



# Summer vacations not only a memory for life, but also a rare infection.

Inês Simão<sup>1</sup>, Iuri Gaspar<sup>2</sup>, Rosário Faustino<sup>2</sup>, Maria João Brito<sup>1</sup>  
<sup>1</sup> Infectious Disease Unit – Department of Pediatrics  
Head of Department: Gonçalo Cordeiro Ferreira

Hospital de Dona Estefânia, Centro Hospitalar Lisboa Central – EPE, Lisbon Portugal  
<sup>2</sup> Otorhinolaryngology Department – Hospital de Santarém – EPE, Portugal

**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 of Central America. The pathogenesis and risk factors are unclear.

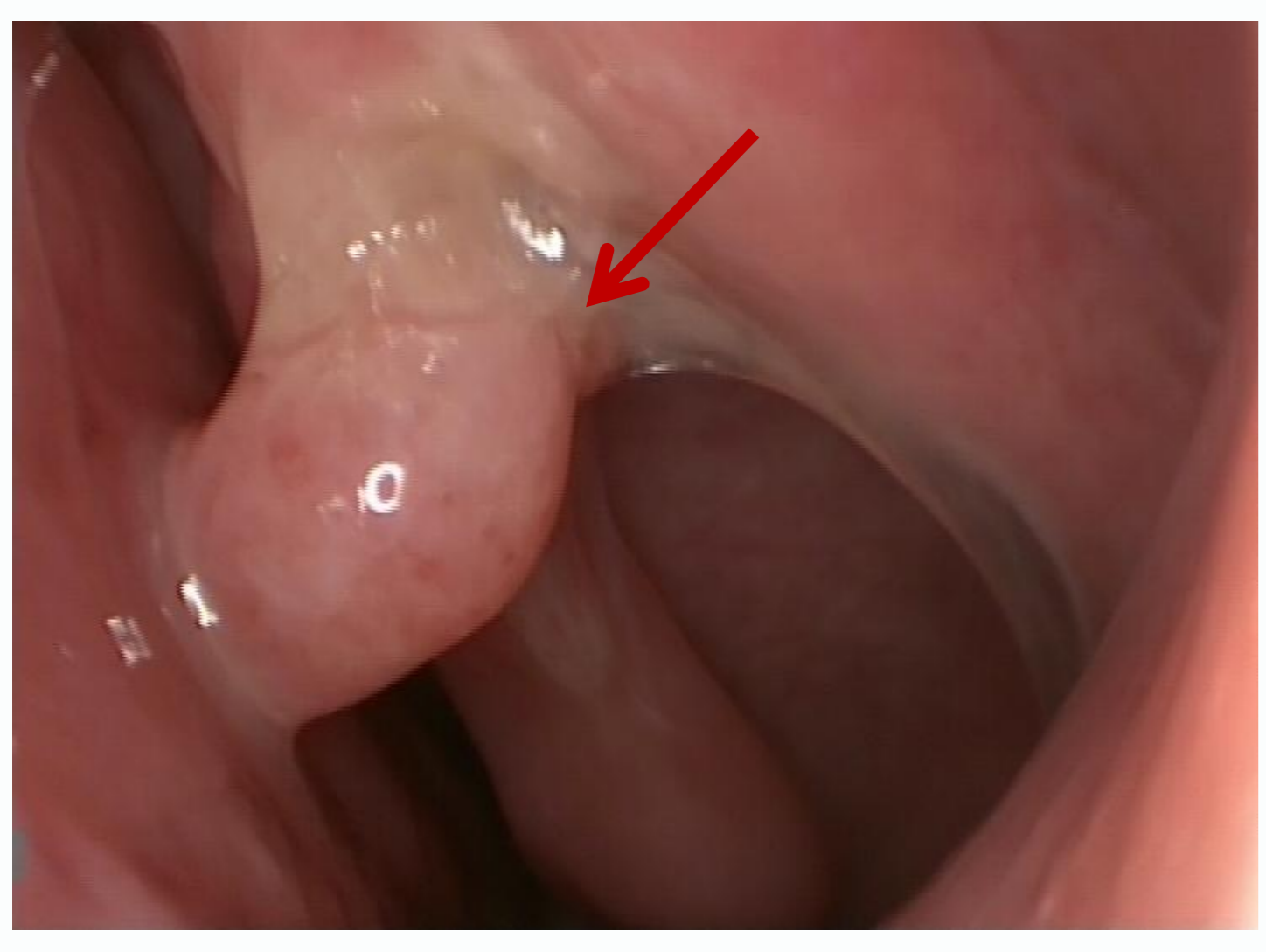
## CLINICAL CASE

### 5 Years old, Portuguese boy

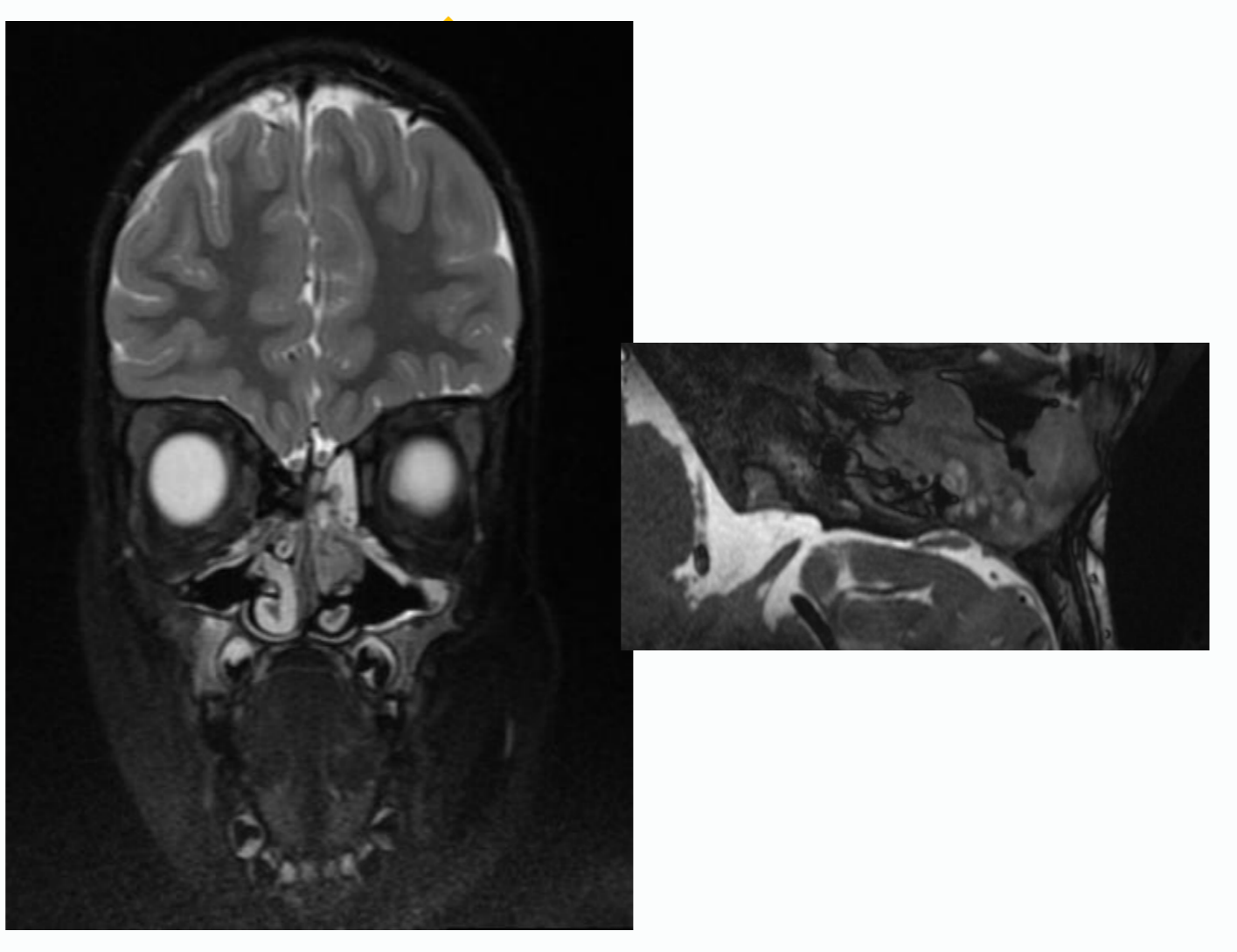
Irrelevant family history  
Previously healthy  
Summer vacations 2009 Dominican Republic

### 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

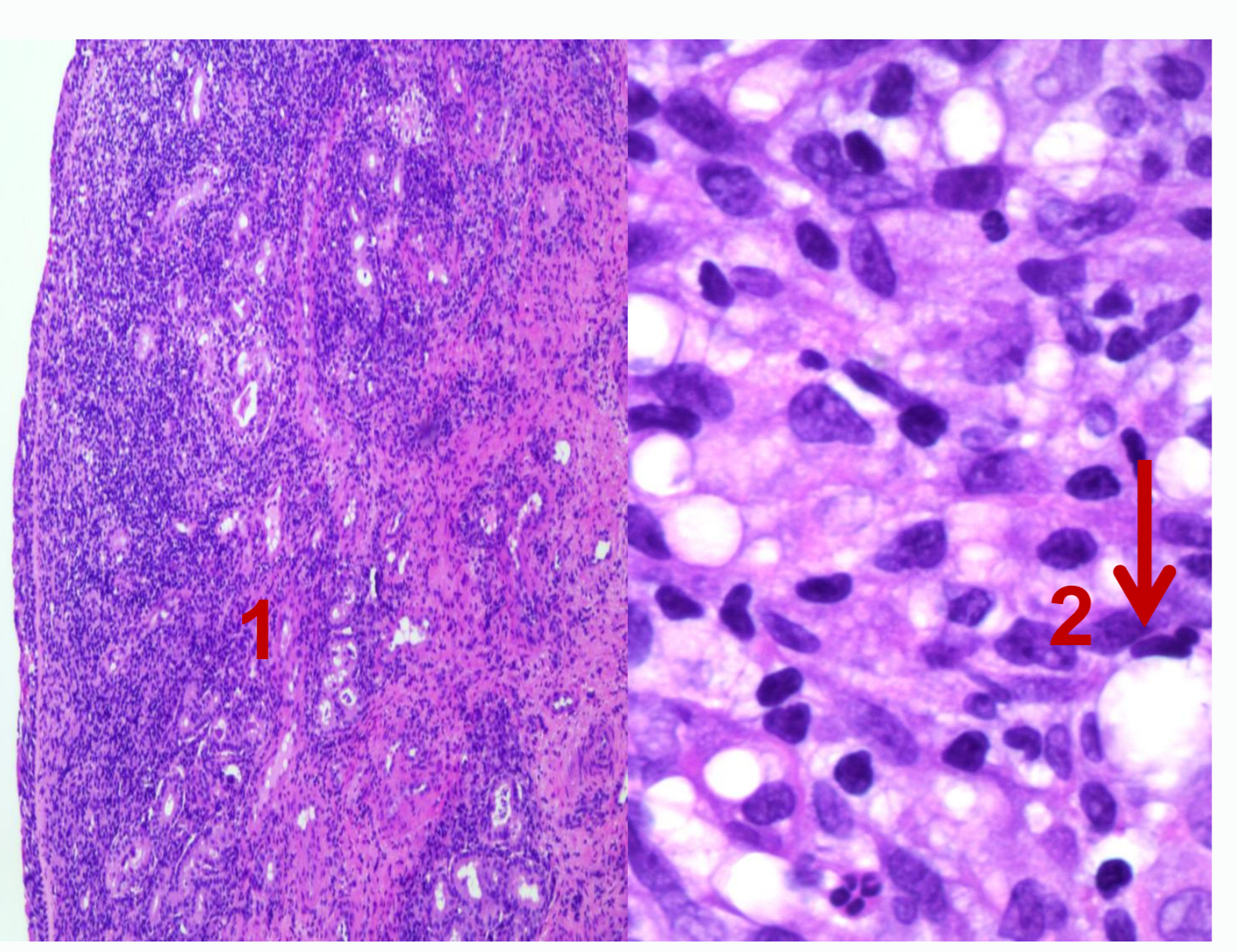


Picture 1 - Rhinoscopy



Picture 2 - MRI

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



Picture 3 - Histopathology

### 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 - ORL and Pediatrician



Picture 3 - Rhinoscopy

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 (3-9 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

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