63 y.o.

- Palpebral oedema and mild legs oedema
- In 2007 BW increase by 10 kg in a few months (oedema of the hands, face, legs, pleural effusion and mild pericardial collection)
Medical history:

- HTA since 1991 treated by β blockers and ACE inhibitors
- Diabetes type II since 1991 treated by Metformin, Amarylle and insulin
- CPAP at night since 1990 (despite uveal ablation)
- Duodenal ulcers in 2006
- Vertebral fracture 1981 (car accident)
- Stent right coronary in 2006
- Chronic prostatitis
- Hypogonadism
- Chronic depression (Fluoxetine 40 mg/day since the age of 30)
- IgG Kappa monoclonality known since 2005
- Cigarettes: 20/day (stop 2006)
- Alcohol in the past (stop 1985)
Physical exams:

- BP 120/80  HR 68/min  T° 36.5  95 kg/174 cm
- No organomegaly – ascites (?)
- Neurology: normal clinically (reflexes, …)
- Major face/hand/legs oedema
- Hand hyperpigmentation (?)
- Hair loss (axillary, pubic, sourcil)
Medications:

- Diabetic and low salt diet
- Metformin 850: 2/day
- Mixtard 30/70: 50-50
- Prozac 20 mg: 2/day
- Omic 0.4 mg: 1/day
- Asaflow 80 mg/day
- Atenolol 50 mg/day
- Coversyl 5 mg/day (Atacand 8 mg/day)
- Burinex 1 mg/day
- Canrenol 50 mg/day
- Omeprazol 20 mg/day
- Simvastatin 40 mg/day
- Alendronate 70/week
Exams (Biology):
Normal cardiac/hepatic and renal function (no proteinuria)

- **APTT** 28.2 s
- **Glycemia** 117 mg/dl
- **PT** 82%
- **Urea** 37 mg/dl
- **Fibrinogen** 400 mg/dl
- **Creat** 0.8 mg/dl
- **CRP** 0.4 mg/dl
- **Uric acid** 6.6 mg/dl
- **WBC** 9.2 $10^3/\mu l$
- **Na** 141
- **Hb** 11.6 g/dl
- **Cl** 106
- **Plat** 331 $10^3/mm^3$
- **CO_2T** 24
- **MCV** 88.5 fl
- **Bilirubin** 0.15
- **PN** 55%
- **PAS** 56 (N 56-119)
- **Lym** 23.7%
- **GGT** 22 (N 8-61)
- **Sedi** 8 mm/hr
- **SGOT/SGTT** 20/19
- **Cholesterol** 151 mg/dl
- **LDH** 138
- **TG** 90 mg/dl
- **CPK** 15
- **HDL** 61 mg/dl
- **HBA_2** 8%
<table>
<thead>
<tr>
<th>Exams (Biology):</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Calcium</strong></td>
<td>9.2 mg/dl</td>
</tr>
<tr>
<td><strong>Phosphor</strong></td>
<td>3 mg/dl</td>
</tr>
<tr>
<td><strong>Prot</strong></td>
<td>6.1</td>
</tr>
<tr>
<td><strong>Albumin</strong></td>
<td>3.8 g/dl</td>
</tr>
<tr>
<td><strong>Gammaglobulin</strong></td>
<td>0.6 g/dl</td>
</tr>
<tr>
<td><strong>Testosterone</strong></td>
<td>2.1 (N 3-10 ng/ml)</td>
</tr>
<tr>
<td><strong>FSH</strong></td>
<td>23 (N 2-8)</td>
</tr>
<tr>
<td><strong>LH</strong></td>
<td>6 (N 2-10)</td>
</tr>
<tr>
<td><strong>Prolactin</strong></td>
<td>375 (N &lt; 325)</td>
</tr>
<tr>
<td><strong>Cortisol</strong></td>
<td>150 ng/ml</td>
</tr>
<tr>
<td><strong>TSH/T4/T3</strong></td>
<td>N</td>
</tr>
<tr>
<td><strong>NT pro BNP</strong></td>
<td>50 pg/ml (N &lt; 300)</td>
</tr>
<tr>
<td><strong>Aldosteron</strong></td>
<td>61 (40-330 pg/ml)</td>
</tr>
<tr>
<td><strong>Renin</strong></td>
<td>77 (3-33 ng/l)</td>
</tr>
</tbody>
</table>

**Monoclonal gammopathy IGg Kappa**
Exams (Biology):

Urine: UNa > 200 mEq/day (bed rest)
Prot 0.1 g/g creatinin  Urine Bence-Jones [——]

Autoimmune: negative (coeliac, …)

Other exams:
- Lymphography  N
- Bone IRM  N
- α1 antitrypsine  Abnormal (5 times upper limits)
- Pet/CT  (bowel captation) (?)
- Video camera  N-mild oedema
- Biopsia  No telangectasia, no whipple
Diagnostic:  

- Chronic capillary leak syndrome? (Chronic Clarkekson Syndrome?) (Never decrease in protein or increase in Ht) (?)
- Incomplete form of POEMS syndrome (?)
- Other ?
### Diagnostic criteria for POEMS syndrome

<table>
<thead>
<tr>
<th>Major criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polyneuropathy</td>
</tr>
<tr>
<td>Monoclonal plasma cell disorder</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Minor criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sclerotic bone lesions</td>
</tr>
<tr>
<td>Castleman's disease</td>
</tr>
<tr>
<td>Organomegaly (spleen, liver, or lymph nodes)</td>
</tr>
<tr>
<td>Volume overload (peripheral edema, pleural effusion, ascites)</td>
</tr>
<tr>
<td>Endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, pancreatic)</td>
</tr>
<tr>
<td>Skin changes (hyperpigmentation, hypertrichosis, plethora, hemangiomata, white nails)</td>
</tr>
<tr>
<td>Papilledema</td>
</tr>
</tbody>
</table>

**Diagnosis requires the presence of**

- Two major criteria **plus**
- At least one minor criterion (excluding diabetes or thyroid abnormalities)

### Classical criteria: POEMS syndrome

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Mayo series (n = 99)</th>
<th>Soubrier, et al. (n = 25)</th>
<th>Nakanishi, et al. (n = 102)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polyneuropathy</td>
<td>100</td>
<td>100</td>
<td>100</td>
</tr>
<tr>
<td>Organomegaly</td>
<td>50</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>24</td>
<td>68</td>
<td>78</td>
</tr>
<tr>
<td>Splenomegaly</td>
<td>22</td>
<td>52</td>
<td>35</td>
</tr>
<tr>
<td>Lymphadenopathy</td>
<td>26</td>
<td>52</td>
<td>61</td>
</tr>
<tr>
<td>Endocrinopathy</td>
<td>67</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>3</td>
<td>36</td>
<td>25</td>
</tr>
<tr>
<td>Monoclonal plasma cell disorder</td>
<td>100</td>
<td>100</td>
<td>75</td>
</tr>
<tr>
<td>Skin changes</td>
<td>68</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Hyperpigmentation</td>
<td>46</td>
<td>48</td>
<td>93</td>
</tr>
<tr>
<td>Hypertrichosis</td>
<td>26</td>
<td>24</td>
<td>74</td>
</tr>
</tbody>
</table>

This table indicates the percent of patients from three major series of patients with POEMS who satisfied each of the original five components of this syndrome. NA: data not available.

Treatment: ?

2007

• Prednisolone 30 mg/day one week
  15 mg/day one week
  7.5 mg/day one week

⇒ BW decrease by 7 kg!

Relapse after a few weeks, new administration with low efficacy

• Dexamethason (like in myeloma; 40 mg/day/4 days and 4 days without repeated 3 times)
  ⇒ BW decrease again by 6-7 kg!

Relapse without efficacy
Treatment:  

2007

- Theophylin (without efficacy)
- Cyclosporin (without efficacy)
Treatment:

2008

- γglobuline/IV (1 g/kg two days): no effects
- Avastatin (no efficacy; VEGF normal)
Treatment: ?

2009

- Plasmapheresis: good result at the beginning then escape
- Direct fluid removal
Treatment:  

2009

- Fluid analysis of leg oedema
  (Na 140 – Cl 115 – CO₂T 25 – K 4 – Protein 0.4 g/dl)
  (Same in pleural fluid)
  (Insulin normal in the fluid)

- Very high doses of insulin are needed when the patient is in anasarc (> 200 U/day)
Treatment: ?

2010

- Improvement by a better salt restriction diet
  - Decrease of 5 kg (± 90 kg)
- Actually Canrenol 50 mg/dl
  Fludex 1/day
  Diamox 500 mg x 2/2 days
  ⇒ BW 84 kg (still oedema)