

Portuguese Clinical Case Presentation

Greenwich | Saturday 12-09-2009





Clinical History (1)

88-year-old man, caucasian, farmer

- ▶ Full autonomy for daily activities.

Past medical history

- ▶ Hypertension. Benign prostatic hypertrophy.
- ▶ Multiple minor surgeries.
- ▶ No documented history of allergies. No alcohol consumption since the last 3 years (past consumption of 50 g/day). No history of cigarette smoking since 1980.
- ▶ Usual medication: Aspirin® 100 mg id, Telmisartan 80 mg id,

Family history

- ▶ Irrelevant.





Clinical History (2)

88-year-old man, caucasian, farmer

Emergency Department

- ▶ Six-month history of : 1) **progressive leg edema** 2) **face swelling** 3)

INTERNAL MEDICINE DEPARTMENT ADMISSION

- ▶ **Physical Findings:** TA - 100/50 mmHg; soft, nontender abdomen with fluid wave; leg and face edema.
- ▶ **Laboratory Results:** **haemoglobin increase** (19,2 g/dL), **haematocrit increase** (55%), **plasmatic creatinine increase** (1,7 mg/dl), **leukocytosis** ($13 \times 10^9/L$) and **reduced serum albumin** (2,6 g/dL). **Two times increase of AST, ALT and GGT.**
- ▶ **Chest radiograph** revealed a **small bilateral pleural effusion.**



What to think and to do?



88-year-old man with hypertension and benign prostatic hypertrophy.

Six-month history of generalized edema, asthenia, anorexia.

Hypotension. Ascitis.

Haemoconcentration, leukocytosis, hypoalbuminemia, mild renal impairment. Hepatic cytolysis.

Small bilateral pleural effusion.



?

Differential Diagnosis

- ▶ Congestive heart failure
- ▶ Chronic liver disease
- ▶ Nephrotic syndrome
- ▶ Protein-losing enteropathy
- ▶ Angioedema
- ▶ Auto-immune disease
- ▶ Infectious disease
- ▶ Neoplastic disease





Evolution and treatment (1)

Furosemide 40 → 80 mg
iv id Spironolactone 100
mg po id

Human Albumin



1st
day

Progressive clinical

worsening: ► generalized
edema (new onset of scrotal and
abdominal wall edema)

► great volume ascitis

► dyspnea



Without improvement of

laboratorial results





Diagnostic workup (1)

- ▶ **Total proteins** ↓ (min.:6,1g/dL) **Albumin** ↓ (min.:2,1 g/dL)
- ▶ **Urinalysis:** normal values
- ▶ **24h proteinuria:** normal
- ▶ **ESR and CRP:** normal values
- ▶ **Pleural and ascitic fluid analysis:** Transudate
- ▶ **Abdominal Ultrasound:** **Great Volume Ascitis**
- ▶ **Echocardiography:** No significant valvular or structural abnormalities. No pericardial effusion. Normal LVEF
- ▶ **Abdominal and Chest CT:** bilateral pleural effusion and ascitis
- ▶ **Endoscopy study of GI Tract:** Chronic gastritis; Diverticulitis; Colic Polyp (villous adenoma)





Diagnostic workup (2)

- ▶ **Blood Gases:** pO₂ 70 mmHg Sat O₂ 94%. **EPO:** normal
- ▶ **Auto-immune panel and serologies:** normal
- ▶ **Complement studies:** normal

- ▶ **Immunoglobulin profile:** **IgG** ↑ (2,5g/dL)
- ▶ **Serum protein electrophoresis:** **Band in the Gamma region**
- ▶ **Imunoelectrophoresis:** **IgG K M component** (100 mg/dL)
- ▶ **Bone marrow biopsy:** normal phenotype. Plasma cells < 2%





Systemic Capillary Leak Syndrome

- ◆ 1st description by Clarkson in 1960. Also called **Clarkson disease**.
- ◆ **Evolution** in 3 phases:
 - ◇ **1) prodrome** (asthenia, anorexia, hypotension, GI symptoms)
 - ◇ **2) edema** (generalized edema, hemoconcentration, hypoalbuminemia, monoclonal gammopathy)
 - ◇ **3) resolution** (clinical and laboratorial normalisation)
- ◆ **Diagnosis:** clinical signs and laboratory findings. The presence of paraproteinemia is very evocative!





Systemic Capillary Leak Syndrome

- ◆ **Pathophysiology:** not fully understood. Viral infections as a trigger? Importance of paraproteinemia, cytokines (IL-2) and leukotriene B4 as determinants of capillary hyperpermeability.
- ◆ **Different treatments** have been tried: theophylline, aminophilyne terbutaline, salbutamol, steroids, loop diuretics, plasmapheresis, calcium antagonists, gincobiloba extracts, IVIG.
- ◆ **Possible Complications:** shock, rabdomyolysis, compartment syndrome, renal insufficiency
- ◆ Possible progression to multiple myeloma.




Evolution and treatment (1)

Furosemide 40 → 80 mg
iv id Spironolactone 100
mg po id Human
Albumin

1st
day

13th
day

 Systemic capillary leak syndrome



- Progressive clinical worsening:**
- ▶ generalized edema (new onset of scrotal and abdominal wall edema)
 - ▶ great volume ascitis
 - ▶ dyspnea



Prednisolone 25 mg iv
id, Aminophylline 225
mg po bid, Furosemide
60 mg iv id

**Without improvement of
laboratorial results**

Evolution and treatment (2)



Discharge



Prednisolone 20 mg po id
(progressive tapering)

Aminophylline 225 mg po
bid

Furosemide 20 mg po id

At this
moment

24th
day

40th
day

3th
month

1st / 2nd
years

3th
year

Fast clinical improvement

- ▶ Peripheral edema resolution.
 - ▶ Abdominal and pleural effusion reduction
- (~ 9kg in one week: 69,3 > 60,7 Kg)
- ▶ Haemoglobin, serum creatinine, plasmatic protein, AST and ALT values normalisation



▶ peripheral edema worsening and pleural effusion

▶ Same laboratorial pattern

Admission

▶ Peripheral edema worsening

Outpatient management



No recurrences



- ▶ Systemic capillary leak syndrome: **if you know it, you won't miss it!**
 - ◆ Chronic forms may occur and our clinical case provides a demonstration of it.
 - ◆ Hypotension, generalized edema, hemoconcentration, hypoalbuminemia. **Monoclonal gammopathy is a hallmark of the disease** (without evidence of multiple myeloma)
 - ◆ Acute forms can rapidly evolve to hypovolemic shock and death. **Keep it in mind!**
 - ◆ Possible evolution to Multiple Myeloma. **Annual surveillance!**
 - ◆ No defined etiology, no curative treatment! **Supportive therapy**. Prophylactic therapy under study.



Message to take home



Portugal is a
beautiful

