

ESIM Case Presentation



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Case presentation



76-year old male patient

Referred for elaboration cardiomyopathy

Symptoms: Dyspnea

Diarrhea

Syncope with eyelid haematoma

Case presentation



History

- Appendectomy
- Bilateral meniscus surgery
- Liver cyst, benign?
- Atrial fibrillation
- TIA
- Hypertrophic cardiomyopathy

Medication

- Diuretic:
Burinex 1mg/d
- ACE-inhibitor:
Lisinopril 20 mg/d
- Beta-blocker + diuretic:
bisoprolol 2,5mg +
hydrochloorthiazide 6,25mg
- Anticoagulant
Marevan (~INR)

Clinical investigation



General: Eyelid haematoma, L 183cm; W 72kg

Heart: Heart rate: 77/min, Blood pressure: 100/80mmHg

S1S2, irregular, no cardiac murmurs

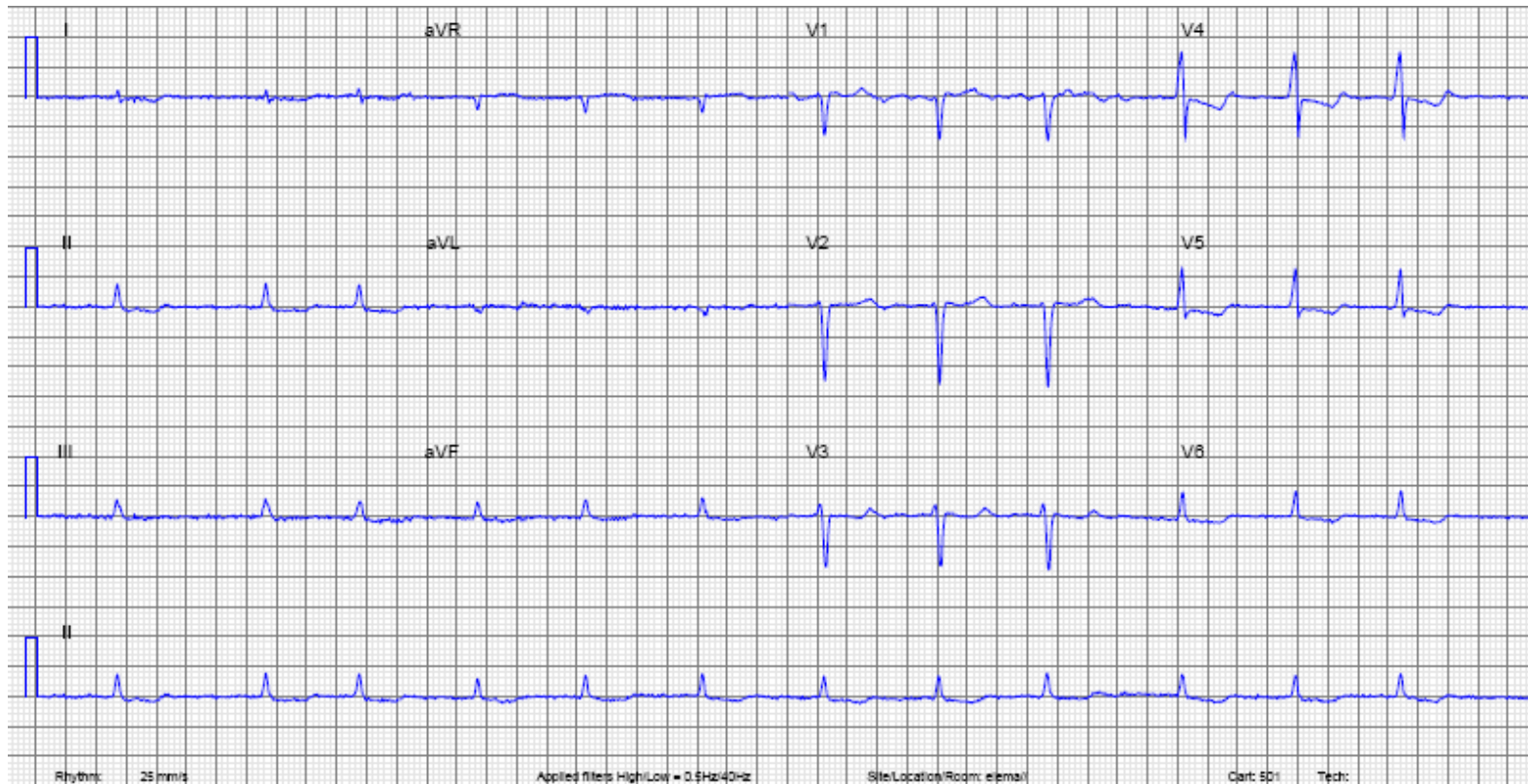
increased CVP (12 cm)

Lung: Bilateral normal respiratory sounds

Liver: normal palpation, positive hepatojugular reflex

Limbs: Pitting oedema lower limbs

ECG





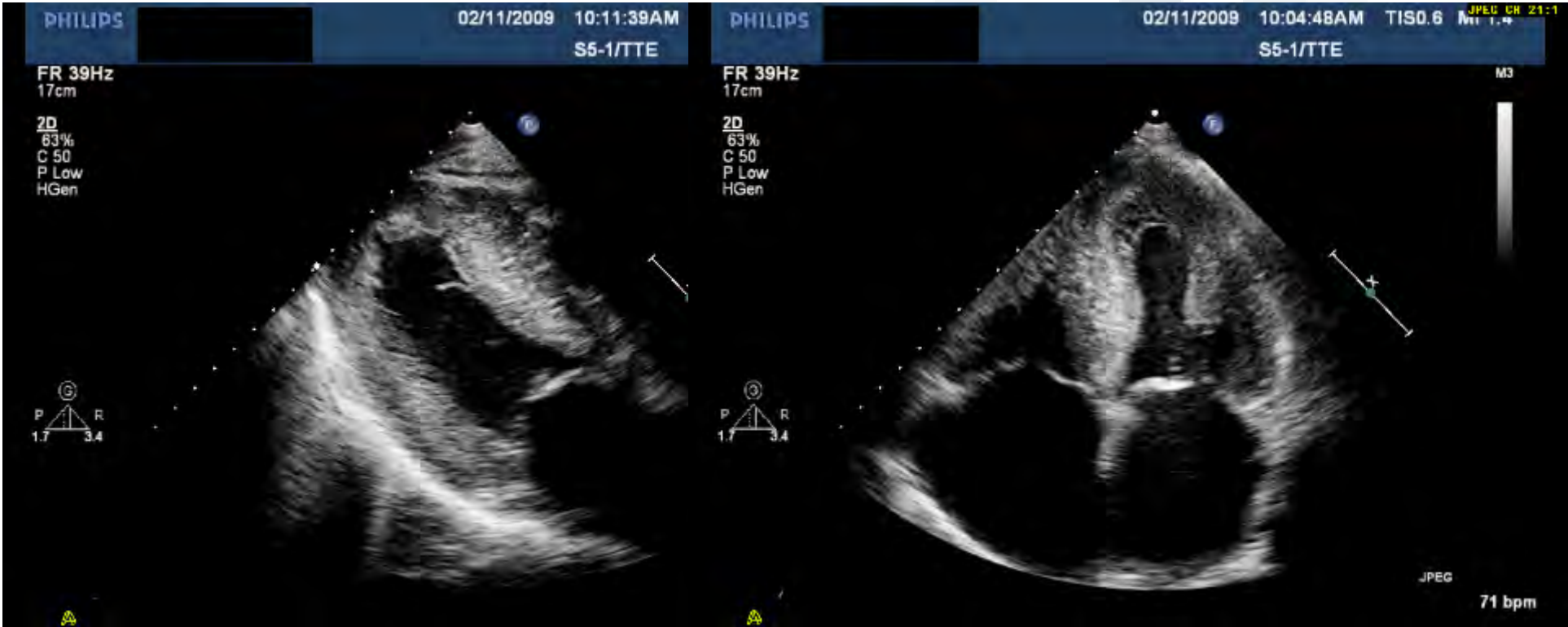


TABLE 64–6**Classification of Types of Restrictive
Cardiomyopathy According to Cause****Myocardial*****Noninfiltrative***

Idiopathic cardiomyopathy*
Familial cardiomyopathy
Hypertrophic cardiomyopathy
Scleroderma
Pseudoxanthoma elasticum
Diabetic cardiomyopathy

Infiltrative

Amyloidosis*
Sarcoidosis*
Gaucher disease
Hurler disease
Fatty infiltration

Storage Disease

Hemochromatosis
Fabry disease
Glycogen storage disease

Endomyocardial

Endomyocardial fibrosis*
Hypereosinophilic syndrome
Carcinoid heart disease
Metastatic cancers
Radiation*
Toxic effects of anthracycline*
Drugs causing fibrous endocarditis (serotonin, methysergide,
ergotamine, mercurial agents, busulfan)

*These conditions are more likely than the others to be encountered in clinical practice.

From Kushwaha S, Fallon JT, Fuster V: Restrictive cardiomyopathy. *N Engl J Med* 336:267, 1997. Copyright 1997, Massachusetts Medical Society.

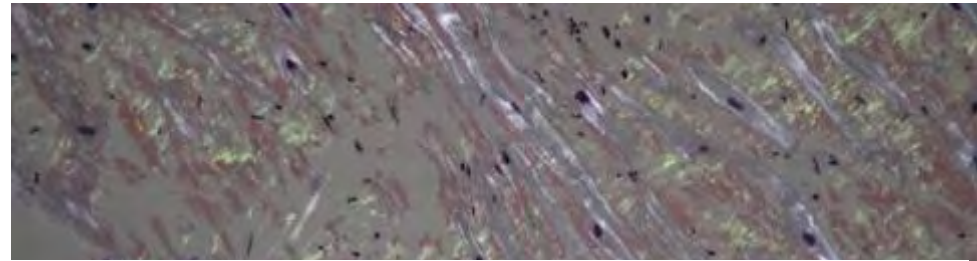
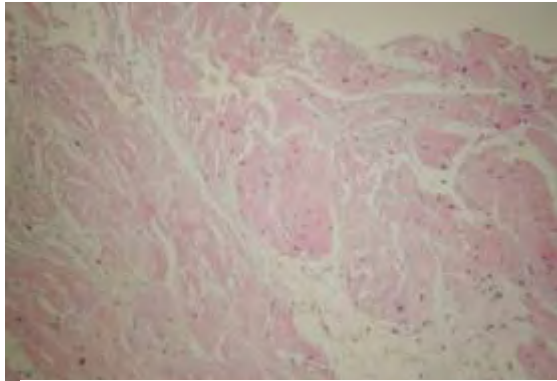
Diagnostic work-up



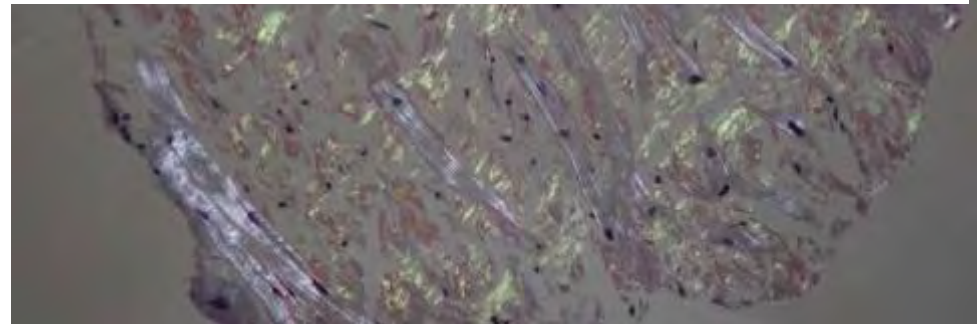
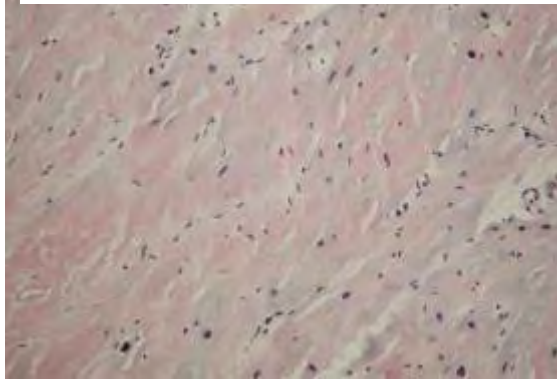
Coronarography:

Decreased systolic left ventricle function. No critical coronary lesions.

Endomyocard biopsy



Cardiac amyloidosis (non-AA)



Diagnostic work-up (2)

- Blood tests:
 - » GFR MDRD 50ml/min/1.73m²
 - » LDH: 872 U/l (> 618), Alk Fosfatase: 115 U/l (>95), gamma-GT: 147 U/l (>45)
- Light chain proteins in serum / urine: negative
- X-ray spinal column: osteoporosis, no pathologic lesions
- Bone marrow biopsy: no arguments for malignancy
- Echo abdomen: Heterogenic mass in right liver lobe, focal defect lesions in left liver segment
- Ileocoloscopy with biopsies:
 - » congo-red staining: negative
 - » Tumor rectum: invasive adenocarcinoma

Diagnosis

- Restrictive cardiomyopathy caused by non-AA amyloidosis
- Adenocarcinoma rectosigmoid
- Liver metastases?

Treatment

- Supportive treatment: diuretics, ACE-inhibitors, beta blockers, anticoagulant
- Referral to gastro-enterology department for staging: pT3pN2pM1 => low anterior resection followed by palliative chemotherapy

Cardiac amyloidosis: learning points

- Amyloidosis as a cause of hypertrophic cardiomyopathy
- Extra-cardial symptoms (neuropathy, hepatomegaly, renal dysfunction)
- Biopsies of suspect tissue essential

Thank you for your attention!

