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Dilated Cardiomyopathy
- A Ghost from the Past

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I have nothing to disclose
History

• 42 years lady presented with progressive shortness of breath;
  – class II for last 2 years
  – class III for 2 months
  – subsequently symptomatic even at rest

• Diagnosed a month back as Dilated Cardiomyopathy (DCM) with severe LV dysfunction at another hospital
History…

• Presented to Medical Emergency with acute decompensated heart failure (ADHF)

• Had asystolic cardiac arrest requiring CPR, was successfully revived

• Required mechanical ventilation for 48 hrs and high dose inotropes
History...

No history of

– Angina, palpitation
– Fever, cough, expectoration

Past History

– No history of DM, HT, CAD
– No history of joint pain, rashes
Examination

• **General Examination** –
  – Cold extremities, pallor and raised JVP

• **CVS Examination** –
  – Cardiomegaly
  – Pulse – 110/min, regular, low volume equally palpable in all four limbs
  – BP – 90mmHg systolic (on inotropes)
  – Auscultation – $S_1$ & $S_2$ normal, LV-$S_3$, PSM at apex
Examination...

• **Respiratory System** – Bilateral basal crepitations

• **Gastrointestinal System** – 3 cm hepatomegaly

• **Central Nervous System** – P/extubation - Normal
Routine Blood Investigations

- Hemoglobin – 9.5 gm%
- TLC, DLC, Platelets – Normal
- Peripheral Smear – Normocytic normochromic anemia
- S. Na+ – 124 mmol/l
- S. K+ – 4.8 mmol/l
- S. Ca++ – 9.2 mg/dl
- Bl. Urea – 32 mg%
- S. Creatinine – 1.2 mg%
- LFT’s & S. Protein – Normal
- Troponin I – Negative
Non specific ST-T changes in anterior leads
Chest X-ray

Cardiomegaly with CT ratio 70%
Echocardiography

- Dilated left atrium and left ventricle
- Severe global LV systolic dysfunction, EF 20%
- Moderate mitral regurgitation
- No evidence of AS/Co-A/pericardial effusion
M-Mode

1. IVSd = 0.92 cm
   LVIDd = 5.69 cm
   LVPWd = 1.15 cm

2. IVSs = 1.27 cm
   LVIDs = 5.18 cm
   LVPWs = 1.21 cm

EF: 20%
FS: 9%
Color Doppler
Summary

- 42 year lady diagnosed as “dilated cardiomyopathy”
- Presented with acute decompensated heart failure (ADHF)
- Cardiac Arrest
- Persistent hypotension requiring inotropic support

All poor prognostic markers and harbinger of very poor survival
Unusual findings noted while in ICU

• Absence of tachycardia despite hypotension (on triple inotropes)

• Persistent postural symptoms with a fall of >30 mmHg of systolic BP associated with tachycardia

• Absence of pubic and axillary hairs

• Dry, coarse skin

• Facial depigmentation
Further Investigations

• Iron Studies
  – S. Ferritin – 318.04 ng/ml (29–248)
  – S. TIBC – 321 mcg/dl (251–406)
  – S. Iron – 104 mcg/dl (41–141)

• Thyroid profile
  – S. TSH – 1.92 mIU/ml (0.35–4.94)
  – S. T3 – 0.37 ng/ml (0.58–1.59)
  – S. T4 – 3.56 mcg/dl (4.87–11.72)

Suggestive of central hypothyroidism
Obstetric History

- Post-partum hemorrhage after last delivery 14 years ago
- Failure of lactation
- Secondary amenorrhea

??? Sheehan’s Syndrome
## Pituitary hormonal levels

<table>
<thead>
<tr>
<th>Hormone</th>
<th>Value</th>
<th>Normal Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prolactin (ng/ml)</td>
<td>&lt;0.6</td>
<td>5.18–26.53</td>
</tr>
<tr>
<td>FSH (mIU/ml)</td>
<td>2.26</td>
<td>26.7–133.41</td>
</tr>
<tr>
<td>LH (m/IU/ml)</td>
<td>0.76</td>
<td>10.39–64.75</td>
</tr>
<tr>
<td>GH (ng/ml)</td>
<td>0.5</td>
<td>0.5–17</td>
</tr>
<tr>
<td>Fasting Cortisol (µg/dl)</td>
<td>6.0</td>
<td>5–25</td>
</tr>
</tbody>
</table>

FSH: Follicle Stimulating Hormone; LH: Luteinizing Hormone; GH: Growth Hormone.
Final Diagnosis

Sheehan’s Syndrome with Dilated Cardiomyopathy
DCMP patient presenting with ADHF

Diagnosed as Sheehan’s syndrome

Stress leading to hypoadrenal shock

Cardiogenic + Hypoadrenal Shock
Further management

• IV Glucocorticoids & oral Thyroxine

• Hypotension improved and was weaned off inotropes within 2 days

• Continued on oral hydrocortisone & fludrocortisone, levothyroxine & oral contraceptive pills

• Marked improvement in symptoms

• After stabilization, underwent MRI pituitary
Pituitary MRI

T₂- sagittal image showing CSF filled sella turcica suggestive of pituitary atrophy
Sagittal post-contrast T1 image showing absence of pituitary gland tissue in the sella turcica
Discharge & Follow Up

• At Discharge - NYHA class II
  – On heart failure medicines & hormone replacement

• At 1 month - NYHA class I, LVEF - 45 %

• 3 months later – LVEF was normal
Echocardiography at 3 months
At presentation

Follow-up
At presentation

Follow-up
DCMP patient with EF 20% in Cardiogenic Shock

Survivor of SCD

Diagnosed as Sheehan’s syndrome

Heart failure Rx + hormone replacement

LV function normalized
Sheehan syndrome is a pan-hypopituitarism due to pituitary necrosis occurring usually secondary to post-partum hemorrhage.

In 1937, Sheehan published his classic description of its occurrence following postpartum hemorrhage and vascular collapse.
Pathogenesis

Discussion…

• The extent of pituitary damage determines the severity and time of presentation of pituitary hypofunction

• The presentation varies from acute pan-hypopituitarism leading to cardiovascular collapse to gradual onset of partial or complete pituitary insufficiency over months to years
Discussion…

• Usual symptoms of Sheehan’s syndrome include

  – Failure of lactation  \[\text{Most common}\]
  – Secondary amenorrhea
  – Loss of pubic or axillary hair
  – Slowed mental function
  – Weight gain
  – Low blood pressure
  – Fatigue
Cardiomyopathy & hormone deficiency

- Cardiac abnormalities and hypothyroidism

  - Almost one-third of patients have pericardial effusion, which resolves with correction of the hypothyroid state

  - Additionally reversible cardiomyopathies, dilated or hypertrophic have been rarely reported that improve with replacement of levothyroxine

Cardiomyopathy & hormone deficiency

• Cardiomyopathies have also been reported in patients with **pituitary hypoadrenalism** – with reversal on hormone supplementation


• Reversible cardiomyopathies have also been reported in patients with **growth hormone deficiency**

  Chest 1992;102;326-327
Cardiomyopathy & Sheehan’s syndrome – are they linked??

• Till date only two cases of Sheehan’s syndrome with reversible DCM have been reported
  – A 33 year female, symptomatic within 2 weeks of delivery*
  – A 25 year young female, symptomatic 2 years after her last delivery**

• Our patient was different from the above cases in terms of very late presentation after post-partum hemorrhage

*Shu-Yi Wang J Chin Med Assoc; August 2005 : Vol 68, No 8
Conclusions

• Sheehan’s syndrome adds to the list of “treatable cardiomyopathies”
  – (Importance of digging out relevant past history which cannot be overemphasized)

• Consequently it may be prudent to screen for Sheehan’s syndrome in appropriate clinical settings

• This may masquerade as peri-partum cardiomyopathy