WCC 2016-115 : Cardiac Angiosarcoma of the Right Atrium: Two Cases: Puligundla Krishna Chaitanya

Background:

Cardiac angiosarcoma is a rare cancer that accounts for less than 10 percent of all resected primary tumors of the heart. Primary tumors of the heart themselves are found in less than 0.3 percent of autopsies. Angiosarcomas usually originate in the right atrium and are associated with a poor prognosis, since distant metastases are present in the majority of patients at the time of diagnosis. Because of the rarity of cardiac angiosarcoma, there are no standard applicable guidelines for treatment. Surgery, adjuvant chemotherapy, and radiation have been used. Despite aggressive treatment, the prognosis is poor, and death usually occurs within one year after the diagnosis. Early heart transplantation has been proposed but may not be possible because of the frequent presence of metastases at the time of the diagnosis.

Case Summary -1:

A 33-year-old previous healthy male presented to a local hospital with complaints of dyspnea of 4 month duration. No H/O cough/chest pain/palpitations. A chest X-ray detected cardiomegaly and bilateral pleural effusion. Computed tomography (CT) of chest showed findings suggestive of soft tissue density noted in Right atrium adherent to the wall showing patchy enhancement with contrast. A transthoracic echocardiogram (TTE) was performed and detected a lobulated right atrial (RA) mass of 6 x 4.5 cm with irregular borders. CT pulmonary angiogram was negative for pulmonary thromboembolism and a 6 x 3.2 cm mass along the anterior wall of right atrium with involvement of crista terminalis was detected.

Excision of the right atrial mass along with anterior right atrial wall followed by pericardial patch augmentation of right atrium was performed. Intraoperative findings were mass arising from right atrial wall just above tricuspid valve, infiltrating the right atrial wall. Tricuspid valve septum were free from tumor. Dense adhesions were found between cardia and pericardium, aorta and pulmonary artery.

Histopathology sections showed small interconnecting vascular channels lined by large polygonal epithelioid cells. The tumour cells showed vesicular nuclei with prominent nucleoli and abundant eosinophilic cytoplasm. Significant cellular pleomorphism with brisk atypical mitosis was observed. There were large areas of tumor necrosis. IHC report – Pan cytokeratin-negative, CD 31 - positive, CD34- negative. Features consistent with Angiosarcoma.

Post operative 2D ECHO – S/P right atrial excision and no residual mass in right atrium. Computed tomography (CT) of chest suggestive of heterogeneously enhancing nodular lesion along the anterior wall of atrium measuring 2x2 cm in size – S/O ? residual tumor and post operative changes in the mediastinum.

Treated with Inj Paclitaxel 175mg/m2 q3 weekly for 6 cycles . Post chemotherapy evaluation was done with cardiac MRI which showed features S/O poorly enhancing soft tissue density nodule of size 1.7 x 1.8 cm over the epicardial surface abutting the anterior wall- ? residual lesion / post operative changes. Irregular thickened pericardium along the anterior aspect / anterior wall of right atrium and right ventricle . Thin loculated pericardial effusion at right ventricular apex , measured 6mm in thickness. Patient is now asymptomatic and on regular follow up.

Case Summary -2:

45 year old female presented with complaints of pain in the right hip and leg since 8 months. H/O shortness of breath of grade III. No H/O trauma / cough/ chestpain/. MRI lumbo sacral spine S/0 altered signal intensity lesion in the right iliac bone in close proximity to right sacro-iliac joint- likely chondrosarcoma/ plasmacytoma/ metastasis.

Biopsy from soft tissue componenet of right iliac bone – S/O mesenchymal neoplasm. IHC showed tumour to be CD31 and CD34 positive favouring Angiosarcoma. 2D ECHO showed a large mass within the right atrium compressing the right ventricle. Interventricular septum is hypokinetic with mild LV dysfunction.

HRCT chest showed features suggestive of a large mixed dense lesion in the anterior mediastinum causing mediastinal shift to left. The lesion measuring 10.1 x 10.4 x 8.9 cm showed multiple hyperdense foci within suggestive of haemorrhage. Tumour was found to involve the adjacent pericardium, encasing aorta, pulmonary artery and SVC. Superiorly it was extending into manubriosternum joint and inferiorly upto the diaphragm. Multiple hypodense lesions in the liver were detected suggestive of ?cardiac sarcoma with liver metastasis. Bone scan showed increased tracer uptake in right ilium, focal uptake in right frontal skull, manubrium, D11 vertebra and left 3rd rib anteriorly. The patient was treated with 6 cycles of Inj Paclitaxel 175mg/m2 q3 weekly for 6 cycles. Post chemotherapy evaluation with CECT chest and abdomen showed lobulated heterogeneous enhancing mass lesion arising from right atrium extending into the anterior mediastinum. Multiple liver and bone metastasis detected. Bone scan showed new

bony lesions compared to the previous scan suggestive of Progressive disease. The patient is currently on second line chemotherapy with Inj. Liposomal Doxorubicin 50 mg/m2 q 3weekly.