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## CASE STUDY

### AN INCIDENTALLY DIAGNOSED RARE CASE OF PERICARDIAL FIBROMA PRESENTING AS CARDIOMYOPATHY, CARDIAC FAILURE AND VENTRICULAR TACHYCARDIA IN A ELDERLY MALE ALCOHOLIC DIABETIC COPD PATIENT

<sup>1</sup>Rajeev, T., <sup>\*</sup><sup>2</sup>Himanshu, V., <sup>1</sup>Mrinda, J. and <sup>1</sup>Anurag, A.

<sup>1</sup>Department of Pulmonary Medicine, SRMS-IMS, Bareilly

<sup>2</sup>Department of Anaesthesia, SRMS-IMS, Bareilly

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#### INTRODUCTION

Primary cardiac neoplasms are a rarity in clinical practice (0.02-0.56%) (Lam *et al.*, 1993). Pericardial neoplasms account for a very small portion of cardiac neoplasms and are usually due to neoplastic involvement of pericardium by direct invasion or by metastasis. Primary pericardial tumors are rare (0.001-0.007%) (Meng *et al.*, 2002) and are usually benign, the most common being pericardial cyst followed by teratoma, lipoma, fibroma etc (Patel *et al.*, 2004). The spectrum of clinical presentation is highly variable and at one end, may range from being asymptomatic to presenting with vague non-specific symptoms like chest pain, dyspnea, palpitation, fever, weight loss and on the other end may present with cardiac failure, arrhythmias and sudden death (Lamba and Frishman, 2012). Chest x ray and transthoracic echocardiography have a limited role in diagnosing pericardial tumors. CT & MR imaging play an important role in detection, characterization and staging of pericardial tumors (Kaminaga *et al.*, 2003).

#### Case report

61 yrs old male presented to us with cough & expectoration on and off for 6months duration. Fever since 3 days which was intermittent, high grade associated with chills and rigor.

**\*Corresponding author: Himanshu, V.**

Department of Anaesthesia, SRMS-IMS, Bareilly.

#### ABSTRACT

Pericardial fibroma is a very rare occurrence in clinical practice (< 0.0001%) (Lam *et al.*, 1993). They may be asymptomatic or can present as case of cardiac failure or even lead to sudden death (cardiac arrhythmias). Many a times they are encountered while patient is being evaluated for other reasons (Meng *et al.*, 2002). We present here, a incidentally diagnosed and successfully managed case of 61 yr old male COPD patient with pericardial fibroma (known diabetic, chronic smoker and chronic alcoholic) who presented to us in shock and respiratory distress and who during his hospital stay had various cardiac manifestations (cardiac failure, ventricular tachycardia, sudden cardiac arrest) and respiratory complications (bilateral pneumothorax).

Chest pain and breathlessness since 1 month but had increased in last 24 hours due to which the patient was brought to the hospital. Patient was a known case of type 2 diabetes & COPD on irregular oral medication. History of smoking of more than one pack /day for last 20 yrs and patient was a chronic alcoholic – around 1 quarter/day for 20 years.

His examination revealed pulse of 87/min, blood pressure - 76/50 mm Hg in right upper limb in supine position, respiratory rate - 35/min, thoraco-abdominal & accessory muscles of respiration being used. Bilateral pedal edema (pitting type) and cyanosis was present. SpO<sub>2</sub> =70% off O<sub>2</sub> and 85% on O<sub>2</sub> face mask @ 5lit/min, bilateral fine creptitations with occasional rhonchi were present in chest. Patient shifted to ICU for respiratory and hemodynamic support & monitoring. ABG pH-7.24, pO<sub>2</sub>-58, pCO<sub>2</sub>-64, hCO<sub>3</sub>-21. Right subclavian central venous line inserted for noradrenaline infusion & CVP monitoring. CXR revealed – Right lower lobe consolidation with left side pleural effusion and a mass lesion in left lower zone (Fig 1). ECG was normal

Patient was put on BiPAP support IPAP - 10/ EPAP - 5 with O<sub>2</sub> @3 l/min, noradrenaline infusion (2amp in 50ml 5%D) @ 4ml/hr, nebulization with duolin 4 hrly and inj cefoperazone + sulbactam and Inj. Solumedrol 40mg tds, iv inj furosemide 20mg bd were started. The patient showed symptomatic improvement.

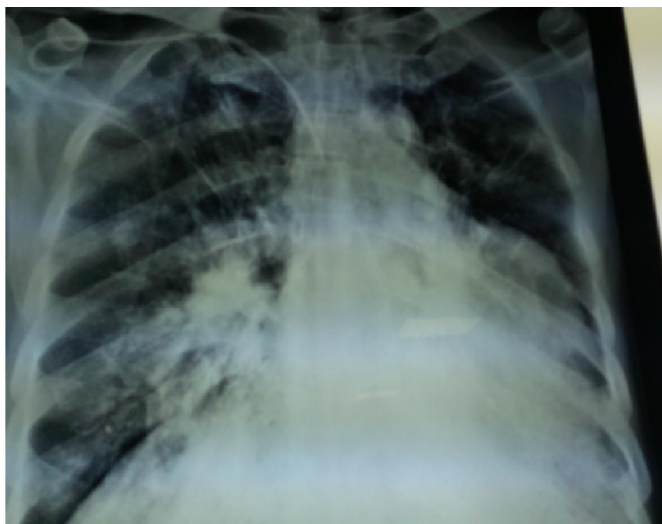


Fig.1.

After 8 hrs of admission patient had an episode of ventricular tachycardia for which patient was started on iv amiodarone bolus and infusion for 24hrs. The rhythm reverted back to normal sinus rhythm. Urgent cardiology opinion sought and 2 D-ECHO revealed dilated cardiomyopathy with dilated & globally hypokinetic left ventricle and severe left ventricular systolic dysfunction and LVEF - 15 %. No pericardial effusion. No intraventricular clots, mass, vegetation or RWMA was seen. Patient was started on tab dytor 10 mg bd, tab aldactone 25mg od, tab lanoxin 0.25 mg OD (5/7) and dobutamine infusion. Hba1c was found to be 6.4%. Patient was started on insulin according to sliding scale. Iv steroids and furosemide stopped. Initial investigations were normal except for raised liver enzymes (SGOT – 580 IU/L, SGPT – 256 IU/L). Day 2-5 ICU : CECT thorax was subsequently done which revealed - a large broad based soft tissue attenuation mass lesion (9.8cms SI X8cms AP X6.2cms TR) along left heart border suggestive of pericardial neoplastic lesion (d/d – fibroma & paraganglionoma). It also showed multiple patches of consolidation involving both the lungs with bilateral mild effusion (Fig 2).

Patient improved thereafter and BiPAP support and inotropes tapered & stopped. TLC increased to 16,900/mm<sup>3</sup> & antibiotics inj levofloxacin 500 mg od and inj clarithromycin 500 mg bd started. Patient shifted to high dependency unit.

Day 5-7 HDU : TLC count declined to 12,000. CT guided biopsy of pericardial lesion done and small iatrogenic pneumothorax was present post biopsy which was managed conservatively with high flow oxygen.

Day 8-10 HDU - Patient had sudden onset of breathlessness with fall in blood pressure and spO<sub>2</sub> for which patient was shifted to ICU. Immediate CXR revealed left sided pneumothorax and ICD insertion (Fig 3) was done and high flow oxygen given. Patient suffered sudden cardiac arrest, patient intubated and CPR as per recent ACLS guidelines started & revived within 45s and put on mechanical ventilation (Pressure regulated volume control mode, FiO<sub>2</sub>-60%, RR-15/min, PEEP-5, TV – 400ml, I:E – 1:2.2).

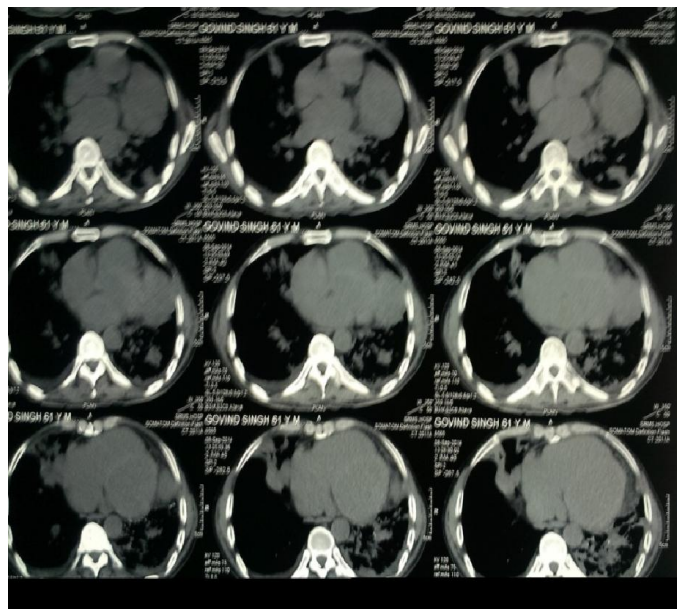


Fig.2.



Fig. 3.

Left subclavian central venous catheterization was done. Inotropes (noradrenaline and dobutamine) were started. Antibiotic escalation to inj piperacillin-tazobactam 4.5g qid, inj clindamycin 600mg tds and inj levofloxacin 500mg od done. Repeat cultures (urine, blood & sputum) sent.

Day 11- 13 ICU - Noradrenaline and dobutamine infusions were slowly tapered. Lower ICD left side was clamped, observed and removed. Biopsy report was S/o pericardial fibroma. ET culture was s/o pseudomonas, klebsiella & ESBL (E.coli) - for which inj.imipenem 500mg tds, inj colistin 1MU tds and neb colistin 1MU tds started and for candida guillereonidi - Amphotericin B(Lipid emulsion) was started.

Day 14-15 ICU - Surgical emphysema was noted on left side. Chest x ray revealed ICD in situ on left side and pneumothorax on right side (Fig 4). Right side ICD insertion was done immediately. CXR showed expansion of right lung and bilateral pneumonia was also seen (Fig 5). Since hypotension

was also present infusion vasopressin was added @1-2 units/hr to noradrenaline and dobutamine infusion. Patient was managed on ventilatory and inotropic support thereafter and on day 15 tracheostomy was planned but due to deranged coagulation profile and negative consent by relatives it was not done.

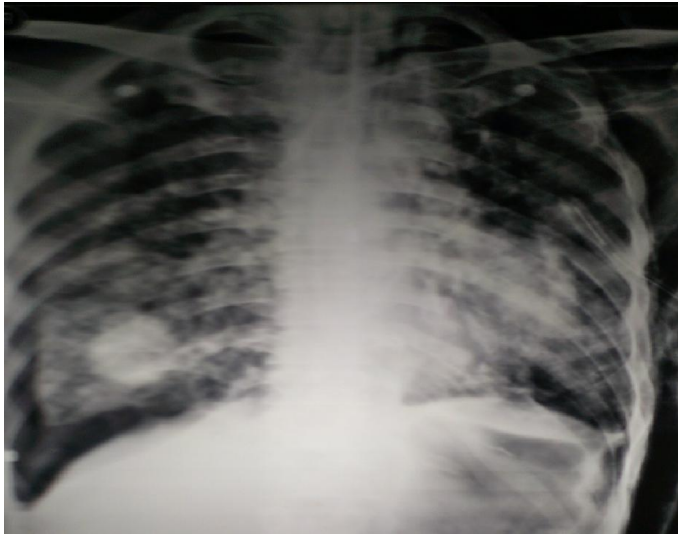


Fig. 4.

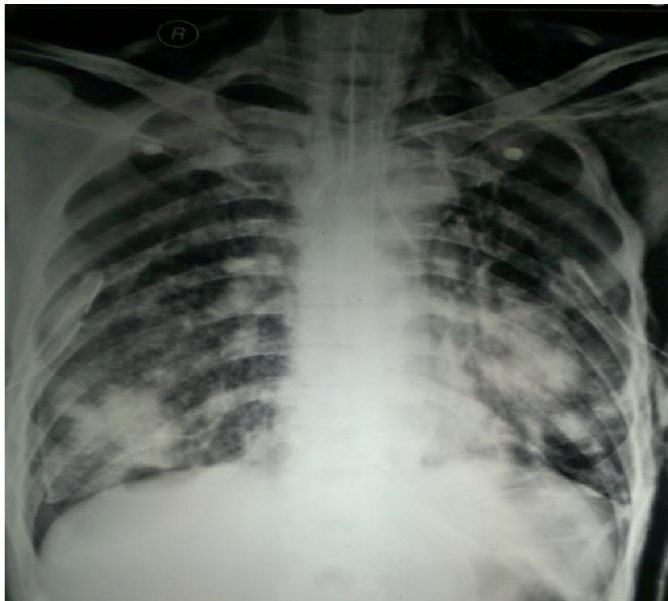


Fig. 5.

Day 16-20 ICU - Patient showed symptomatic improvement over next two days. Vasopressin then noradrenaline was tapered & stopped. Patient weaned off from ventilator support and shifted to BiPAP 12/6 and dobutamine support.

Day 20-24 ICU – left ICD was removed and couple of days later right ICD was removed. CXR showed bilateral expanded lungs with resolving pneumonic patches. Inotropes were tapered off and stopped and patient was shifted out of ICU.

Day 24-27 - Patient shifted out of HDU, started orally and discharged on 27<sup>st</sup> day to follow up with MR imaging of chest

and a referral for higher centre with cardio-thoracic surgical facility given to the patient.

## DISCUSSION

Primary cardiac neoplasm are a rare occurrence in clinical practice with prevalence of 0.02-0.056% (Lam *et al.*, 1993). Primary pericardial tumors account for 6-7-12.8% (Reynen, 1996) of above occurrence and so there prevalence ranges from 0.001-0.007%. The most common benign pericardial mass is pericardial cyst followed by lipoma (Patel *et al.*, 2004). Patients with primary pericardial neoplasm present with diverse non-specific symptoms ranging from exertional dyspnea, chest pain, cough, palpitation, fatigue, fever, facial and lower extremity edema which are usually because of rhythm abnormalities of heart, pericardial effusion, cardiac failure etc (Lamba and Frishman, 2012). Pericardial tumors may usually be encountered incidentally in a asymptomatic patient or during workup for an unrelated illness. Initial workup begins with radiological assessment, chest x ray which may reveal enlarged cardiac silhouette, an abnormal mediastinal contour or discrete mediastinal mass. Transthoracic echocardiography has a limited role in diagnosing pericardial tumors except for showing pericardial effusion and thickened pericardium. In case of pericardial fibroma, it may reveal an isolated mass with variable echogenicity and occasional calcification (Jeung *et al.*, 2002).

Computed tomography of the thorax may show location of tumor, its relation to neighbouring structures and invasion of adjacent structures (Wang *et al.*, 2003). Magnetic resonance imaging is a very powerful tool in diagnosing pericardial tumor. It provides useful information about myocardial invasion and the functional impact of neoplasm (Randhawa *et al.*, 2011). PET –CT is an important tool in staging the tumor and demonstrates distant and loco-regional spread (Ost *et al.*, 2008).

We present here a extremely rare case of pericardial fibroma and the wide array of respiratory, cardiovascular and infectious complications we encountered in our way of managing the patient in our ICU.

In our patient of pericardial fibroma, shock and respiratory distress can be attributed to pericardial fibroma as the cardiac failure (left ventricular failure – left ventricular systolic dysfunction LVEF – 15% and dilated cardiomyopathy) led to pulmonary odema and respiratory distress. The subsequent ventricular tachycardia was because of involvement of conduction system.

The other reason for sudden respiratory distress was probably because of acute exacerbation of COPD as was evident by increased PaCO<sub>2</sub> – 58 mm Hg which could have been precipitated by lower respiratory tract infection (as was seen by raised TLC and CXR – right lower zone consolidation).

Subsequent occurrence of pneumothorax on left side could have been because of iatrogenic (CT Guided Biopsy) reason. The pneumothorax on right side was probably because of

rupture of subpleural emphysematous bullae as the patient was a case of COPD on irregular treatment.

Later in the course of hospital stay, patient developed hospital acquired infections leading to sepsis which needed antibiotic escalation. Left pneumothorax which needed emergency ICD insertion and subsequent emergency intubation, CPR and number of days on mechanical ventilation led to development of VAP (*Pseudomonas*, ESBL *E.coli*, *Klebsiella* and *Candida guilliermondi*) which required use of imipenem, colistin, tigecycline and lipid emulsion amphotericin B and vasopressor and inotropic support (noradrenaline, dobutamine and vasopressin) which was gradually tapered off and patient was successfully weaned off ventilator support and shifted out of ICU and subsequently discharged.

### Conclusion

Because of the very rare occurrence and varied presentation of pericardial fibroma, initially diagnosing and then managing a case of pericardial fibroma is a challenge for any clinician.

The wide array of cardiovascular manifestations, respiratory and infectious complications we encountered during our critical care management of the patient along with the fact that very limited published literature makes the path of treating physician even more treacherous. In future, with the advent of advanced cardiac imaging techniques and with the use of minimally invasive methods and improved histochemical staining techniques, management of such cases will become safer & easier.

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