28th Annual Scientific Congress of the Hong Kong College of Cardiology

### Interesting Cases Sharing and Discussion - the cardiologist's perspective

Dr Julia Shi Resident Specialist Department of Paediatric Cardiology Queen Mary Hospital





Referred for management of heart failure in Oct 2012

## **History of AML**

#### July 2003 (13 months old)

- Myelomonocytic AML diagnosed in July 2003 (13 months old)
- UK AML chemotherapy protocol (Hospital A)
- Pre-chemo echo: normal, FS 27.4%, LVIDd 2.65cm
- Cardiotoxic medications received:
  - o Daunorubicin
  - Amsacrine
  - Mitozantrine
- Equivalent doxorubicin 260mg/m<sup>2</sup>
- In clinical remission since November 2003 (almost 9 years)
- Otherwise good past health, no family history of cardiomyopathy



### **Decompensated Heart Failure**

#### Sep 2012 (10 years old)

- Admitted to Hospital B (Regional hospital)
  - Vomiting and diarrhoea x 2 days
  - On & off abdominal pain x 1 month
  - Decrease appetite
  - Puffiness of eyelids
  - Displaced cardiac apex
  - Gallop rhythm, 2/6 PSM at apex radiating to axilla
  - Hepatomegaly
- CXR CT ratio 0.65





### **Investigations for Cause of HF**

- Slight Troponin I (0.16ng/ml) (normal up to 0.04ng/ml)
- Normal CK
- Echo : LVEF 30%, moderate MR

 $\Delta$  ? Acute myocarditis

 $\Delta \mbox{?}$  Chronic heart disease with decompensation

## **Initial Management**

- Inotropes : Dobutamine (10mcg/kg/min) & Milrinone (1mcg/kg/min)
- Captopril, diuretics
- Transferred to Hospital A PICU after 12 days

## **Initial Management**

Hospital A

- Viral study -ve, metabolic screening -ve
- Tn I 0.2-0.3 ng/ml
- Echo : LVEF 36% , severe MR
- Developed hypotension, nausea and vomiting upon weaning of inotropes

Could not wean off inotrope Transferred to us after 4 weeks

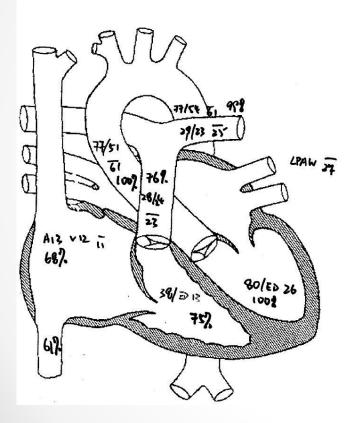
# **Further Investigations**

• Echo : dilated LV (LVIDd 5.2cm)

LVEF ~ 24%, pericardial effusion 4-6mm

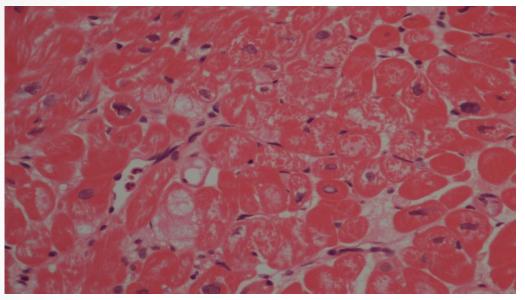
• Borderline BP

#### **Cardiac catheterization**





#### • Endomyocardial biopsy



Courtesy of Dr. WH Shek

Interstitial fibrosis, cytoplasmic clearing suggestive of myofibrillar loss ,vacuolated cells No significant lymphocytic infiltration

#### EM : Nonspecific changes

# Diagnosis

- No feature of myocarditis
- Although nonspecific, features can be seen in anthracycline cardiotoxicity

#### $\Delta$ Late-onset anthracycline cardiomyopathy

## Treatment

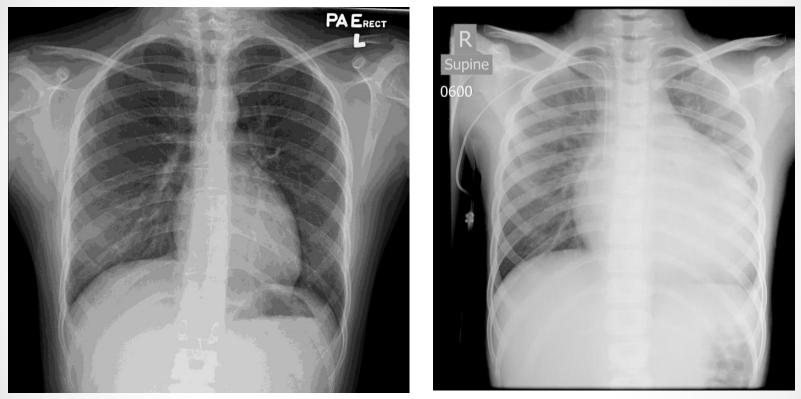
- Levosimendan infusion x 24 hours
- Gradually weaned off inotropic infusions
- Discharged 2 weeks after admission
- Medications : Enalapril, Carvedilol, Digoxin, Frusemide, Spironolactone

- Functional class I-II on last follow-up
- Can join PE lesson, slowly walk up 5FOS

Date	LVIDd (cm)	LVEF %	MR
18/4/2013	4.3	45	Mild to mod
26/3/2015	4.4	52	Mild
27/12/2018	4.3	54	No
8/8/2019	4.3	56-60	Trace



#### CXRs



# Summary

- Late-onset anthracycline-induced dilated cardiomyopathy
- Favorable response to medications
- Functional class  $IV \rightarrow I-II$
- LV reverse remodeling

# WCH, 13y/M

Referred from Macau for post-chemotherapy dilated cardiomyopathy in Jan 2014

### History of AML . Post-chemo DCM

- Known AML post chemotherapy complicated with dilated cardiomyopathy
- On multiple antifailure treatment for 10 years (since 2004)
  - o Digoxin
  - Captopril
  - Carvedilol
  - Diuretics
- AML in remission, no oncology FU now

### **Complicated with Stroke**

- Severe headache and left sided hemiplegia including left facial palsy in Jan 2014, admitted to hospital in Macau
- MRI brain showed ischemia change in Rt basal ganglion region
- Intensive physiotherapy was given with neurological improvement, regained almost full limb power
- Aspirin was started

### **Clinically Worsened HF**

Functional class I-II  $\rightarrow$  III

Physical Findings on admission to our unit

- Displaced apex
- Hepatomegaly 4cm
- Left facial palsy UMN lesion, other CNs NAD
- Left upper and lower limbs hyperreflexia, power 5-/5

## Investigations

• Echo:



- Systolic dysfunction: LVEF 30-35%
- Features of diastolic dysfunction, MV E/A 3.0, E/e' 7.1
- Holter: frequent premature ventricular beats, up to 11% of total, no runs of ventricular tachycardia

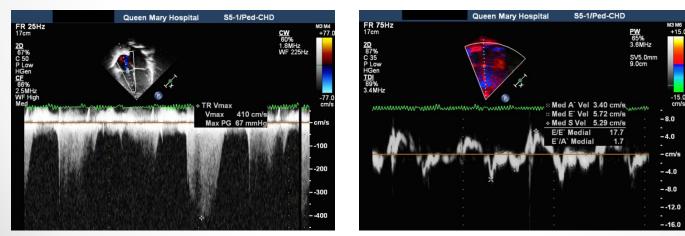
## **Initial Management**

- Inotropic infusion: Dopamine 5mcg/kg/min
- Levosimendan x 48 hours
- Optimized carvedilol, digoxin, enalapril and diuretics doses
- Ventricular ectopics controlled with amiodarone
- Weaned off dobutamine after 2 days
- Discharged after 2 weeks
- Echo upon discharge: LVEF ~43%

# Follow-up Echo

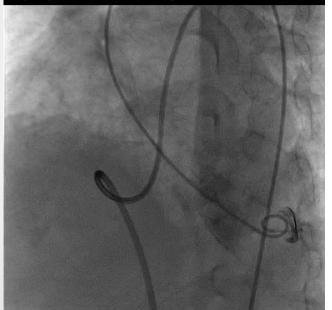
#### Nov 2015

- Dilated left atrium
- Mildly dilated left ventricle, LVIDd 4.3cm
- Mildly impaired LV and RV systolic function, LVEF~50%, TAPSE 16mm
- Evidence of diastolic dysfunction, mitral E/A 2.6, E/e' 13
- Evidence of pulmonary hypertension, TR gradient 67 mmHg



## **Cardiac Catheterization**

ossy compression - not intended for diagnosis



LV Function	LVEF 45.5% LVEDp 26-30mmHg
RV function	Impaired systolic motion RVEDp 13-14mmHg RVSWI 667mmHg/mL/m2
CI	1.8L/min/m2
PASP	70-75% systemic
mPAP	45-48mmHg
PVR/PVRI	7.8-10/ 10.6-13.7

\*Unable to tolerate nitroprusside test due to hypotension

### Outcome

- Added Sildenafil and Bosentan
- Clinically progressive heart failure symptoms, NYHA class III-IV, SBP lowish at 90-100mmHg
- Received transplantation in mainland China (details unclear), and passed away during early post-operative period

# Summary

- Post-chemotherapy cardiomyopathy with prominent diastolic dysfunction
- Poor outcome with progression to pulmonary hypertension
- May require heart or heart-lung transplantation for refractory cases

28th Annual Scientific Congress of the Hong Kong College of Cardiology

### Interesting Cases Sharing and Discussion - chemotherapy-induced cardiomyopathy

Dr Julia Shi Resident Specialist Department of Paediatric Cardiology Queen Mary Hospital

