

Primary cardiac angiosarcomas- A case report

Randhawa PK

ABSTRACT

Primary cardiac tumors are a very rare occurrence and their incidence was found to be only 0.0017% in one of the largest consecutive autopsy series conducted by Straus and Merliss. Only 7.5% to 8% of primary cardiac tumors are malignant of which angiosarcomas form a part.

Angiosarcomas usually occur between 3rd to 5th decades of life and are characterized by rapidly proliferating, extensively infiltrating anaplastic cells derived from blood vessels that line irregular blood-filled spaces. They are aggressive tumors and the rate of tumor-related death is high with a mean survival of 6-12 months.

This is the report of a middle aged woman with primary cardiac angiosarcoma. This case highlights an impressive survival period of 4 years after diagnosis, without resection of the primary tumor and the role of multidisciplinary management in improving the prognosis.

Keywords: cardiac tumors, angiosarcoma,

INTRODUCTION

Primary cardiac angiosarcomas are a rare entity.¹ It accounts for around 33% of all primary malignant cardiac tumors, which make up approximately 25% of the primary cardiac tumors. The diagnosis is delayed for the reason of its symptoms being non specific and very low incidence. Due to their rapid and aggressive behaviour; the tumors have commonly metastasized by the time of diagnosis, resulting in a poor prognosis. This tumor shows preference for male gender and the most common site of occurrence is found to be the right atrium.

Suitability of therapies of primary angiosarcoma of heart has remained controversial so is the increase in the survival period in regard to the adjuvant therapy besides surgery being regarded as treatment of choice. Two schools of thoughts prefer to dominate on each other. So far a study with highest level of evidence is awaited to establish the benefits or losses of aiding adjuvant therapy.

CASE REPORT

A 40 year old white woman presented to the hospital with dizziness and presyncope. Previously, she had a several month history of nonspecific chest discomfort that till now had been diagnosed as pneumonias or pericarditis. On presentation, she was noted to have a ventricular tachycardia and was placed on amiodarone.

ACT scan was performed which showed a mass like thickening of the right atrial wall that was suspicious for a highly infiltrating neoplasm. There was a small pericardial effusion and multiple pulmonary nodules. A colored Doppler Scan showed mild mitral and tricuspid regurgitation and a mildly dilated right atrium. An MRI revealed a 5.4 x 6.2 cm irregularly shaped mass in the right interventricular groove, which appeared highly vascular. All other chambers of the heart were normal.



Fig.1 Heterogeneous mass involving the lateral aspect of the right atrium, which extends to involve the inferior aspect of the superior vena cava and the interatrial septum, measures approx 2.6x4.8 cm

Subsequently she underwent a right video-assisted thoracoscopy and thoracotomy with wedge resection biopsy from the right lower lobe. The pathology of the mass revealed a poorly differentiated angiosarcoma positive for CD31 and CD 34. It was negative for keratin (AE1/AE3). Scattered tumor cells stained for p53. The tumor showed high proliferation activity by Ki-67 immunostain.

Her treatment was begun with Paclitaxel but due to severe hypersensitivity reactions to Paclitaxel and also Taxotere, she was discharged on weekly gemcitabine with aggressive premedications to minimize hypersensitivity. She continued with chemotherapy with intermittent holidays and liposomal doxorubicin.

After a couple of years she presented with various constitutional symptoms like fevers, night sweats, fatigue and increased pleuritic chest pain. Work up showed progression of the disease. This was followed a few months later by lower back pain radiating to her left leg, and a history of multiple UTIs. An MRI showed multiple metastatic lesions to the thoracic and lumbar spine. However, there was no acute spinal cord compression. She then received

3000 cGy radiation to the lower spine for about a year.

The back pain and paresthesias continued and a few months later she presented to the emergency department with abdominal pain, watery diarrhea and severe rectal bleeding. This was 3 days after the last dose of her radiotherapy. An echocardiography performed demonstrated an EF = 57% with some abnormality of septal motion, increase in RV size and mild MR and TR. Her EKG showed a first degree A-V block. And an X-Ray showed stool in the entire colon with non obstructive bowel gas. The symptoms were thought to be an exaggerated side effect of the radiation she received. She was treated with a stool softener, pantoprazole and kept NPO.

This gave her some relief but three months later, she again presented with abdominal discomfort and incomplete evacuation of the bowel or bladder. There was tenderness in the left lower quadrant of the abdomen without guarding. Labs showed a rise in WBCs, BUN and Cr. A CT scan showed right-sided hydronephrosis with perinephric fat (Fig.2). There was stool seen throughout the colon (Fig.3). There was no evidence of obstruction or intraperitoneal free air. The patient was managed conservatively and discharged.



Fig.2: CT scan showing perirenal fat



Fig.3: CT abdomen showing stool throughout the colon.

Only after three days she returned with continuing abdominal pain, fevers and chills. Her abdomen was diffusely tender with guarding. Cardiac examination showed a normal S1 and S2 without murmurs or JVD. An X-Ray showed free air under the diaphragm and a diagnosis of a perforated recto-sigmoid and intra-abdominal sepsis was made. She underwent Hartmann's Procedure the following day.

During her stay at the hospital in the following days, she was noted to have bradycardia, with her heart rate between 46-52 on an average and dipping to the 20s on several occasions. Subsequently, a pacemaker had to be implanted for stabilizing the heart rate. She was kept under observation in the hospital for the next few days and finally discharged on 17 October. After that, her condition however continued to deteriorate and she couldn't fight the disease much longer.

DISCUSSION

Being an extremely rare occurrence, primary cardiac angiosarcomas have, for long, drawn attention to them. These tumors are more common in men; male: female ratio of 3:1.²

and the most common site of occurrence is found to be the right atrium.³ In our case, the tumor was typical in arising from the right atrium and for the age of presentation (between the 3rd and the 5th decade). However, our patient is a female, which is the lesser affected gender for this tumor.

In our patient, the tumor had already metastasized to the lung by the time of detection. The presenting symptoms were those of chest discomfort, dizziness and presyncope. Though there were complaints of bilateral swelling of feet about 2 years down the course of the disease, no signs of overt heart failure were noted - the JVP was not found to be raised during any of the examinations and no murmurs were appreciated. However, the 2nd degree heart block with intermittent 3rd degree block and the resulting bradycardia led to the need for a pacemaker implantation.

Owing to the aggressive behavior of angiosarcomas, treatment options are limited. However, Complete resection is the treatment of choice.⁴ The role of chemotherapy is not well established. However, chemotherapy is recommended after surgical resection, owing to the high rate of tumor metastasis.⁵ Liposomal doxorubicin (PLD) has been shown to be useful.⁶ In our case, gentamycin and liposomal doxorubicin were the chemotherapeutic agents used and radiotherapy was used to manage the tumor metastasis. It is worthy of mention that there was no surgical procedure done on the heart itself.

CONCLUSION

Advent of enhanced imaging techniques aided with accurate understanding of these tumors it has been made possible to prolong the survival

period in these cases. Clinicians should always keep this rare tumor in one of their differentials when a right sided heart mass is detected, especially if arising in the posterior wall and unattached to the interatrial septum. If, fortunately, the tumor is recognized before metastasis, surgery remains the treatment of choice. Multidisciplinary treatment may help provide greater hope for survival in these patients.

AUTHOR NOTE

Puneet Kaur Randhawa.

Email: puneet.gemini.16@gmail.com

Yale New Haven Hospital, Connecticut, USA.
(Department of Cardiology)

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