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Case Report

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Search the Culprit - Case Report on Malignant Pericardial Effusion with Life Threatening Cardiac Tamponade

Chandra Shekara Reddy[1], Arun Srinivas[2], Guru Prasad[3]

[1]Junior Consultant, Apollo BGS Hospital, Mysore, Karnataka - 570023, India; [2]HOD - Dept. of Cardiology, Apollo BGS Hospital, Mysore, Karnataka - 570023, India; [3]Senior Consultant Cardiologist, Apollo BGS Hospital, Mysore, Karnataka - 570023, India.

ABSTRACT

Introduction: Malignant massive Pericardial Effusion (PE) with Tamponade is life threatening and prompts immediate management.

Case Report: We report a case of massive malignant PE with Tamponade in a 40-year-old female with uncommon symptoms. She was immediately survived after an emergency Pericardiocentesis. It's HPE analysis, confirms presence of Mesothelioma. With multi-modality approach - we extensively searched for primary source of malignancy with Specific blood tests, Pleural & pericardial Fluid analysis, CT thorax & Pulmonary angiograms, Upper GI endoscopy and Sigmoidoscopy. With all negative reports, then referred to Oncology center, where diagnosed as adjacent left lower lobe of lung Mesothelioma by PET Scan. Underwent successful Left Lung Lobectomy and clinical follow-up confirms recovery.

Conclusion: Malignant Massive Pericardial Effusion with Tamponade is a serious condition. Primary goal of management is immediate Pericardiocentesis, followed by extensive search for Primary Metastatic Neoplastic locations. Survival rate increases with early diagnosis and prompt management.

Key Words: Mesothelioma, Pericardiocentesis, Primary Pericardial Malignancy, Secondary Metastasis, CT thorax, Pulmonary Angiograms

INTRODUCTION

Malignant Pericardial Effusion (PE) is a serious manifestation in advanced malignancies associated with significantly decreased patient survival.[1] Symptomatically often present with cardiac tamponade, which can rapidly lead to cardiovascular collapse or death.[1] Thus, rapid diagnosis and timely treatment is necessary.[1] There are limited number of randomized controlled trials (RCTs).[2] Therefore the number of class I level A indications are limited.[2]

CASE REPORT

A 40-years-female was admitted with history of untreated recurrent cough since 2 months and generalized weakness & breathing difficulty for 4 days. Associated with recurrent vomiting and epigastric pain, which radiates to left shoulder and posterior regions. There was no history of chest pain, giddiness, fever, significant weight loss, use of oral contraceptives or any surgery in recent past. Clinically she was afebrile, tachycardia (HR - 110 / min), muffled heart sounds, tachypneic, desaturation (SaO_2 - 88%) and mild bi-basal crepitations.

Preliminary Investigations: ECG records Sinus Tachycardia with low voltage complexes, Chest X Ray shows Cardiomegaly with Bilateral Cardio Phrenic angle haziness and 2D Echocardiography revealed Massive Pericardial Effusion with Tamponade (**Figure 1**) and so emergency Pericardiocentesis was done and drained around 600 ml of hemorrhagic fluid was drained. The Histopathological examination (HPE) of Pericardial fluid showed clusters of Malignant cells – suggesting Mesothelioma as shown in **Figure 2 A**. Biochemically it was Exudative with Total Protein of 5.8 gm/dl (Normal - < 3), increased LDH - 778 U/L (Normal - < 480) and with no growth on Culture. Other **Important investigations** are Complete Blood Count, Liver Function

Corresponding Author:
Dr. Arun Srinivas, Interventional Cardiologist, MBBS, MD, DM (Card), FESC, FACC, HOD, Dept. of Cardiology, APOLLO BGS Hospital, Adichunchanagiri Road, Kuvempunagara, Mysore, Karnataka, India - 570 023.

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Tests, Coagulation profile tests and Thyroid function tests - all were within Normal limits.

Specific investigations: She also underwent few specific investigations which includes Serum Cancer Antigen (CA) 125 was 256.1 U/ml (Normal - < 35) confirmed the presence of malignancy. Her d - Dimer was 4467 ng/ml (Normal < 500) due to Pro thrombotic state induced by malignancy. To rule out Pulmonary Embolism (PE), CT Pulmonary Angiogram was done, showed as Acute PE involving segmental branches as in **Figure 2 B**. Since the primary malignancy of the Pericardium is rare, we then extensively searched for Primary source. CT Abdomen **(Figure 2 B)** - reported as Hypodense lesions in Liver VII Segment and Mild bilateral Pleural effusions. Then left sided Pleural Effusion was drained and its HPE confirmed malignant (Mesothelioma) cells contamination. Ultra Sound Breasts **(Figure 3 A)** - reported as few tiny cysts in lower outer quadrant of Left Breast with same side few axillary lymphadenopathy. Also showed thrombosed Left Brachiocephalic, Subclavian, Axillary and Brachial veins till elbow. Upper GI Endoscopy **(Figure 3 B)** - reported as Pangastritis. Colonoscopy **(Figure 3 C)** - reported as normal except, multiple Superficial Ulcers of Terminal ileum (10 cm), biopsy taken and its HPE analysis as in **Figure 3 C** - reported as Small mucosal ulcerations with edematous Lamina Propria densely infiltrated by Lymphoplasma cells with fair number of Eosinophils (12 - 15 / hpf). Complete reports were mentioned along with respective figures. Despite of extensive search for Primary source for Neoplasm, none of the investigations were confirmative.

Differential Diagnosis: The important causes for Differential Diagnosis are Primary Neoplasm of Pericardium and Tubercular Pericarditis. Primary Neoplasm of Pericardium is rare. Although Tuberculosis is common in our society with no history of exposure to Tuberculosis, non-smoker, Non-Diabetic and negative Mantoux test - rules out Tuberculosis.

Management: As preliminary investigation confirmed Massive PE with Tamponade, underwent emergency Pericardiocentesis, in Cathlab under Fluroscopic guidance. Procedure done using Pigtail catheter, introduced through subcostal approach. Under Local Anesthesia and aseptic precautions, drained 600 ml of hemorrhagic pericardial fluid. Pigtail Catheter was kept in-situ for next 48 hours, but drained nil. Then cardiac functions were improved drastically as confirmed by her symptoms, ECG and 2D Echo (**Figure 1**). Also treated with O2, Antacids, Antiemetics, IV Fluids and Antibiotic. She had mild pre-renal failure, corrected (S. Creatinine from 1.6 to 1). On 4th day, she was discharged with stable hemodynamics and referred to Oncology center for further evaluation and management. We followed up clinically and it was informed that she had Primary Neoplasm from Left Lower Lobe of lung as confirmed by PET Scan. She was then treated by Surgical excision (Left Lower Lung Lobectomy), Chemotherapy and Radiotherapy. She was symptomatically better thereafter.

Discussion: PE is a common finding in clinical practice either as an incidental finding or as a manifestation of a systemic or cardiac disease.[3] It is uncommon to present as cause for Malignancy.[4] Malignant PE is usually secondary to adjacent lung cancers and rarely presents with hemodynamic instability.[5] Primary malignancy of the pericardium is rare and requires high index of suspicion.[5] The definitive etiological differentiation of malignant PE is of particular therapeutic and prognostic importance.[1] Establishing the etiology of PE remains challenging and difficult to assess on the basis of clinical assessment only.[1] Patient's presentation and development of symptoms depends on three principle factors - the volume of the PE, the rate of accumulation and the elasticity of the pericardium. Larger volume effusions are better tolerated if the rate of accumulation is slow and the pericardial elasticity is high.[4] Severity of symptoms depends on rate of accumulation and not amount of volume of Pericardial fluid.[4] Drainage should be limited to <1L at a time.[6] Larger volumes may cause right ventricular dysfunction.[6] The drain should remain until draining less than 25 mL per day, with prolonged drainage reducing the recurrence rate from 23% to 6%.[6] Pericardial sac (Fibrous layer which covers the outer layer of heart) contains only about 25 mL of fluid (20 to 60 mL).[7] Increased production, impaired drainage, or both will lead to excessive fluid accumulation (Pericardial Effusion).[7] Requires Percutaneous aspiration - a technique called as Pericardiocentesis.[7] Echocardiography and Fluoroscopy -guided percutaneous pericardiocentesis has become the standard intervention for evacuating pericardial effusions, as well as an essential tool in the diagnostic work-up of an unexplained pericardial effusion.[7]

European Society of Cardiology (**ESC**) provides guidelines endorsed by European association for cardio-thoracic surgery (**EACTS**) provides list of all possible Class of recommendation, Level of evidence and Reference(s) supporting recommendations for Management of Malignant PE as mentioned in **Table 1**.[2] Despite the magnitude of this serious problem, little progress has been made in early diagnosis and treatment of PE secondary to malignant disease.[1]

Typical features of Cardiac Tamponade:6

- Clinical diagnosis + 2D Echocardiographic confirmation with Pulsus paradoxus (highly sensitive (98%) and specific (83%)).
- Beck's triad (Raised JVP, Muffled heart sounds and Hypotension) may be present.
- ECG - Low voltage QRS (<5 mm QRS amplitude in all limb leads) and electrical alternans.
- Chest X Ray - Increased Cardio Thoracic (CT) ratio.

Learning Points

- Malignancy is one of the rare causes for Massive PE with Tamponade.
- Severity of symptoms depends on rate of accumulation and not volume of Pericardial fluid.
- Emergency Pericardiocentesis is Lifesaving.
- Extensive search for both Primary source and Metastatic lesions of malignancy is must.
- Management should be tailored according to etiology and must be referred to higher center at the earliest for further evaluation and management if required.

Limitations

- No PET Scan
- No high-grade tissue, blood or pericardial fluid analysis

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Table 1: Recommendations for Management of Malignant Pericardial Effusion.2

Recommandations	Class[a]	Level[b]	Ref.[c]
Pericardiocentesis is recommended for cardiac tamponade to relieve symptoms and establish the diagnosis of malignant pericardial effusion	I	B	
Cytological analyses of pericardial fluid are recommended for the confirmation of malignant pericardial disease	I	B	191
Pericardial or epicardial biopsy should be considered for the confirmation of malignant pericardial disease	IIA	B	
Pericardial or epicardial biopsy should be considered for the confirmation of malignant pericardial disease	IIA	B	
Tumour marker testing should be considered for distinguishing malignant from benign effusions in pericardial fluid	IIA	B	193
Systemic antineoplastic treatment is recommended in confirmed cases of neoplastic aetiology	I	B	
Extended pericardial drainage is recommended in patients with suspected or definite neoplastic pericardial effusion in order to prevent effusion recurrence and provide intrapericardial therapy	I	B	

Table 1 ***(Continued)***

Intrapericardial instillation of cytostatic/ sclerosing agents should be considered since it may prevent recurrences in patients with malignant pericardial effusion	IIA	B	197 - 204
Intrapericardial cisplatin should be considered in pericardial involvement in the course of lung cancer and intrapericardial instillation of thiotepa should be considered in breast cancer pericardial metastases	IIA	B	197 , 198, 200, 204
Radiation therapy should be considered to control malignant pericardial effusion in patients with radiosensitive tumours such as lymphomas and leukaemias	IIA	B	
Pericardiotomy should be considered when pericardiocentesis cannot be performed	IIA	B	205
Percutaneous balloon pericardiotomy may be considered for the prevention of recurrences of neoplastic pericardial effusions	IIB	B	
Pericardial window creation via left minithoracotomy may be considered in the surgical treatment of malignant cardiac tamponade	IIb	B	207
Interventional techniques should consider seeding of neoplastic cells, patient prognosis and the overall quality of life of the patients	IIa	C	

[a]Class of recommendation.
[b]Level of evidence.
[c]Reference(s) supporting recommendations.

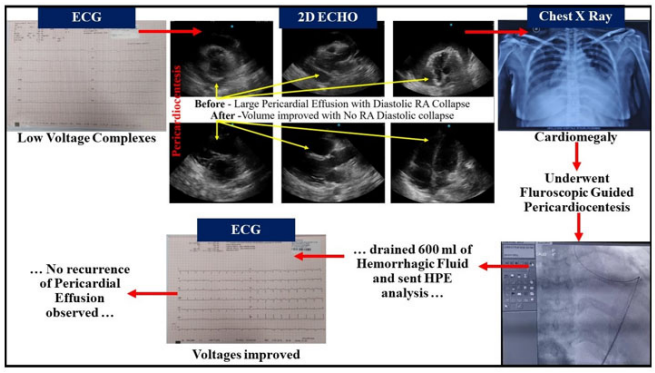
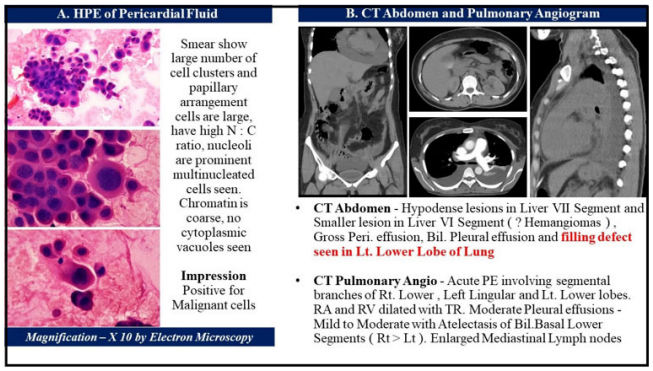


Figure 1: ECG and 2D Echo Findings of Pericardial Effusion and its Tapping.



Figures 2: **Figure 2A:** Histo Pathological Examination (HPE) Analysis of Pericardial Fluid.

Figure 2B: Computerized Tomography of Abdomen and Pulmonary Angiogram.

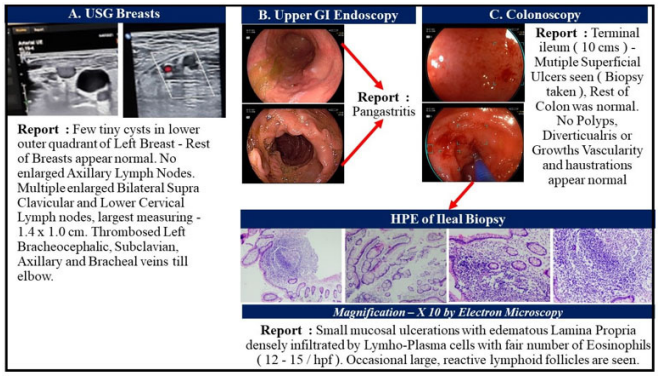


Figure 3: **Figure 3A:** Ultra Sound of Breasts.

Figure 3B: Upper Gastro Intestinal Endoscopy.

Figure 3C: Colonoscopy and HPE Analysis of Ileal Biopsy.