A Deceptive Diagnosis: Constrictive Pericarditis Masquerading as Liver Disease

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Patient Presentation

- 50-year-old male
- Chief Complaints:
  - Bilateral pedal edema since 2 months,
  - Dyspnea on exertion (NYHA II) since 1 month
  - Abdominal distension since 15 days

- Medical History:
  - Hypertension since last 10 years on regular medication
  - Chronic alcohol intake (since last 15 years)

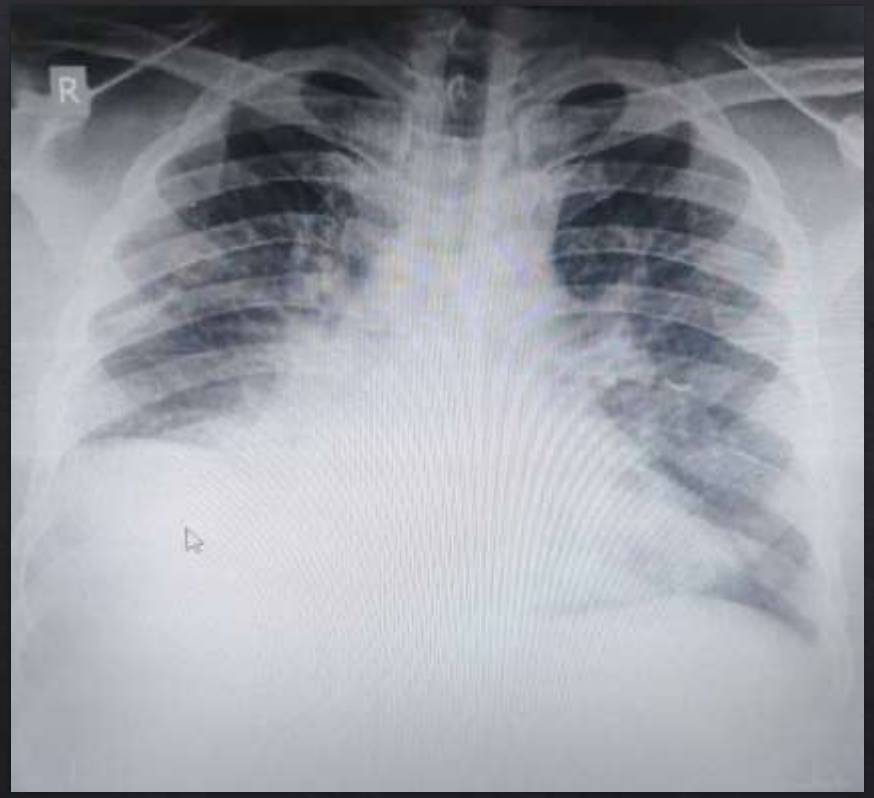
Physical Examination on presentation

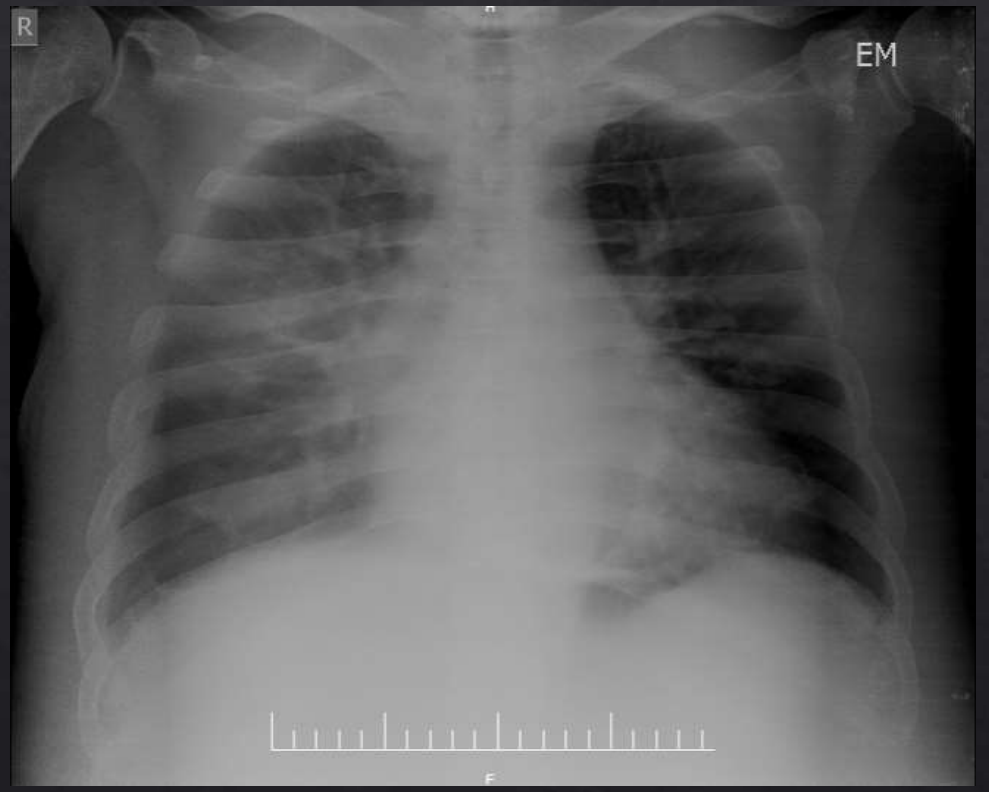
- Vital Signs: Normal pulse, blood pressure with tachypnea
- General Appearance: Bilateral pedal edema, Raised JVP, Abdominal distension, Scrotal swelling
- Cardiovascular: Normal S1 and S2 , No murmurs , No additional sounds
- Respiratory: Bilaterally air entry present with Bilateral basal crackles

Laboratory Investigations

- Routine Blood Tests (CBC,RFT,LFT,ELECTROLYTES) : Unremarkable
- Among cardiac Markers: troponin-negative, normal CK-MB

Chest X- RAY





USG ABDO/PELVIS

- Hepatomegaly with coarse echotexture with surface nodularity s/o liver parenchymal disease
- Gross ascites

USG INGUINO-SCROTAL

- Mild hydrocele and bilateral diffuse scrotal wall edema

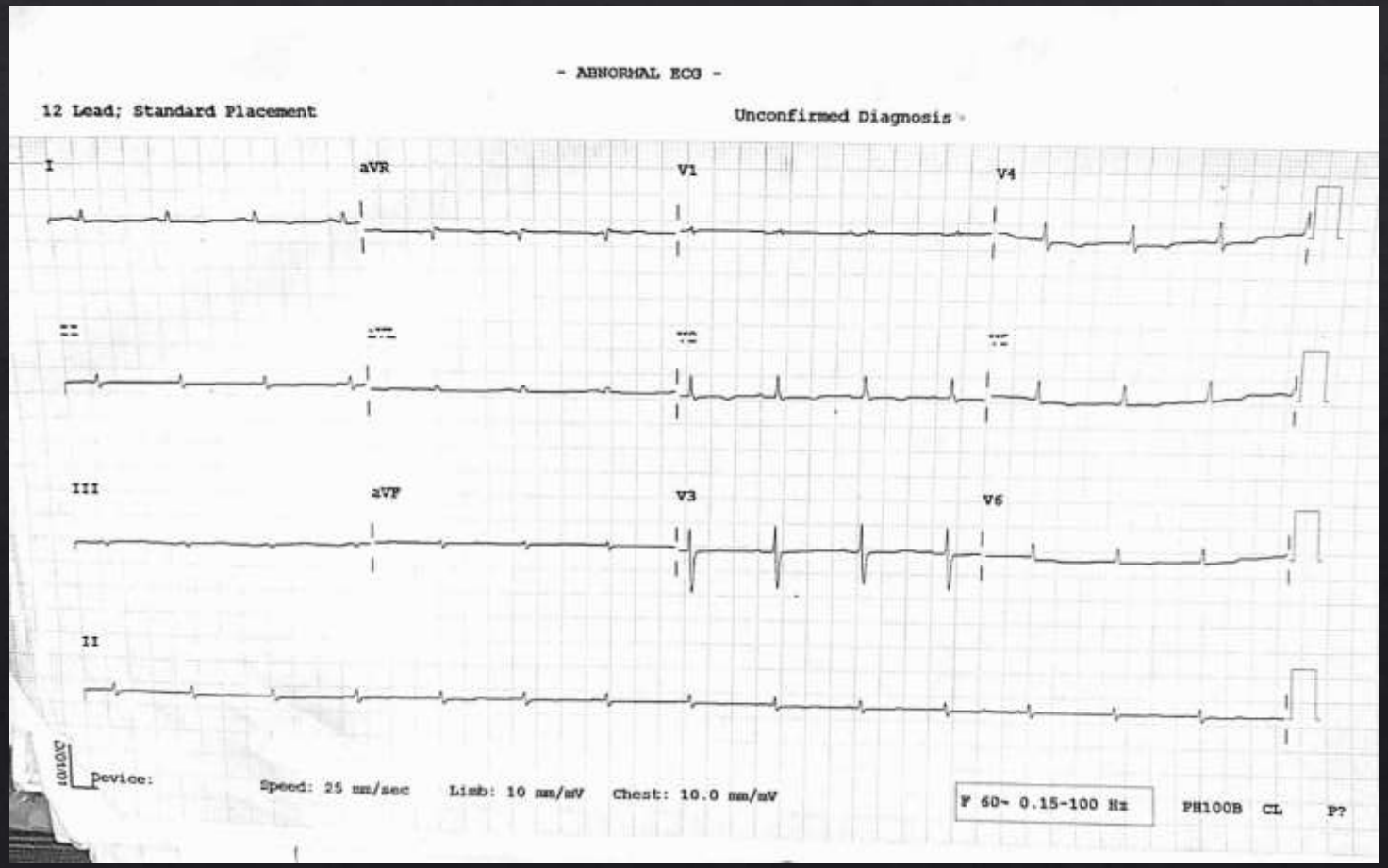
- Patient was admitted in medicine ward with the above complaints with a provisional diagnosis of Chronic liver disease.

- Patient was referred to cardiology for 2D echocardiography.

- On further investigation: NT-proBNP was Elevated (16055 pg/mL)

ECG

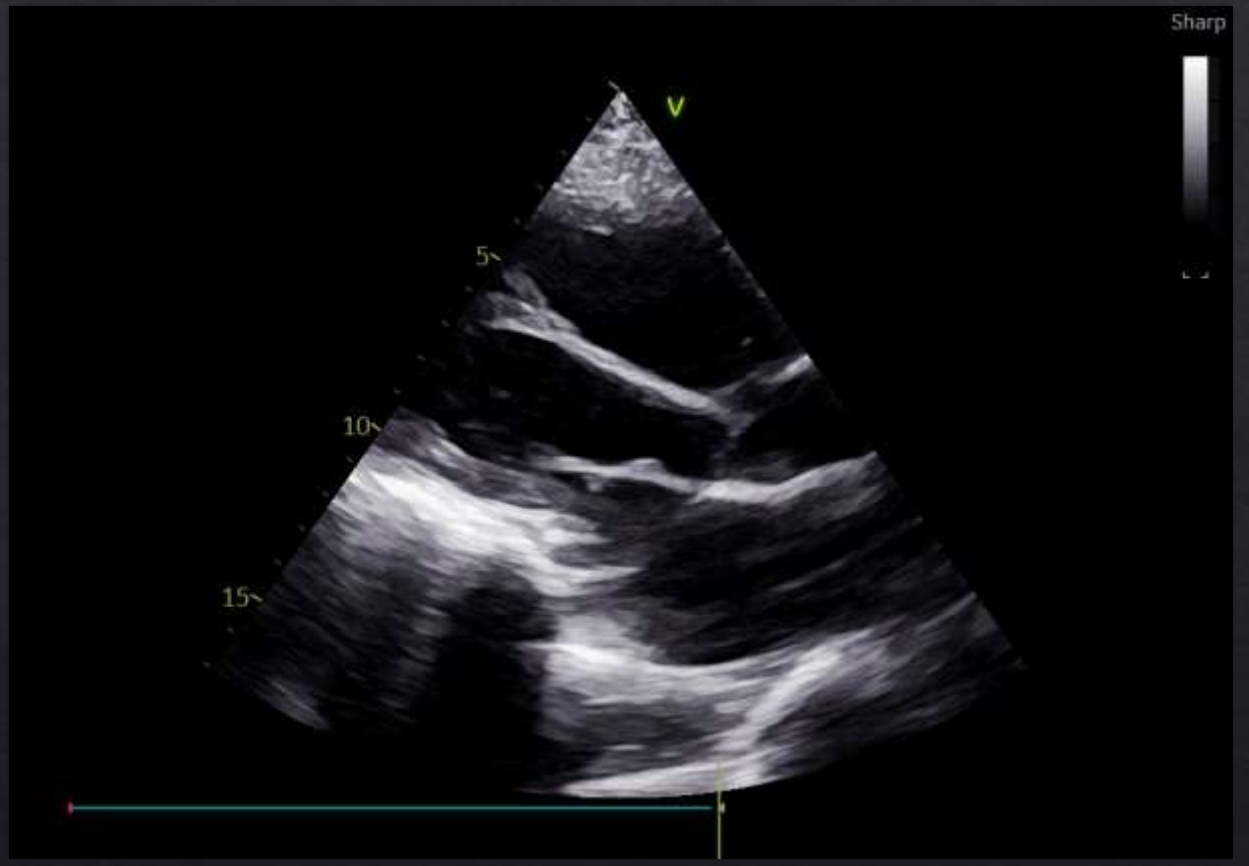
- Normal sinus rhythm, low voltage



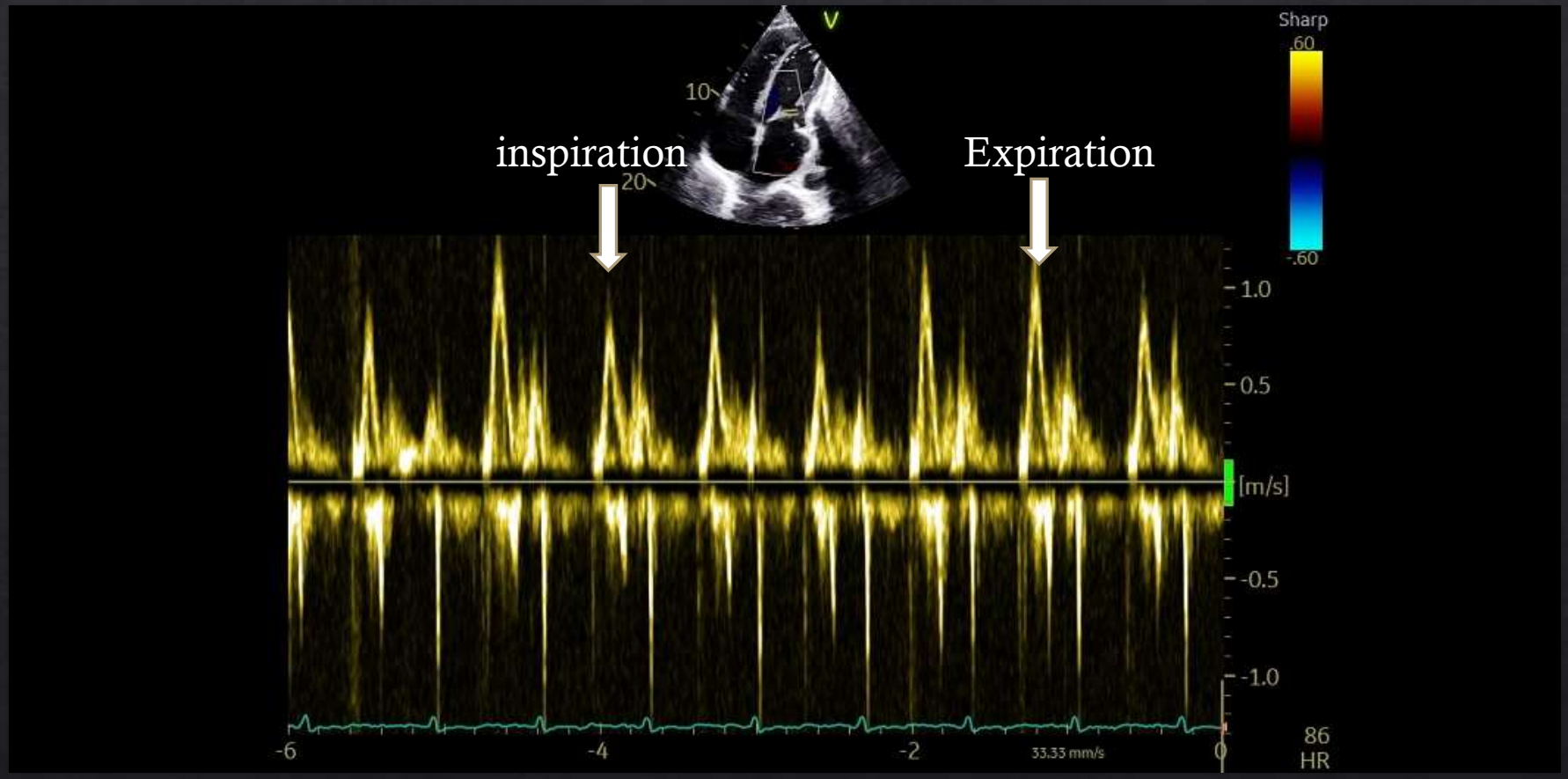
Echocardiography Findings

- Fair Left Ventricular Systolic Function (EF 55%)
- Bi-atrial Enlargement
- Septal Bounce
- Respiratory Variation of Mitral Inflow
- Annulus Reversus (on tissue doppler imaging)
- Hepatic Venous Diastolic Flow Reversal
- Depressed Right Ventricular Function (TAPSE <10)
- Left Ventricular GLS of -15
- Right Ventricular GLS of -8.6

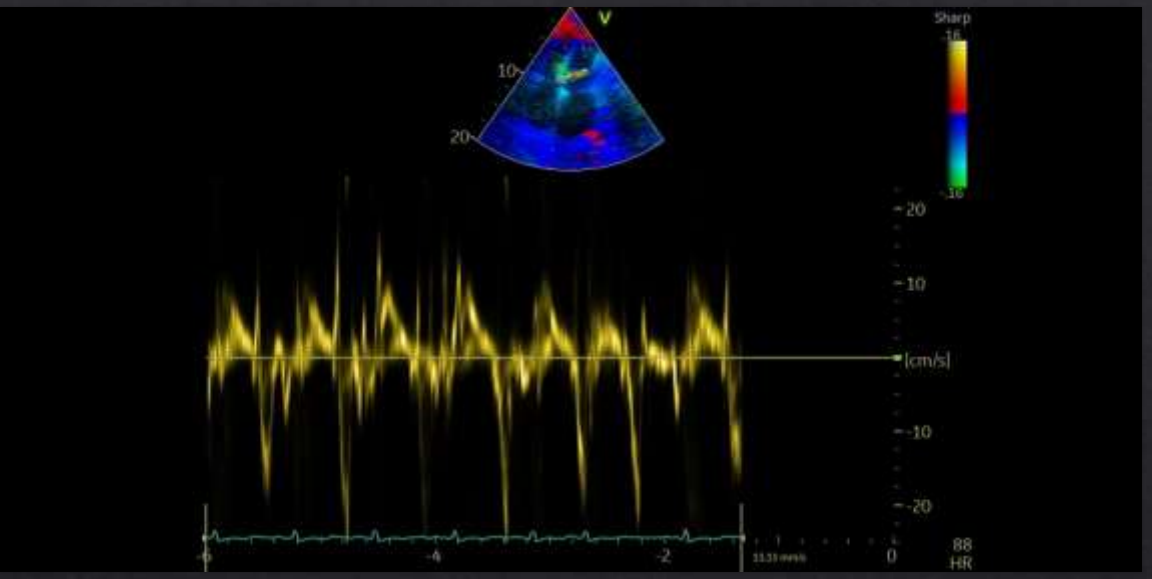
Septal bounce



Respiratory variation of mitral inflow

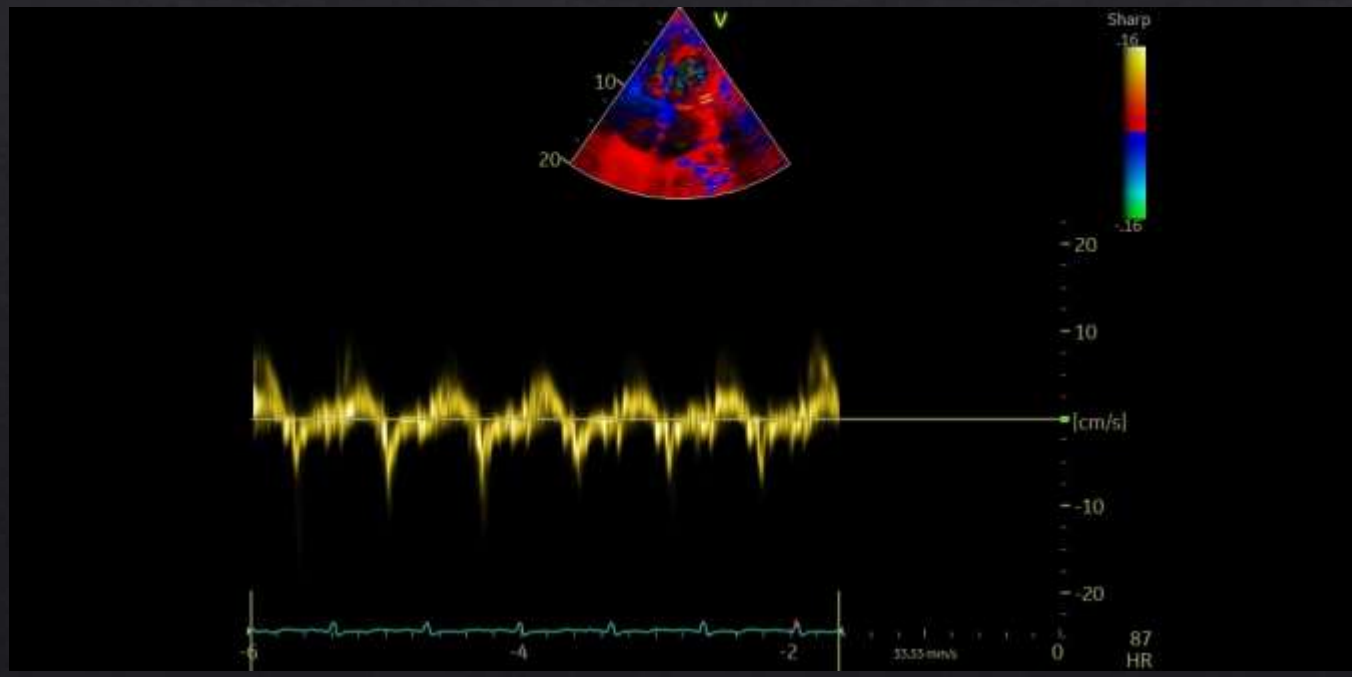


Annulus Reversus

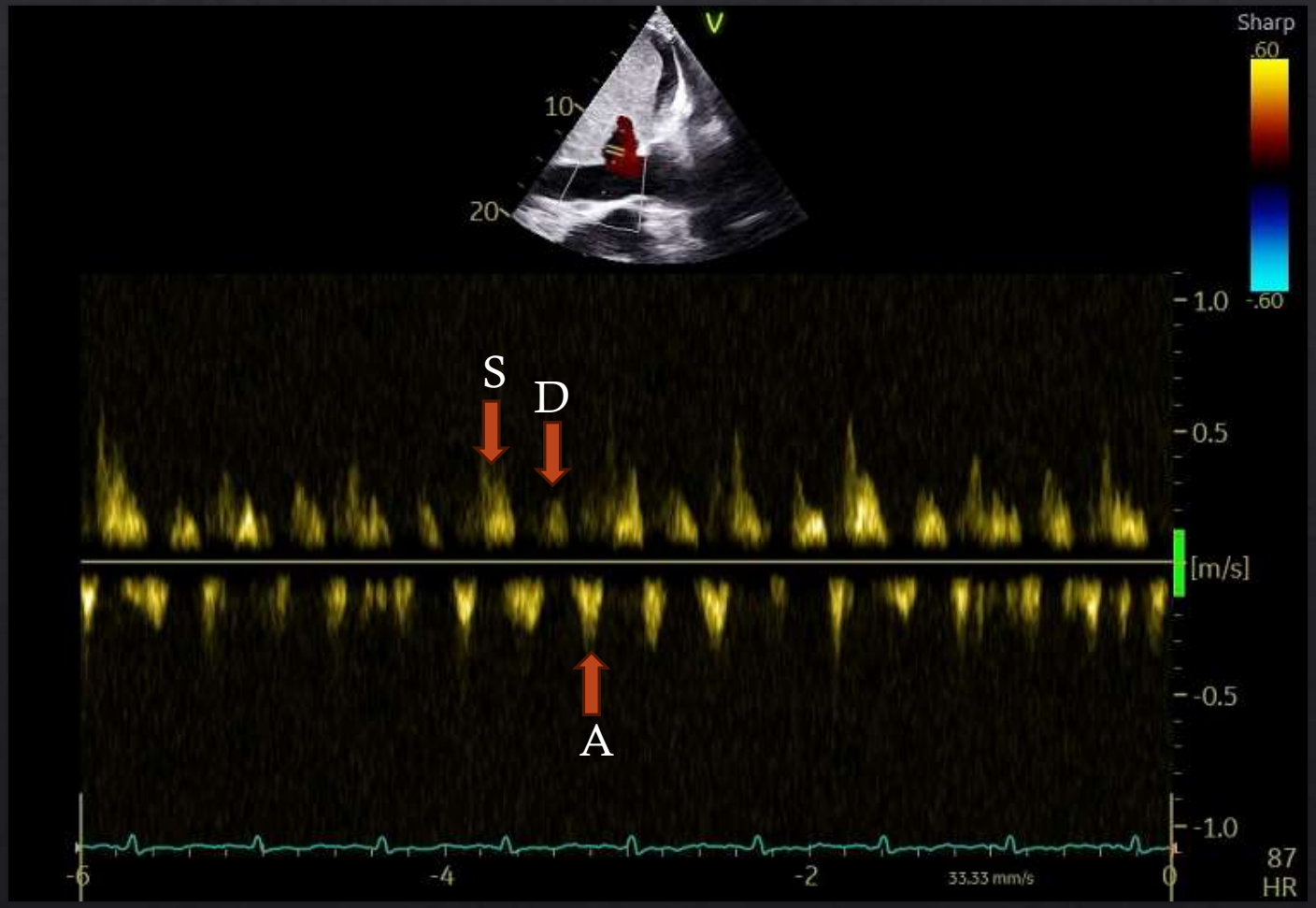


Septal annulus TDI

Lateral annulus TDI

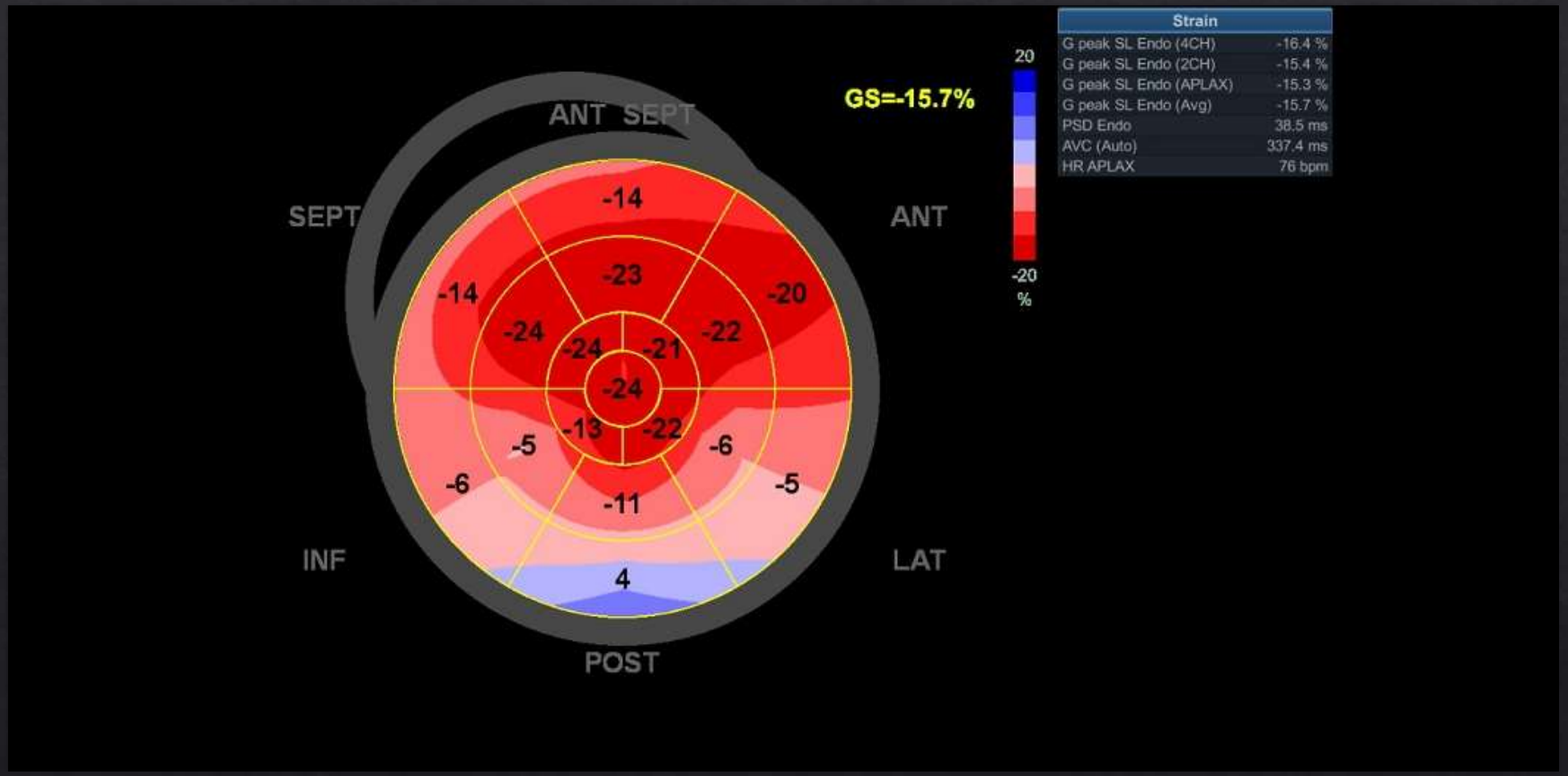


Hepatic Venous Diastolic Flow Reversal



Global longitudinal strain

(Left Ventricular GLS of -15 with Hot Septum Sign)

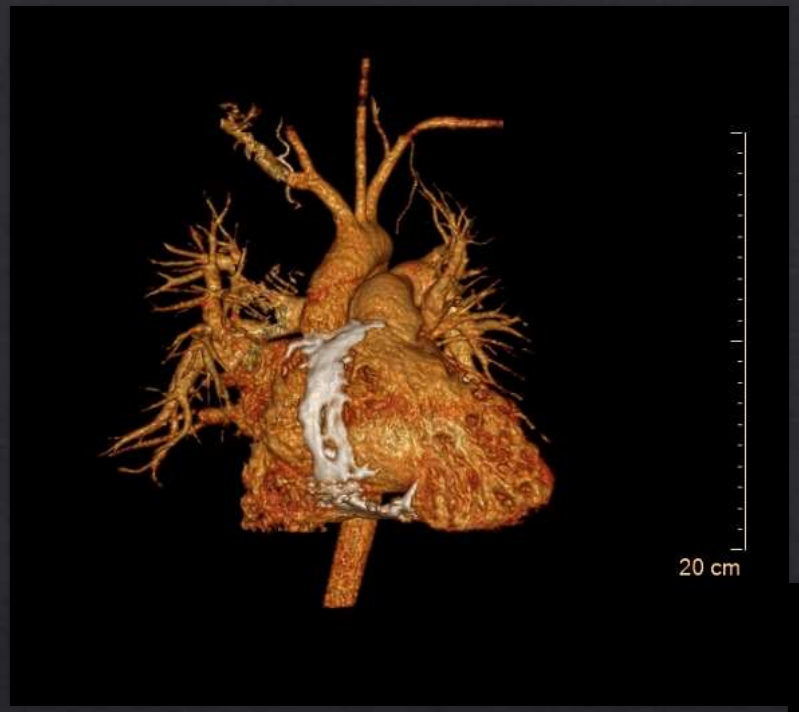


Cardiac CT Findings

Pericardial Thickening and Calcification

Focal Fan-like Calcifications Invading Myocardium noted in lateral and inferior wall of left ventricle at basal and mid level, right ventricular free wall and inferior wall at basal level, right atrioventricular groove.

Findings consistent with Chronic Calcific Constrictive Pericarditis



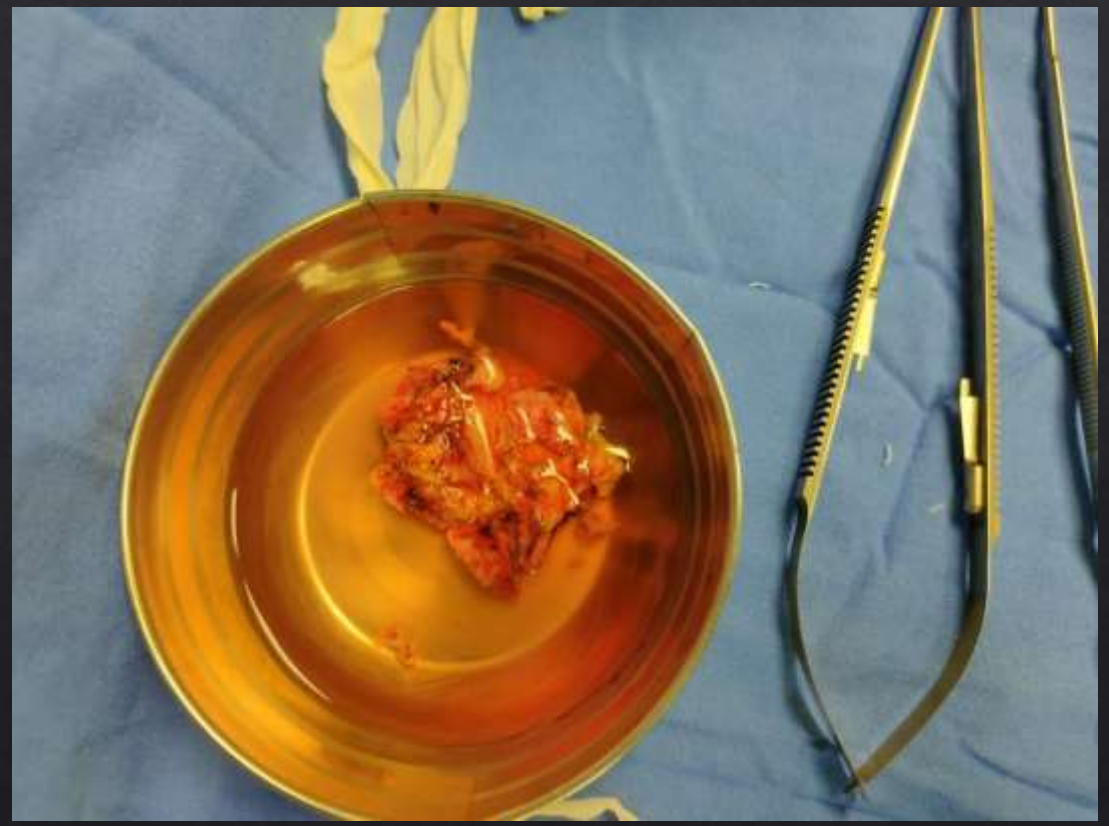
Diagnosis and Treatment

- Final diagnosis : Chronic Constrictive Pericarditis with Cardiac cirrhosis.

- Medical Management for Heart failure
  - Diuretics (furosemide)
  - Aldosterone antagonists (spironolactone)

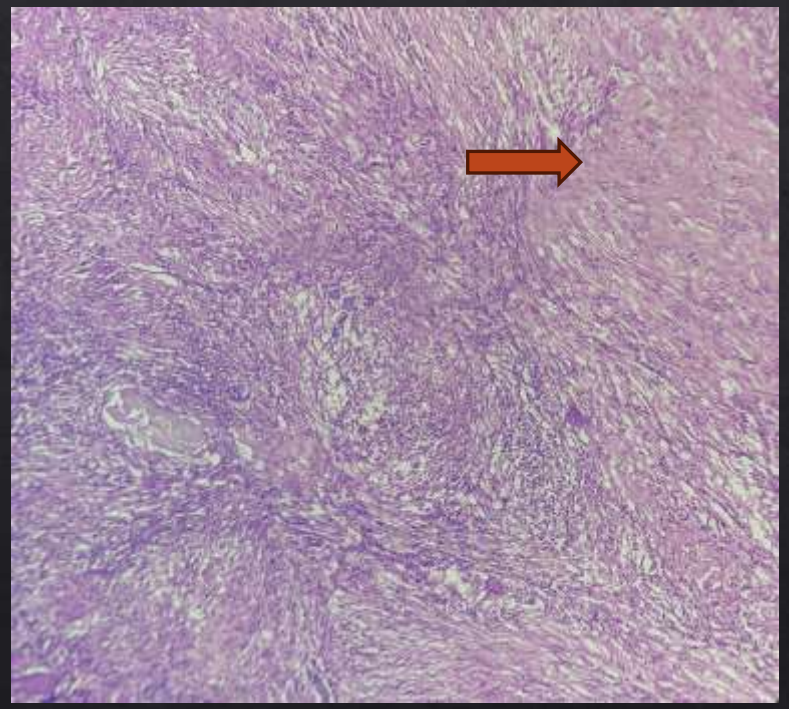
- Surgical Management (Definitive):
  - Pericardiectomy

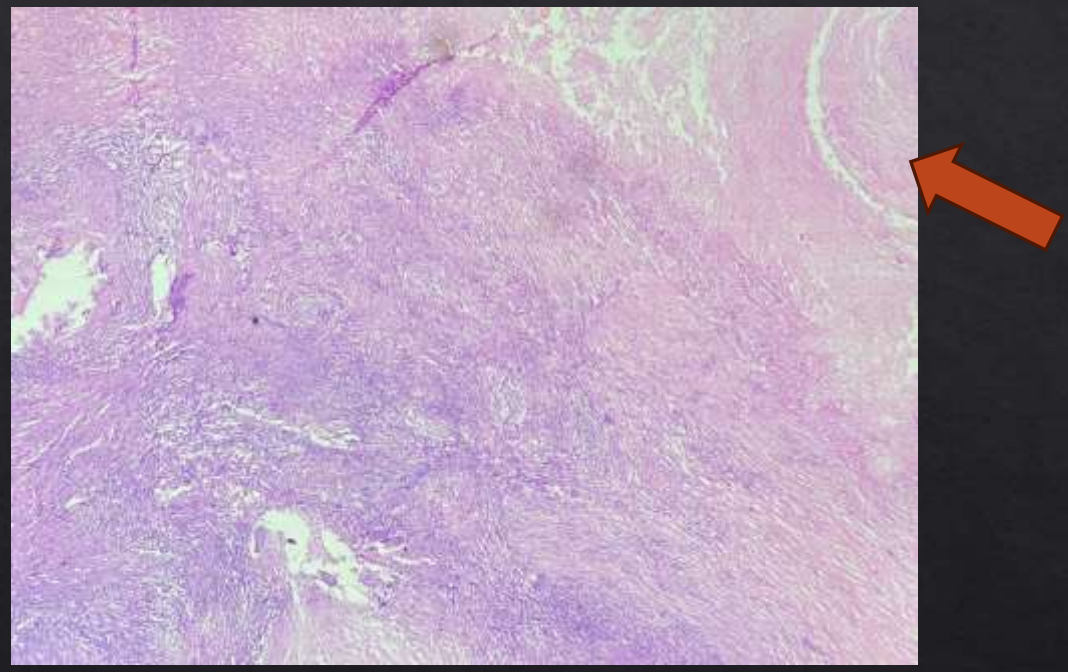
- Intraoperative Findings
- Thickened and Calcified Pericardium
- Severely Thinned Right Atrial Wall
- Calcification Invading Right Atrial Myocardium



HISTOPATHOLOGY REPORT of PERICARDIAL TISSUE

- Suggestive of tuberculous pericarditis with calcification





show well formed **Epitheloid cell granuloma**, areas of **Caseous necrosis** and Langhan's type of giant cells. Also seen are extensive areas of calcification.

Postoperative Course

- Initial Recovery:
  - Intubated with inotropic support
  - Gradual weaned and extubated
  - Hemodynamically stable and discharged on heart failure management
  - Anti Tubercular treatment was started after HPE reports.

- On discharge patient was clinically improved. 2D echo s/o fair LV systolic function with signs RV dysfunction.

Late Course

- Patient was readmitted after 1 month with right heart failure symptoms.
- Under ICU care, was initiated on inotropic and ventilatory support.
- Finally patient had an unfavorable outcome (death of patient).

Discussion

- Chronic constrictive pericarditis is a rare but serious condition that results from chronic inflammation and fibrosis of the pericardium, leading to impaired diastolic filling of the heart.
- The pathogenesis of constrictive pericarditis involves various factors, including infections, autoimmune diseases, cancer, radiation induced and post cardiac surgery.
- This inflammatory process becomes chronic, leading to the formation of a rigid, fibrotic pericardium, which restricts the heart's ability to expand and fill with blood during diastole, ultimately reducing cardiac output and leading to heart failure.
- D/D of CCP are restrictive cardiomyopathy.

	CONSTRICTION	RESTRICTION
Prominent *y* descent in venous pressure	Present	Variable
Paradoxical pulse	Approximately 1/3 cases	Absent
Pericardial knock	Present	Absent
Equal right-left side filling pressures > right	Present	Left at least 3–5 mm Hg
Filling pressures >25 mm Hg	Rare	Common
Pulmonary artery systolic pressure >60 mm Hg	No	Common
"Square root" sign	Present	Variable
Respiratory variation in left-right pressures/flows	Exaggerated	Normal
Ventricular wall thickness	Normal	Usually increased
Pericardial thickness	Increased	Normal
Atrial size	Possible left atrium enlargement	Biatrial enlargement
Septal "bounce"	Present	Absent
Tissue Doppler E' velocity	Increased	Reduced
Speckle tracking	Normal longitudinal, decreased circumferential restoration	Decreased longitudinal, normal circumferential restoration

- Early diagnosis and prompt intervention are crucial for improving symptoms, long-term outcomes in patients with CCP.
- The diagnostic workup typically includes a combination of imaging modalities, such as echocardiography, computed tomography, and cardiac magnetic resonance imaging, to detect pericardial thickening, and calcification, and assess diastolic dysfunction.
- Management include medical management i.e heart failure management. And definitive management include pericardiectomy.

- The treatment of choice for constrictive pericarditis is pericardiectomy, that involves removing the thickened pericardium to relieve the constriction around the heart.
- Pericardiectomy have a relative high perioperative mortality ranging 2-20%.
- Long term result are worst in reduced LVEF, moderate to severe TR, advanced age, impaired renal function.
- In this case despite the successful pericardiectomy, because of myocardial involvement, the patient's clinical course was complicated by a recurrence of heart failure, leading to the patient's demise.

Take Home Message

- 1. Chronic constrictive pericarditis is a rare but serious condition that requires a high index of suspicion and prompt diagnosis and intervention for optimal outcomes.
- 2. Comprehensive diagnostic evaluation, including imaging modalities like echocardiography with global longitudinal strain assessment and cardiac CT is crucial for confirming the diagnosis and guiding management.
- The potential for irreversible myocardial damage in advanced cases highlights the importance of early recognition and timely intervention to improve long-term prognosis.

Thank you

- Discussion
- Pathophysiology of Constrictive Pericarditis
- Diagnostic Challenges and Role of Imaging
- Medical and Surgical Management Strategies
- Prognostic Implications and Future Directions