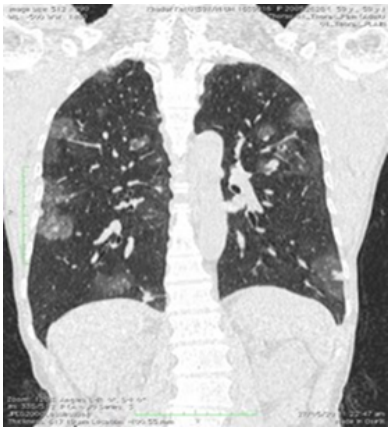

Figure 3a : Coronal view

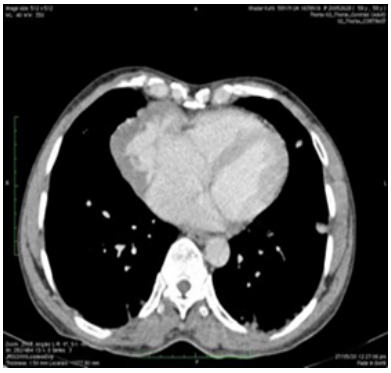

Figure 3b : Transverse view

DISCUSSION:

The occurrence of heart tumor was first identified by Realdo Columbus in 1559. Majority (75%) of the primary cardiac neoplasms are benign and of the remaining 25% of tumors which are malignant, cardiac sarcomas comprise 95% of cases. But, the first case of primary heart sarcoma was reported in 1934. Primary cardiac neoplasms are extremely rare, usually affects male more than the females, in a 2-3:1 ratio. The incidence of primary cardiac sarcoma is 0.0001 to 0.030 per cent based on a autopsy study [1]. It constitutes about 1 in every 500 cardiovascular surgical cases [2].

Nearly 90% of the cardiac angiosarcomas occur in the right atrium where it usually presents as a multicentric mass [3]. It is usually aggressive with a permeating growth within the surrounding myocardial wall, but can project into or fill the atrial chamber and sometimes can invade the vena-cava and tricuspid valve [4]. In less than 5% of cases left atrium and ventricles are involved [5]. Pericardium can be involved in right sided angiosarcoma. Angiosarcomas are usually hemorrhagic and necrotic, with a dark red or brown appearance and if it spread into pericardium, it usually produces a thick gray-black layer [6].

If a trans-thoracic echocardiography fails to detect the tumor, cardiac MRI or a trans-esophageal echocardiogram (TEE) is indicated. TEE has a high resolution (1-3mm) than MRI (5-10mm), but MRI can identify the tissue composition better. Pericardioscopic guided biopsy has a diagnostic value of 93% to 97%. Transvenous endocardial biopsy is less invasive but is often false negative [7]. Chest x-ray often reveals cardiomegaly (due to mass or pericardial effusion) with or without pulmonary congestion. Lung metastasis is very frequent (80%) and can be detected by CT imaging [8]. Despite the availability of modern imaging techniques, the diagnosis of cardiac angiosarcoma is usually late.

It was reported that the prevalence of primary cardiac tumors is between 0.001 to 0.28% in autopsy series[9]. In heart, angiosarcoma, a histopathological sarcoma type, accounts around for 30% of malignant primary lesions[10]. Angiosarcoma is most commonly found in the right atrium and it is usually asymptomatic or associated with nonspecific symptoms until the tumor becomes large which makes the diagnosis difficult and tardy[11]. Middle-aged adult males are the most affected although there is a study of series of primary cardiac sarcomas that reported a higher incidence in females[12].

In our case, diagnosis of angiosarcoma has not been confirmed by histopathology. However, with typical radiological features suggesting of right atrial mass lesion with pulmonary metastasis, the diagnosis of angiosarcoma is very certain. Although, the cardiac angiosarcoma is very rare, this can remain asymptomatic for longer period of time until complicated by invasion to vena cava or distant sites like in our patient. Identification of this rare condition all though incidental, high degree of suspicion is required as this condition has high mortality.

The prognosis of angiosarcoma patient is poor due to the aggressiveness of the lesion, as well as the high incidence of metastasis at the time of diagnosis, with a median survival of 6 to 11, There has been a 30-month survival report in patients with combined treatment of chemotherapy, radiotherapy, surgery and transplant. Surgical treatment remains the therapy of choice if no metastasis is evident and if myocardial resection is reparative. Chemotherapy as treatment for angiosarcoma is controversial as there is no randomised study comparing the evolution of metastatic diseases with and without chemotherapy. Multi modality therapy which includes surgery, radiation therapy, and chemotherapy was associated with improved survival [13]. However, there are a review of literature that demonstrated that combined treatment with doxorubicin and ifosfamide was effective in treating metastatic soft tissue angiosarcoma regardless of the primary site[14].

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