UNMASKING CARDIAC SARCOIDOSIS THROUGH RECURRENT VT – A RARE CLINICAL CONUNDRUM

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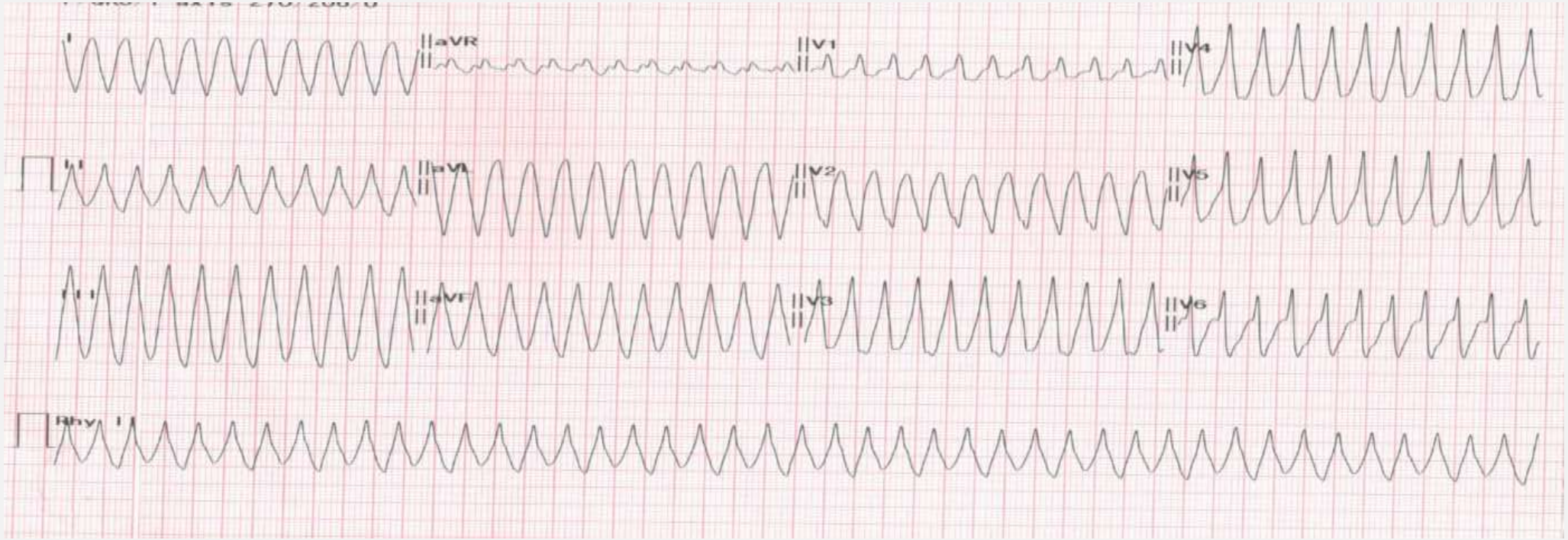
HISTORY AND EXAMINATION

- 40 year old male
- Chief complaints of sudden onset palpitations and dyspnoea for 1 hour.
- No h/o chest pain or syncope.
- No significant medical or surgical history in the past.
- Patient was hemodynamically unstable initially.
- Systemic examination revealed no significant abnormality.

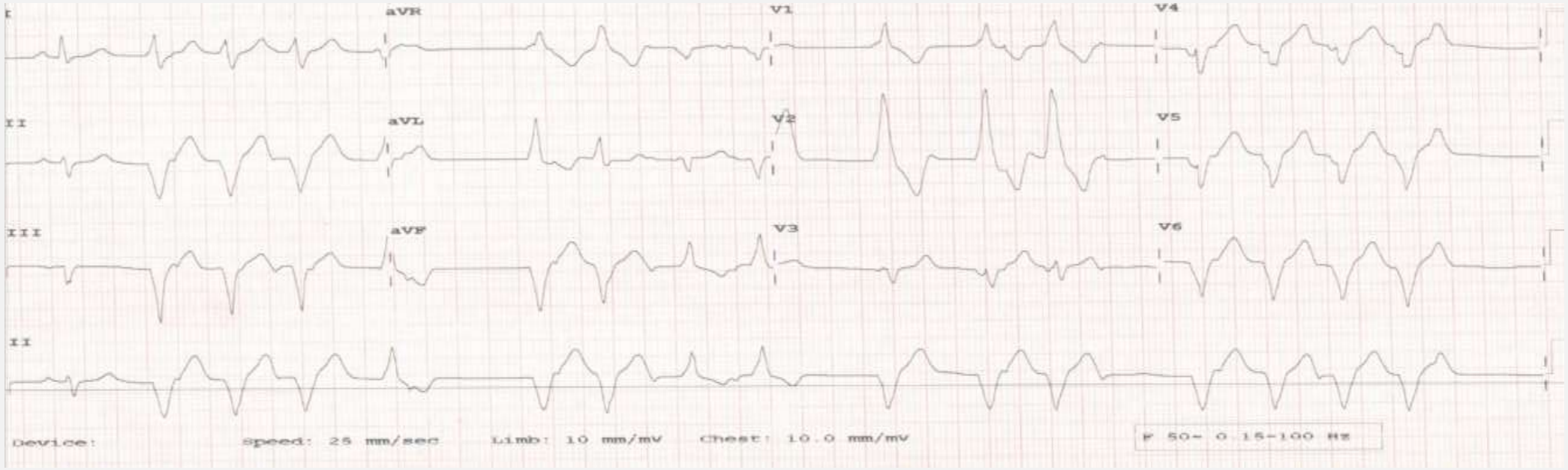
INITIAL ELECTROCARDIOGRAM

- Electrocardiogram showed monomorphic ventricular tachycardia (VT).
- Synchronized electrical cardioversion 150 J was given to terminate VT.



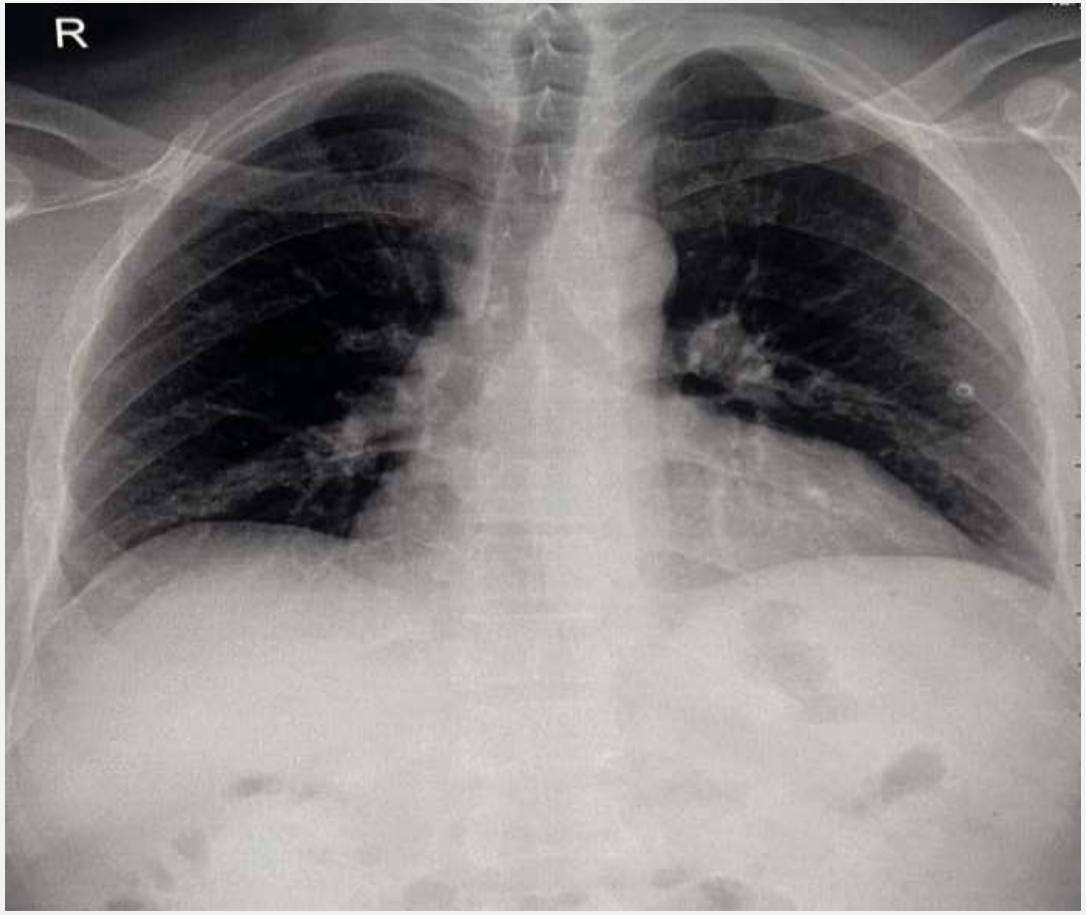
POST CARDIOVERSION

- Post cardioversion, ECG showed multiple ventricular premature complexes of two different morphologies.
- Later, patient had two repeated episodes which terminated spontaneously.



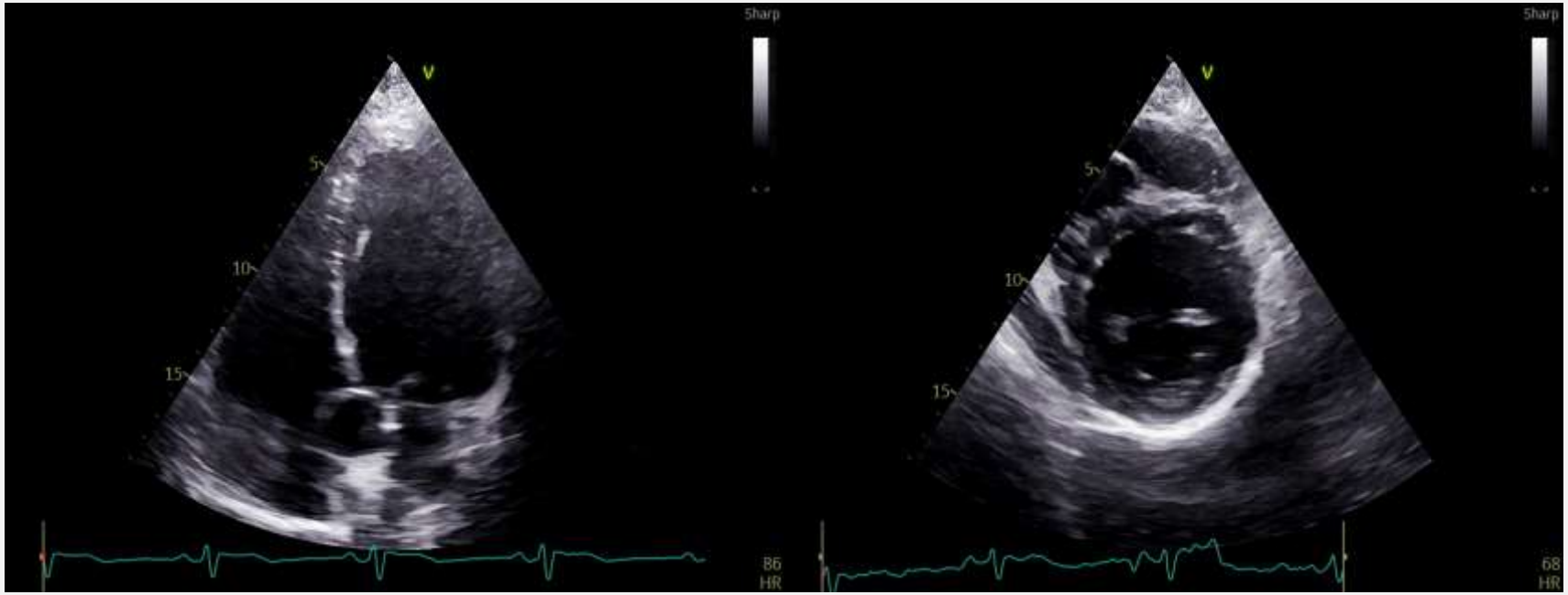
CHEST X-RAY

- No cardiomegaly.
- Normal lung fields.



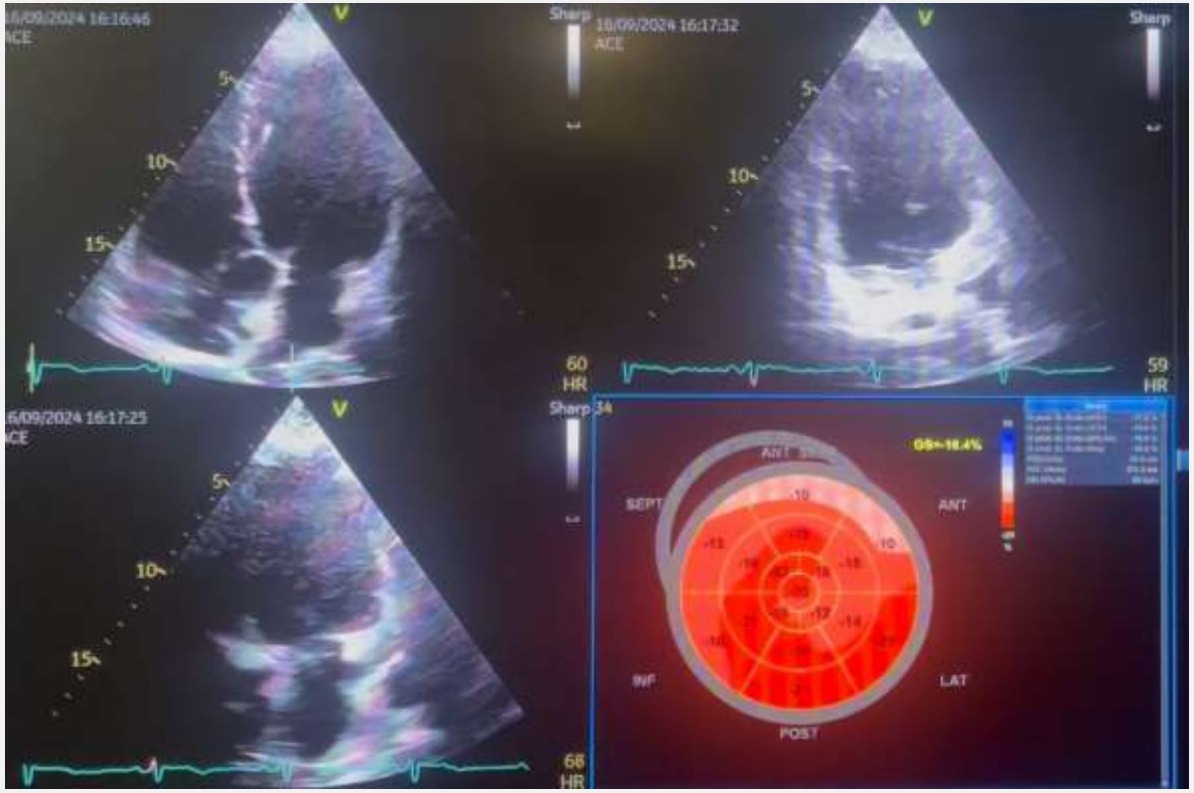
ECHOCARDIOGRAPHY

- Dilated LV
- Mild left ventricular systolic dysfunction with an ejection fraction of 45%.
- Mild mitral regurgitation
- Mild tricuspid regurgitation
- Mild pulmonary arterial hypertension



LV GLOBAL LONGITUDINAL STRAIN

- GLS was mildly reduced (-16.4%).
- There is significant reduction of regional wall longitudinal strain of basal anterior septum and anterior wall.



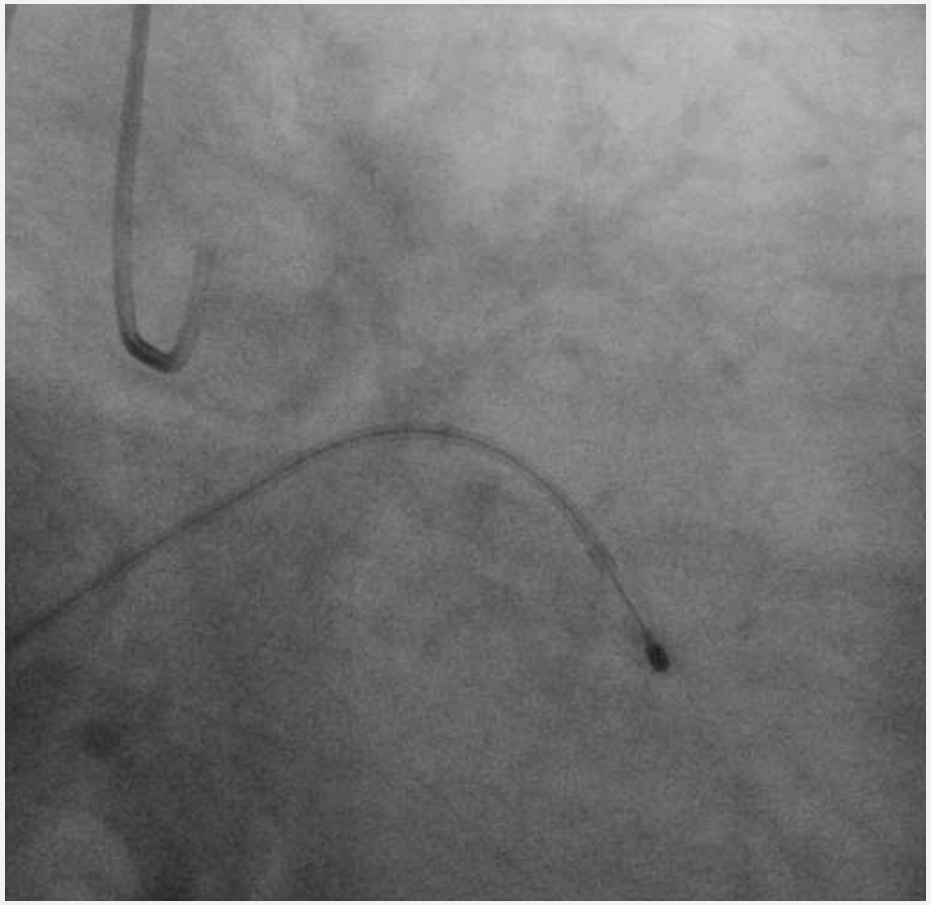
ROUTINE BLOOD INVESTIGATIONS

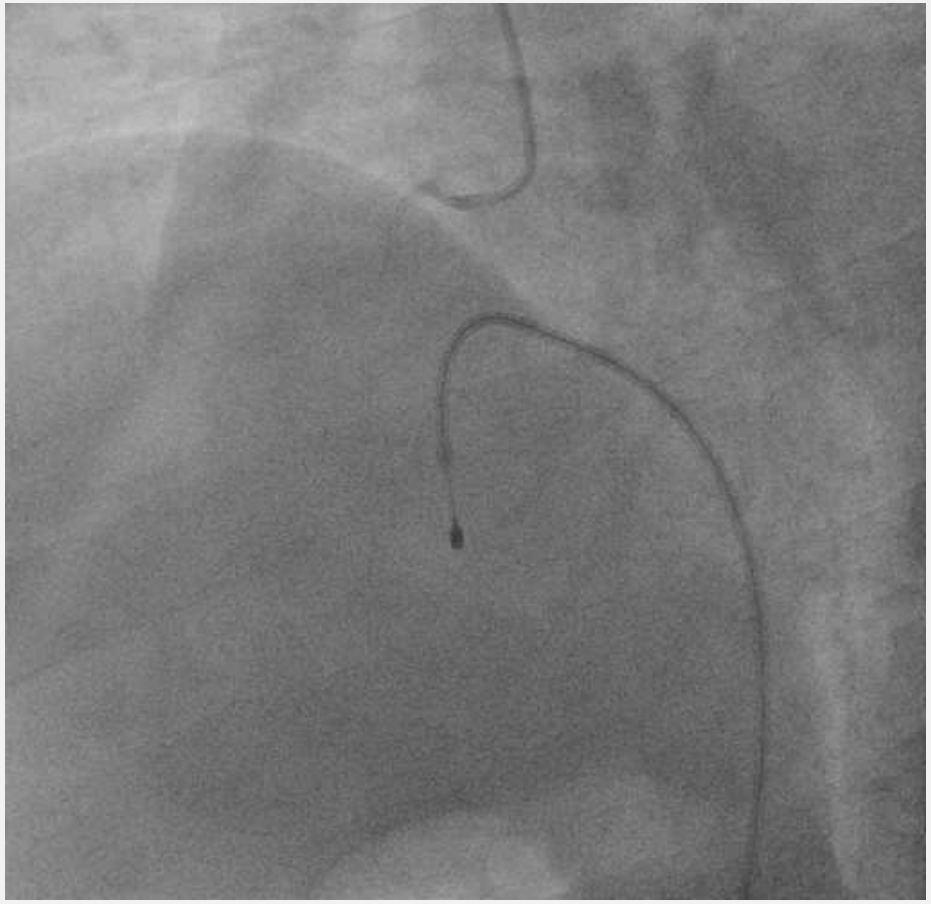
- Hemogram
- Electrolytes
- Liver/ Renal function test
- Random blood sugar
- Thyroid function tests
- ABG, all within normal limits
- Troponin I and NT-Pro BNP levels - Elevated.

DIFFERENTIAL DIAGNOSIS

- Acute coronary syndrome
- Viral Myocarditis
- Inflammatory cardiomyopathy
- Electrolyte imbalance

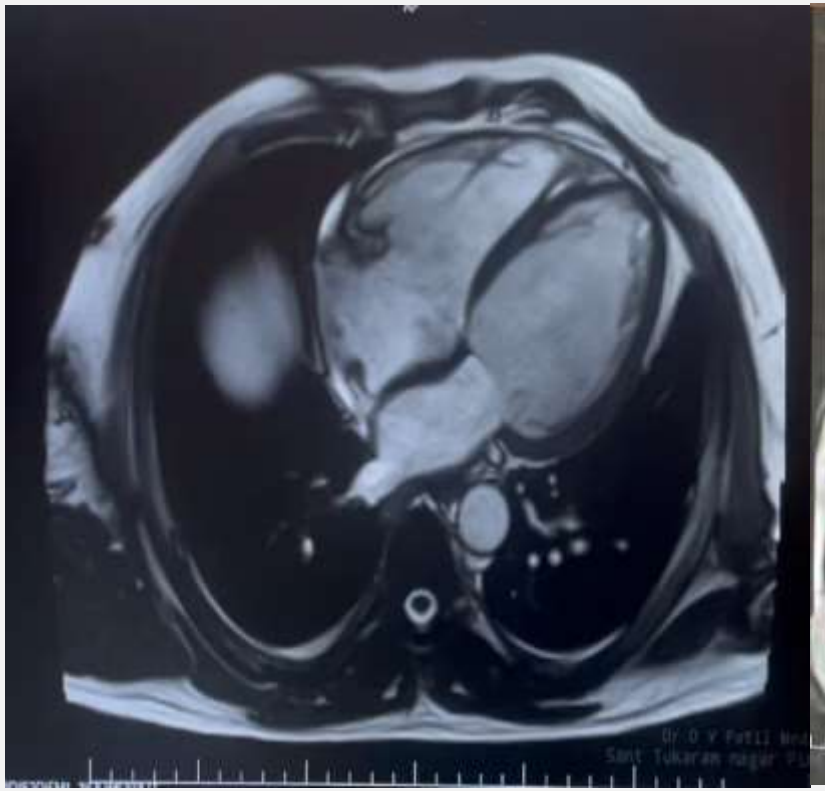
CORONARY ANGIOGRAPHY

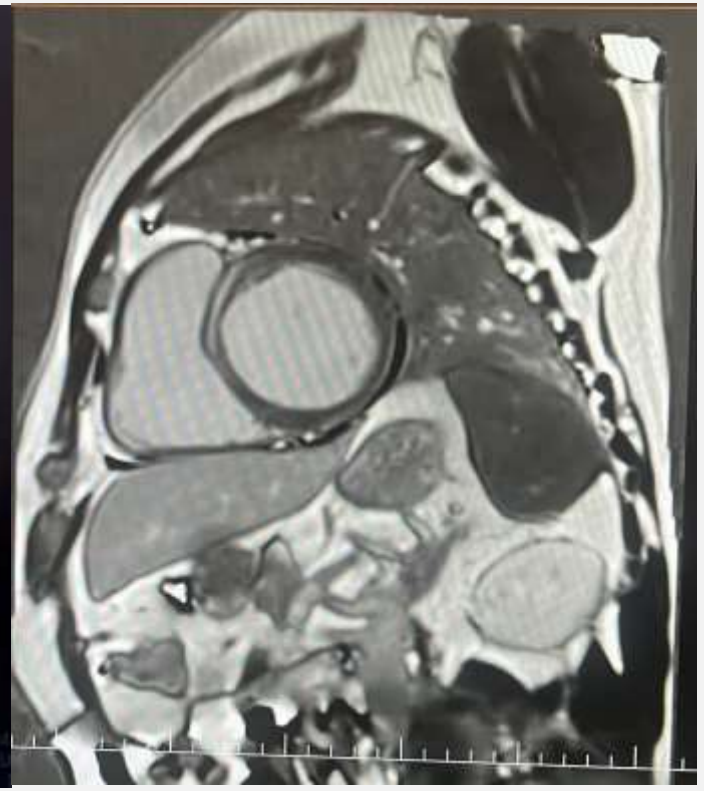




CARDIAC MRI

- Dilated LV with LVEF of 40%.
- Smooth mid myocardial enhancement noted in basal/mid antero-septum, anterior and inferior segments with corresponding mid myocardial oedema.
- No myocardial infarction.
- LGE uptake was 22%.
- Likely ? INFLAMMATORY CARDIOMYOPATHY





SPECIFIC BLOOD INVESTIGATIONS

- ESR and CRP - Elevated
- Serum angiotensin converting enzyme levels - Normal
- Tuberculin test - Negative
- Interferon gamma (TB gold) test - Negative

GA 68- FAPI/ FDG-PET SCAN

- Evidence of increased FAP expression and FDG uptake in the left ventricle consistent with granulomatous and infiltrative aetiology.
- Likely ? SARCOID
- FAPI/ FDG uptake was noted in subcarinal and right lower cervical nodes.

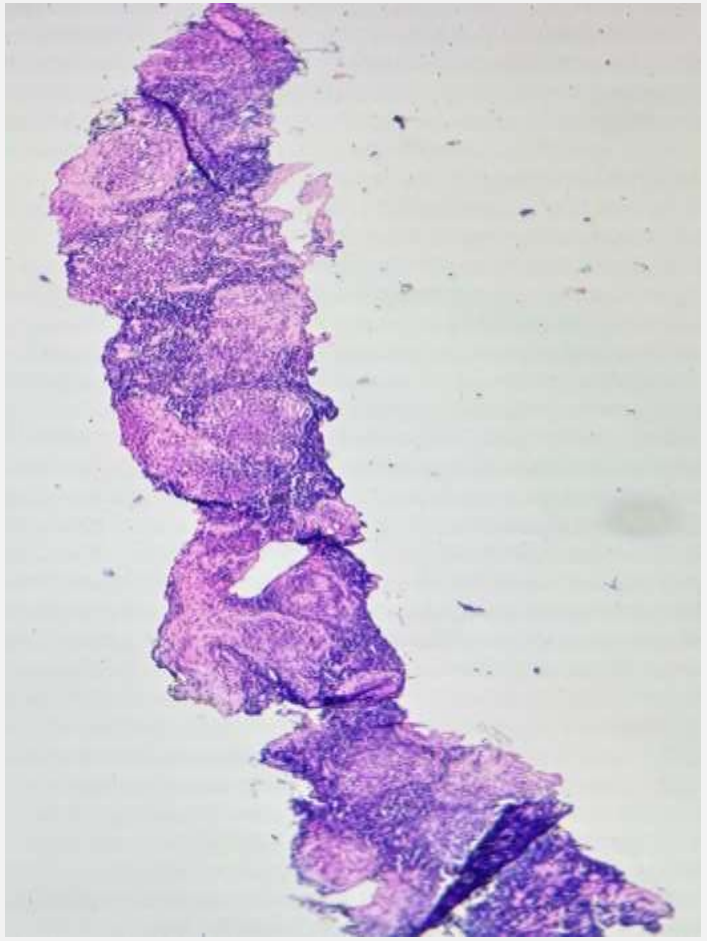
BIOPSY

RIGHT CERVICAL LYMPH NODE BIOPSY

- Core of lymphoid tissue infiltrated by discrete granulomas comprised of epithelioid cells and occasional langhan's giant cells with focal calcifications.
- No evidence of caseous necrosis and malignancy, indicative of chronic non-caseating granulomatous lymphadenitis.

ENDOMYOCARDIAL BIOPSY

- Inconclusive of myocarditis but showed features of fibrosis.



DIAGNOSIS

Based on

- Clinical findings
- ECG
- 2DEcho
- Cardiac MRI
- FDG-PET imaging
- Cervical lymph node biopsy findings

FINAL DIAGNOSIS – CARDIAC SARCOIDOSIS

MANAGEMENT

- Synchronised electrical cardioversion was given to terminate VT.
- Antiarrhythmic medications.
- Beta blockers.
- Counselled for prophylactic implantation of AICD.
- Pulse doses of intravenous methyl prednisolone 1gm/day dose for 3 days.

ON DISCHARGE –

- Oral prednisolone at dose 1mg/kg/day.
- Oral amiodarone at maintenance dose of 400 mg.
- Oral beta-blocker(metoprolol) at a dose of 50mg daily.

FOLLOW UP AFTER 1 MONTH

- Symptom free.
- Resting ECG had multiple ventricular ectopics.
- Holter monitoring had 36% VPC burden.
- Re counselled for AICD implantation.
- Methotrexate was added to the on-going therapy.

DISCUSSION

- Sarcoidosis is a multisystem granulomatous inflammatory disorder of unknown aetiology that predominantly affects the lungs, eyes, skin and lymphoreticular system with cardiac involvement being a rare entity.
- Cardiac sarcoidosis (CS) is an infiltrative cardiomyopathy estimated to be present in 10-25% of patients with systemic sarcoidosis.
- This disease may present as isolated CS or systemic sarcoidosis with cardiac involvement.
- The exact cause remains incompletely understood, may be immune system response to environmental exposure in context of genetic predisposition.
- Majority of patients with CS have asymptomatic involvement, some of them may present with conduction abnormalities, ventricular arrythmias, heart failure and sudden cardiac death (SCD).

- Diagnosis of CS is challenging because of diverse and nonspecific presentations.
- Combination of multimodality imaging and multidisciplinary collaboration is needed to estimate the likelihood of patient having CS.
- Electrocardiogram (ECG) may show nonspecific findings like conduction delays, AV blocks, ventricular premature complexes.
- Ambulatory ECG monitoring may increase the suspicion of CS by identifying transient AV blocks or ventricular arrhythmias.
- Echocardiography may demonstrate reduced left ventricular ejection fraction, regional wall aneurysm, basal septal thinning and abnormal global longitudinal strain.
- Despite limited sensitivity and specificity, ECG and echocardiography are helpful for screening of patients for CS.

- FDG-PET along with CMR is used to assess the disease activity, extracardiac involvement and monitor treatment response.
- CMR imaging is used to localize and quantify areas of late gadolinium enhancement (LGE) as a marker of myocardial involvement from sarcoidosis.
- FDG-PET imaging identifies metabolically active inflammatory lesions.
- Definite diagnosis of CS is made by histopathological examination of cardiac or extracardiac tissue showing non-caseating granulomas containing epithelioid and giant cells.
- Endomyocardial biopsy is considered as gold standard in the diagnosis of CS but has low sensitivity.
- Management of CS aims to slow disease progression, manage arrhythmias, treat heart failure and reduce the risk of SCD.
- Treatment options include immunosuppressive medications like corticosteroids, methotrexate, azathioprine or biological agents.

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Table 10. Diagnostic Guidelines for Cardiac Sarcoidosis

Clinical findings defining cardiac involvement

Cardiac findings should be assessed based on the major criteria and the minor criteria. Clinical findings that satisfy the following 1) or 2) strongly suggest the presence of cardiac involvement.

(See subsection c. "Clinical findings defining cardiac involvement" in the section II.3.2 "Clinical findings suggestive of sarcoidosis involving different organs")

1) Two or more of the five major criteria (a) to (e) are satisfied

2) One in the five major criteria (a) to (e) and two or more of the three minor criteria (f) to (h) are satisfied.

Criteria for cardiac involvement

1. Major criteria
   - (a) High-grade atrioventricular block (including complete atrioventricular block) or fatal ventricular arrhythmia (e.g., sustained ventricular tachycardia, and ventricular fibrillation)
   - (b) Basal thinning of the ventricular septum or abnormal ventricular wall anatomy (ventricular aneurysm, thinning of the middle or upper ventricular septum, regional ventricular wall thickening)
   - (c) Left ventricular contractile dysfunction (left ventricular ejection fraction less than 50%) or focal ventricular wall asynergy
   - (d) ^{67}Ga citrate scintigraphy or ^{18}F-FDG PET reveals abnormally high tracer accumulation in the heart
   - (e) Gadolinium-enhanced MRI reveals delayed contrast enhancement of the myocardium
2. Minor criteria
   - (f) Abnormal ECG findings: Ventricular arrhythmias (nonsustained ventricular tachycardia, multifocal or frequent premature ventricular contractions), bundle branch block, axis deviation, or abnormal Q waves
   - (g) Perfusion defects on myocardial perfusion scintigraphy (SPECT)
   - (h) Endomyocardial biopsy: Monocyte infiltration and moderate or severe myocardial interstitial fibrosis

TAKE HOME MESSAGE

- Inflammatory cardiomyopathies like CS should be ruled out in all patients with recurrent VT.
- Cardiac MRI and FDG-PET are the fundamental imaging modalities for accurate diagnosis of CS.
- High index of suspicion should prompt a search for a positive tissue biopsy in a visualised abnormal area of myocardium or from a non-cardiac site.
- A negative endomyocardial biopsy should not be taken as evidence of absence of cardiac sarcoidosis.
- Due to high risk of recurrent ventricular tachyarrhythmias, strong consideration for prophylactic implantation of AICD should be considered.

THANK YOU