

♀ 53 y.o.

- Chronic major oedema since the age of 10 years
- At the age of 20 years she is followed by the Department of Internal Medicine

Biology at the age of 20 y.o.

	29/10/80	25/11/80	29/11/80	09/12/80	23/12/80	26/12/80	11/08/93
Total proteins (mg/dl)	4.3	3.4	3.4	4.6	3.9	4.1	3.1
Albumin (mg/dl)	-	1.4	-	-	-	1.7	1.2
Ca (mg/ dl)	7.1	6.3	5.4	7.7	7.5	7.0	7.4
Mg (mg/dl)	1.6	1.1	-	1.8	-	-	1.4
P (mg/dl)	-	3.5	-	4.2	3.9	3.7	4.4
WBC (10^3)/mm ³	13.6	4.1	-	-	-	4.5	4.9
% lymphocytes	1	4	-	-	-	1	3
% poly neutro	96	84	-	-	-	94	91
RBC (10^6)/mm ³	5.13	4.9	-	-	-	4.74	
Hemogl. (g/dl)	15.2	14.9	-	-	-	13.9	13.2
Hematocrite vol. %	44.7	42.8	-	-	-	41.4	
SGOT/SGPT	N	N					N
Bilirubin	0.5						
Cholesterol (mg/dl)						120	100
Triglyceride (mg/dl)						50	66
Gammaglobulin (g/dl)						0.5	0.5
Uric acid (mg/dl)						2	2.5
Urine protein						Neg.	Neg.
Na (mEq/l)						8	

Diagnostic?

Lymphography

- Multiple lymphoduodenal fistulas (hypoplasia of the thoracic chanel)
- Protein-losing enteropathy (Alb-I*131: Half-life 2.5 days) (N = 8 to 17 days)

Evolution I

- Many abdominal operations to try to close the fistulas (transient efficacy of a few month – Prot 4.5-5 g/dl)
- Chylous ascites (not after 20 y.o.; peritonitis)
- Frequently hospitalised for severe hypocalcemia (tetany) (despite 1-alpha cholecalciferol; calcium, DCure IM)
Good control with Rocaltrol
- Frequent intravenous administration of Albumin/Calcium/Magnesium/Vitamins (each 4-6 weeks)
- Chronic treatment with diuretics (Spironolactone/Moduretic/Lasix) (Aldosterone 2500 pg/ml) ($\text{N} < 300$)

Evolution II

- 27 y.o.
 - Weber Christian (Panniculitis of the arms)
(Fever during a few weeks)
 - For the first time lymphocytes 30%!
- 40 y.o.
 - Gammopathy IgG Kappa and Lambda (!)
 - Total protein 5.5 g/dl (!) Albumin 2.5 g/dl (!)
Cholesterol 250 mg/dl (!) G Glob 0.7 g/dl Uric 7 mg/dl

Evolution III

- 46 y.o.

- WC 5400 Uric acid 6.1 Alb 3 g/ml
- PN 68% Urea 30 TP 5.8 g/dl
- Ly 25% Creat 0.5 G Glob 0.9 g/dl
- Calcium: 8 mg/dl PTH: Normal Aldosterone: 1200
- Extensive arms warts (despite better immunity)
(Aldara – No efficacy)
- Chronic leg oedema (++)
- Development of a large scarpa adenopathy
(about 5 cm in a few months)
- Ablation – Diagnostic?

Diagnostic

- Localised “Primary amyloidosis”
(bone marrow; skin biopsy; bone IRM, cardiac echography, Aprotinine-Tc99 scan analysis)

Evolution IV

- Lymphocela (6 months)
- 4 years after the diagnosis of primary amyloidosis, still no other localisation
- Last protein 5 g/dl, lymphocytes $600 \times 10^3 / \text{mm}^3$