Introduction: Rhinoscleroma is a rare, chronic, granulomatous disease that most frequently affects the upper respiratory tract, especially the nasal cavity, and sometimes extends through the lower respiratory tract. It is associated with *Klebsiella rhinoscleromatis*, which is endemic in certain geographic regions, namely Central America. The pathogenesis and risk factors remain unclear.

### CLINICAL CASE

#### 5 Years old, Portuguese boy
- Irrelevant family history
- Previously healthy
- Summer vacations 2009, Dominican Republic

**Emergency Department – July 2010**

- Epistaxis
- No other signs or symptoms
- Rhinoscopy: **INTRANASAL BLEEDING MASS**
  
  **MRI – July 2010**
  
  Intranasal mass with extension to ethmoid bone sinus

**Bacteriologic exam of biopsy material** - *Klebsiella Spp.* sensitive *in vitro* to amoxicillin and clavulanic acid

**HISTOPATHOLOGY PATOGNOMONIC FEATURE**

- Granulomas (1) containing Mikulicz cells (2) - cells with *Klebsiella rhinoscleromatis*

**RHINOSCLEROMA**

**GENETIC STUDY**

- Exome sequencing: functional study in immortalized cell lines
- Under course

**IMMUNODEFICIENCY SCREENING:**

- Immunoglobulins; Lymphocyte subpopulations; Phagocytosis test; Oxidative burst; CH100 Normal

**Treatment**

- Amoxicillin plus clavulanic acid

**Follow-up**

- Monthly evaluation - O.R.L and Pediatrician
  
  - Progressive reduction until total disappearing of macro and microscopic lesion
  - Negative bacteriologic exam

**6 Months of Antibiotic therapy**

**8 Months After End of Treatment**

**ASSYMPTOMATIC**

- No evidence of recurrence

**What is said in the literature!!**

- Clinical outcome is variable.
- 3 Stages: catarrhal or atrophic; granulomatous or hypertrophic and sclerotic.
- Antimicrobial therapy must be administered for prolonged periods (3-5 M).
- Needed antibiotics with activity against gram-negative bacilli, intracellular efficacy and low toxicity. Ex: quinolones
- Association with cellular immunodeficiency; weaker antibody responses.
- Genetic predisposition to *Klebsiella rhinoscleromatis* infection may involve a specific pathway.
- Relapse until 3 years.

**Comments:** Globalization and free transit of people to areas far from origin countries where some rare diseases are endemic brings a new challenge to modern medicine. Sometimes vacations bring more than memories.

**Bibliography**