14th ECHO HONG KONG

Mastering the All-Time Clinical Tool

23 - 26 November 2023

Hong Kong Convention and Exhibition Centre

Workshop II with Hands-on Demonstration

Station E
Diagnosis and Assessment on Cardiomyopathy
(HOCM, Fabry's, Amyloid)

Friday 24 November 2023
09:30 – 12:15
Room S424

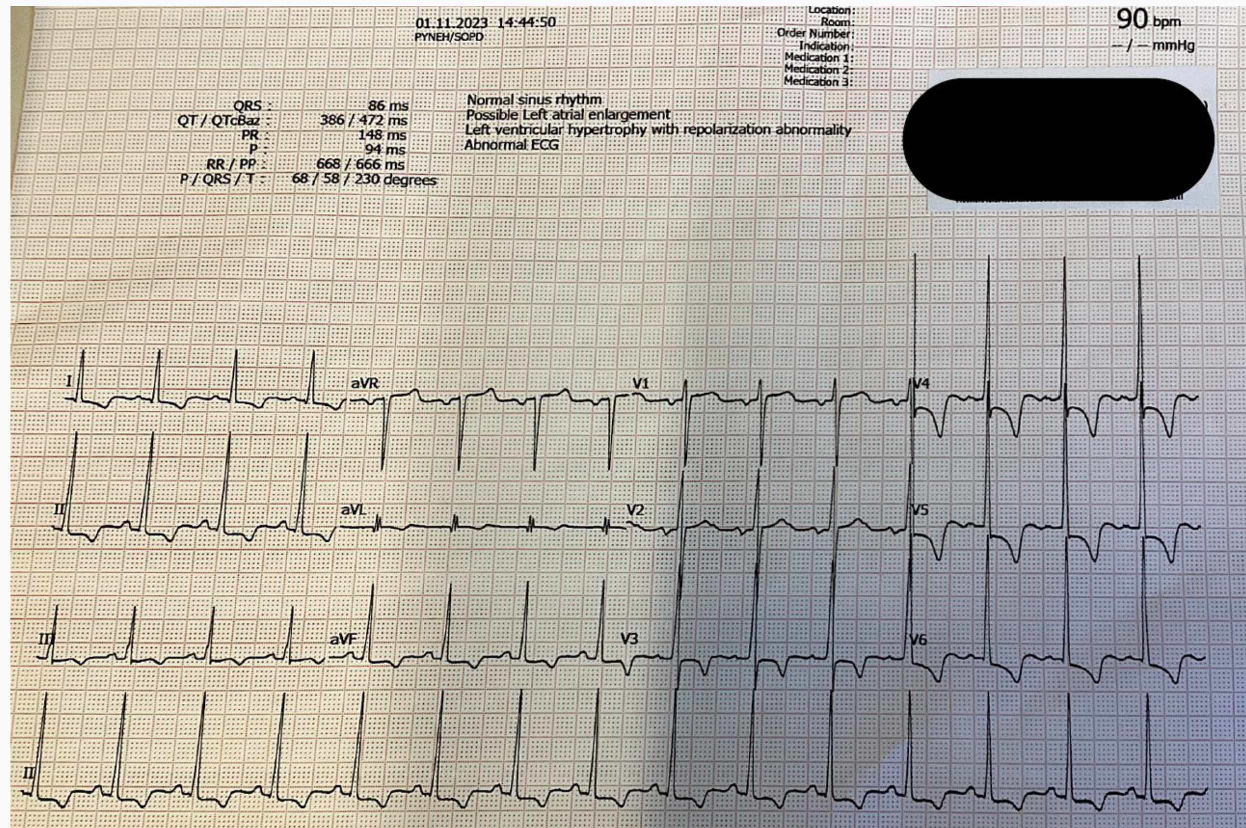
Case E1 - BCM

- 54/M Businessman
- PMHx: HT/Lipid
- FHx: Mother HCM/SCD
- chest discomfort + SOBOE(ET:1-2FOS) x 1yr
- CT coro: mLAD 40% stenosis

Case E1 - BCM (con't)

- ECG:

Giant negative T-wave inversion over precordial and inferolateral leads



Case E1 - BCM (con't)

- Mixed type Apical HCM
- Apical obliteration (septal 1.5cm, apical 2.2cm)
- Mild MR

Case E2 – HKL

- M/74
- TTR-FAP, p.Val50Met, late onset phenotype in non-edemic region
- Well until age 60, started with neuropathy
- NCS: axonal neuropathy
- Sural nerve biopsy: TTR amyloidosis
- Sick sinus syndrome with pacemaker implanted
- PYP scan 2021: Moderately increased update in myocardium (higher than rib), heart to contralateral lung uptake ratio 1.87
- Diflunisal switched to Tafamidis since 2022

Case E2 – HKL (con't)

Echo

- Thickened LV septal and posterior wall
- LV septal diameter 14 mm
- Relatively small LV chamber size
- Midrange LVEF 53% (Simpson A4C)
- Mild LA dilation (LAVI 31 ml/m2)
- Thickened MV leaflets, mean Gr 2 mmHg, trivial MR
- Impaired GLS -14.6%
- Impaired relaxation, E/A 0.7, MV decal time 290 ms, septal E' 3.6 cm/s, lateral 4.35 cm/s, E/E' 22
- RVSP 23 mmHg
- RA and RV leads seen

Case E3 - YWH

- M/58. Retired Policeman. Non-smoker. Social drinker.
- Medical History: Minimal CAD by CTCA 9/10/2021 during health check. Unremarkable past history otherwise.
- Drug history: No regular medication
- Family history: No family history of heart disease / SCD
- First episode syncope on exertion after playing soccer for 15 minutes
- Syncope duration: 1-2 minutes
- Denied any chest pain / palpitation / SOB before syncope. No witnessed convulsion.
- Noted pulseless by peer players – bystander CPR for 1 minute
- Regained full consciousness before ambulance arrival

Case E3 – YWH (con't)

- Physical examination
  - BP/P Stable
  - CVS Examination: Unremarkable
  - Neuro Examination: Unremarkable
  - ECG: Ectopic atrial rhythm . VR 44bpm. Normal PR/QTc. LVH by voltage criteria. T Wave Inversion V3-6.
  - Blood tests electrolytes unremarkable. LDL 3.35.
  - Bedside Echo (V-Scan): Normal LVEF. No RWMA. No obvious valvular lesion. No pericardial effusion.

Case E3 – YWH (con't)

- Echo
  - Mildly dilated LA (4.49cm)
  - Asymmetrical hypertrophy over antero-septum/anterior wall (IVSd up to 2.1cm).
  - Normal LV/RV systolic functions, LVEF ~70% with no RWMA.
  - Impaired LV diastolic function
  - Aortic root size is normal. AV is trileaflet. There is no resting LVOTO or SAM. (LVOT mean/max PG 1.48/2.89 mmHg. AV mean/max PG 4.37/8.07mmHg)
  - Mild TR/MR and trivial PR.
  - No pericardial effusion.
  - Impression: HCM.

Case E3 – YWH (con't)

- CTCA
  - Mild coronary arteries calcification.
  - R dominant.
  - LM/LCx/OM1/OM2/RCA/RPL/PDA: Unremarkable.
  - Proximal LAD minimal disease (<25%) with calcified and non-calcified plaques.
  - Mid LAD myocardial bridging. D1-D3 unremarkable.
  - Extra-coronary: AV trileaflet. Aortic root not dilated. No intra-cardiac shunt. Superior segment of LAA/LSPV not seen. Pulmonary veins drainage normal otherwise. No pericardial effusion.
  - CTCA images reviewed: Significant LV hypertrophy – but not mentioned in the CTCA report

Case E3 – YWH (con’t)

- MRI Heart
  - BP/P Stable
  - Normal chamber sizes. Asymmetrical LV wall thickening involving basal anterior, basal anteroseptal, mid anterior, mid anteroseptal segments. The segments of LV wall thickening measured up to 15mm. Other parts of LV segments have preserved wall thickness. The mid ventricular IVS was 12mm.
  - Normal LV systolic function. LVEF 62.4%.
  - No LV/RV RWMA. No focal dyskinesia or LV aneurysm.
  - No intracavitary lesion /mass / thrombus. No significant valvular abnormality.
  - No myocardial ischemia under adenosine stress (0.14mg/kg/min; resting HR 54bpm; peak stress HR 66bpm).
  - No microvascular dysfunction.
  - No LGE to suggest old infarct/ scar or fibrosis.
  - No pericardial effusion.
  - No extra-cardiac abnormality.
  - Impression: Hypertrophic cardiomyopathy. (Ddx of infiltrative change or acute inflammatory are considered not very likely).

Case E3 – YWH (con't)

- Progress
  - Subcutaneous ICD (Boston Emblem MRI) implanted on 4/3/2023 - R parasternal implant; 2 incision technique with intermuscular pocket
  - Paroxysmal AF: S-ICD detected PAF with inappropriate shock on 7/8/2023
  - Current drugs: Concor 1.25mg QD. Lipitor 20mg QD
  - Last seen well 6 months post cardiac arrest