

Congestive heart failure with rapid onset

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Clinical

- B. D. , male, 63, non-smoker, not consuming alcohol
- Present complaints:
 - dyspnoea with minimal activity
 - weight loss (12 kg/last 2 months)
 - progressive asthenia
 - dysuria

History disease

- Congestive heart failure stage IV NYHA with no history of arterial hypertension or ischemic cardiac disease for about 1 year
- Left adrenalectomy for suspicion of pheochromocytoma 2 weeks prior to admission
- Massive right pleural effusion

Physical examination

- Pale skin,
- dullness to percussion on right site+ inaudible breath sounds,
- BP=110/80 mm Hg, HR-80/min, diminished heart sounds,
- the rest of the exam is normal

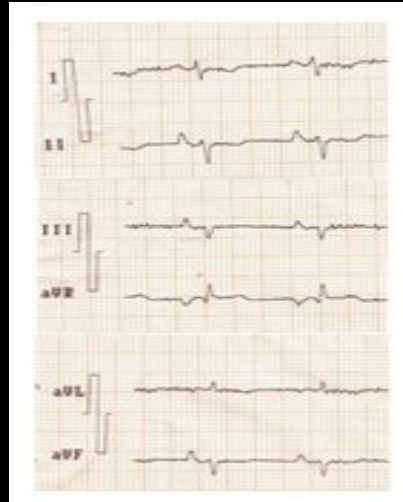
Laboratory evaluation

- inflammatory syndrome (fibrinogen-570 mg/dl, CRP-40 mg/dl)
- increased alkaline phosphatase 340 U/l and LDH 500 U/l
- pANCA, cANCA, C3, C4-normal
- hematuria, proteinuria < 1g/24h urine

- pleural effusion:
 - bacteriological exam is negative
 - cell count: 99% lymphocytes , 1% neutrophils , 100 elem/mm³
 - transudate

Diagnostic procedures

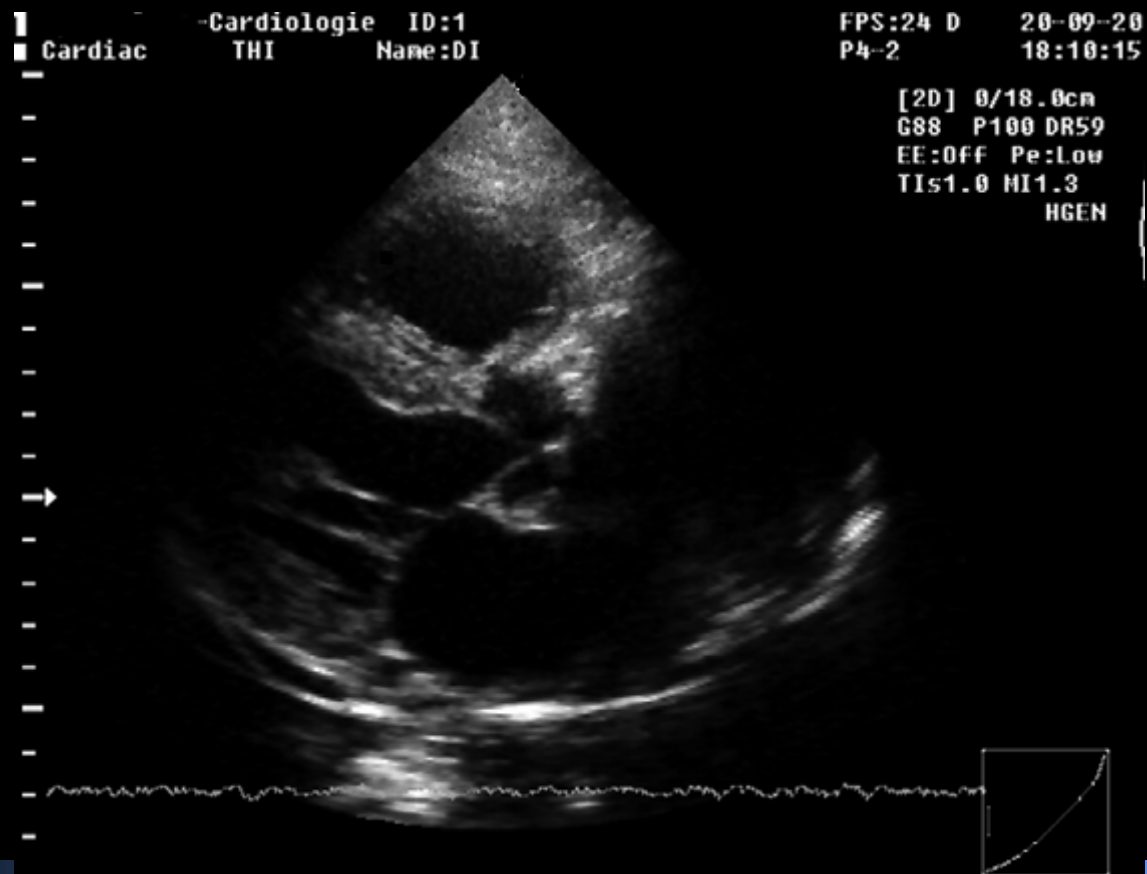
- ECG-low QRS voltage, 80/min



- Chest X- Ray: right pleural effusion in medium quantity
- Abdominal ultrasound: Normal except urinary bladder with thickened walls

Echocardiography

- Typical features of restrictive cardiomyopathy: thickening of the LV walls and restrictive pattern of transmitral flow



What is the next step?

- Lack of risk factors for cardiac disease
- A low voltage on ECG
- An increased thickness of LV walls

Diagnosis?

- Lambda chains in serum increased 20xN
- Kappa chains in serum- normal
- Kappa/lambda ratio-decreased
- Urinary paraprotein present (Bence-Jones)

? Diagnosis: AL Amyloidosis?

Confirmation

- Abdominal fat biopsy-the characteristic apple green birefringence seen when the tissue is stained with Congo Red and viewed under polarised light



Treatment

- Melphalan and Dexametasone were started
- Urology consult- cystoscopy showed urinary bladder tumor that was removed by endoscopy



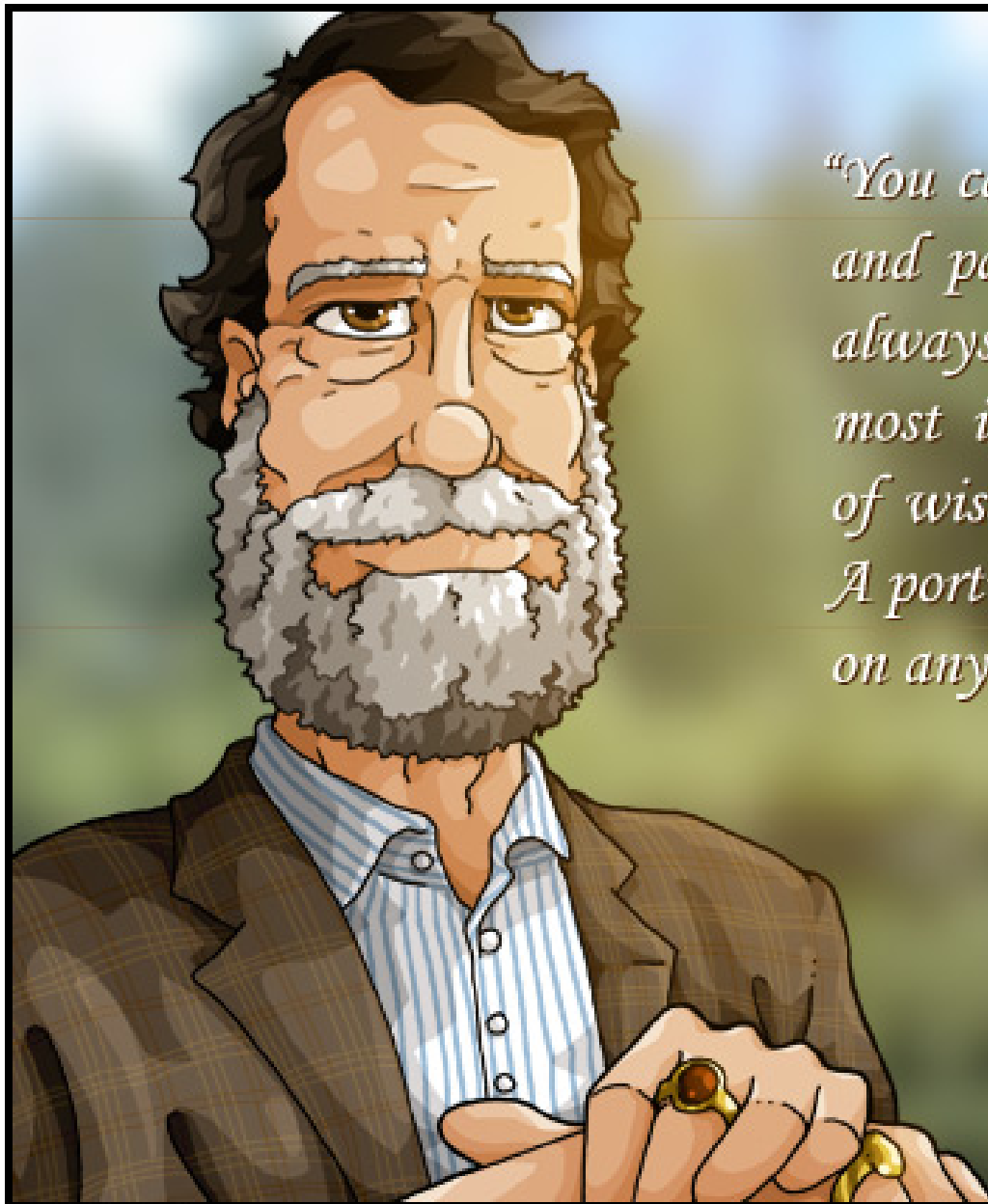
Histology exam+ Congo staining of the bladder tumor +
suprarenal excised prior to admission- amyloid deposits

Evolution

- Introduction in a study treatment with bortezomib
- Worsening of CHF to death

Conclusions

- Final diagnosis: Cardiac amyloidosis
- Should not be overlooked especially in a patient with multi systemic involvement
- The main clue is the inverse relationship between ECG voltage and LV mass
- Thinking at this diagnosis could have been avoided the unnecessary adrenalectomy
- Cardiac involvement has a very poor prognosis-survival less than 8 months



"You can never know everything and part of what you know is always wrong. Perhaps even the most important part. A portion of wisdom lies in knowing that. A portion of courage lies in going on anyway."

*~ Robert Jordan
1948 - 2007*