Case Presentation

E F Post

Introduction

- Case Presentation
- Ideas from audience
- Discuss pathology
- Treatment options

History

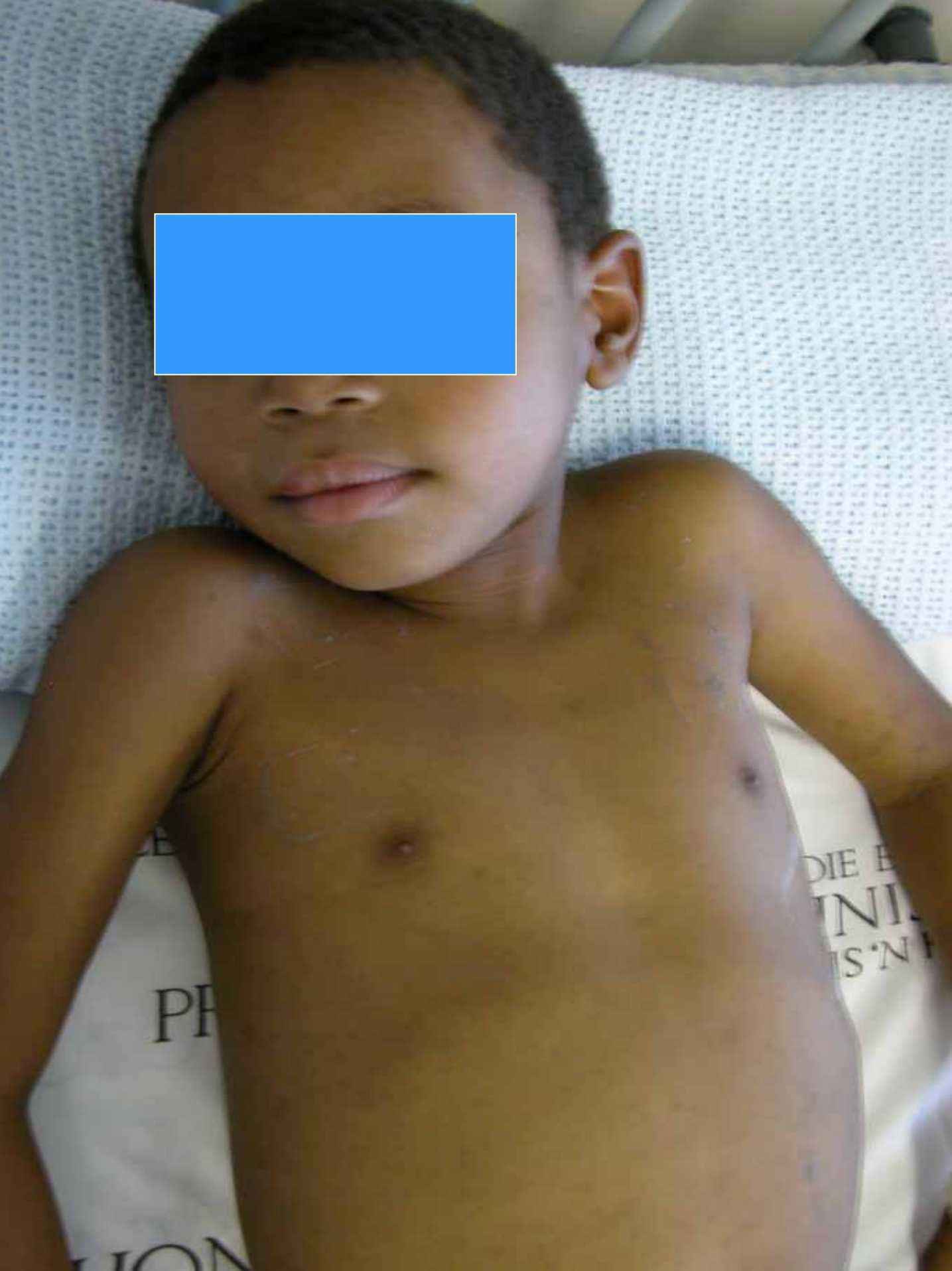
- 5 yo male

- SOB }
- Hoarseness } 3/12
- Cough }

- PMHX: Asthma DX 5/12
- TB contact: Father 3 years ago

Presentation

- Insp. + Exp stridor
- RR 54
- Sats 88% (room air)
- Resp. distress
- Chest: decreased A/E
- Pulsus Parodoxus
- RESPIRATORY COLLAPSE --- ETT



What now?

Special Investigations

- Bloods: U+E (N)
  FBC wcc 14
  LFT (N)
  RVD (+)

- Bronchoscopy: polyp/ granuloma VC

- ENT Flexi Scope: Granuloma VC /
  - Ball/ Valve effect

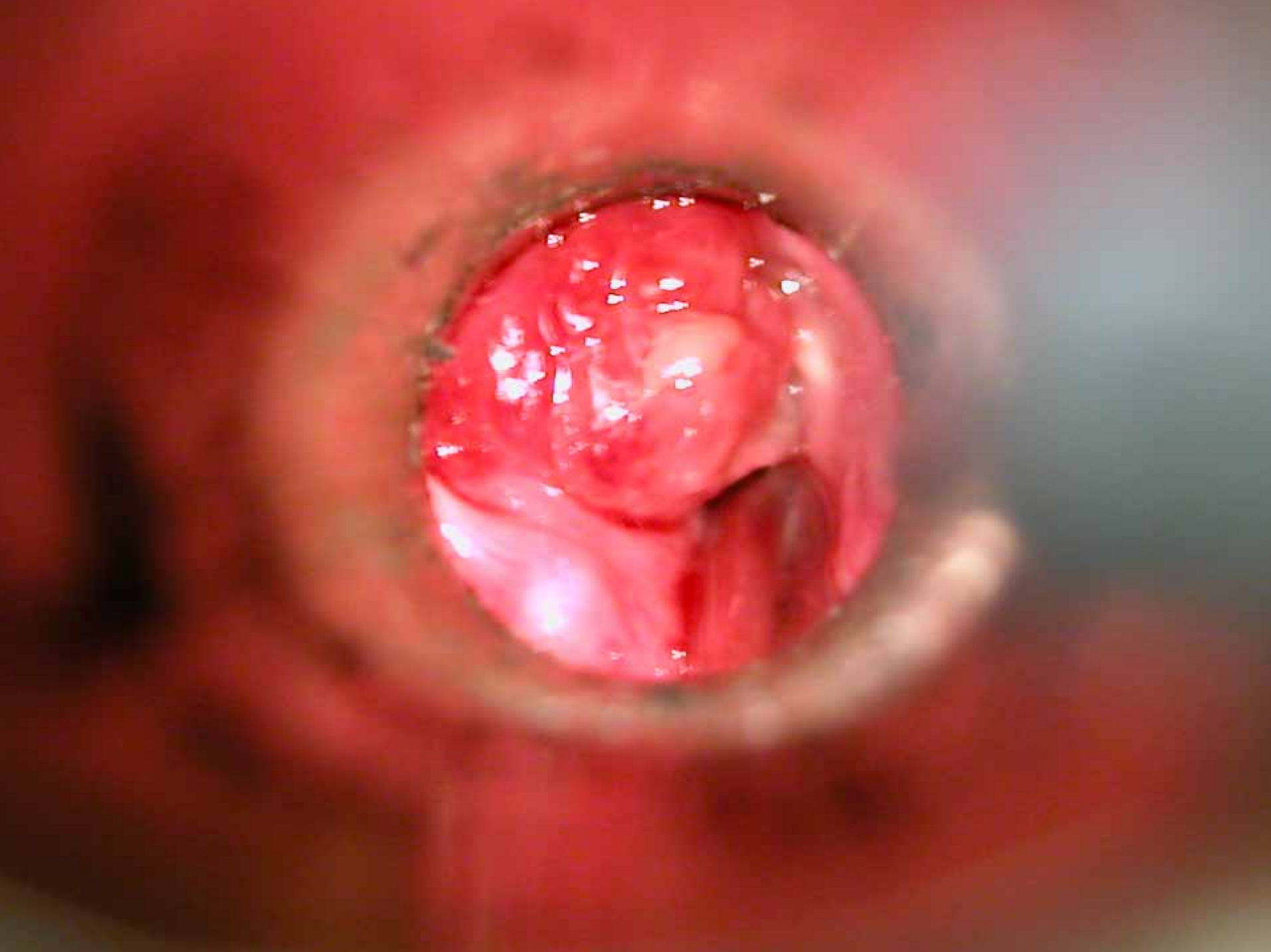
- TB investigate Sputum (-)
  G.aspirate (+)

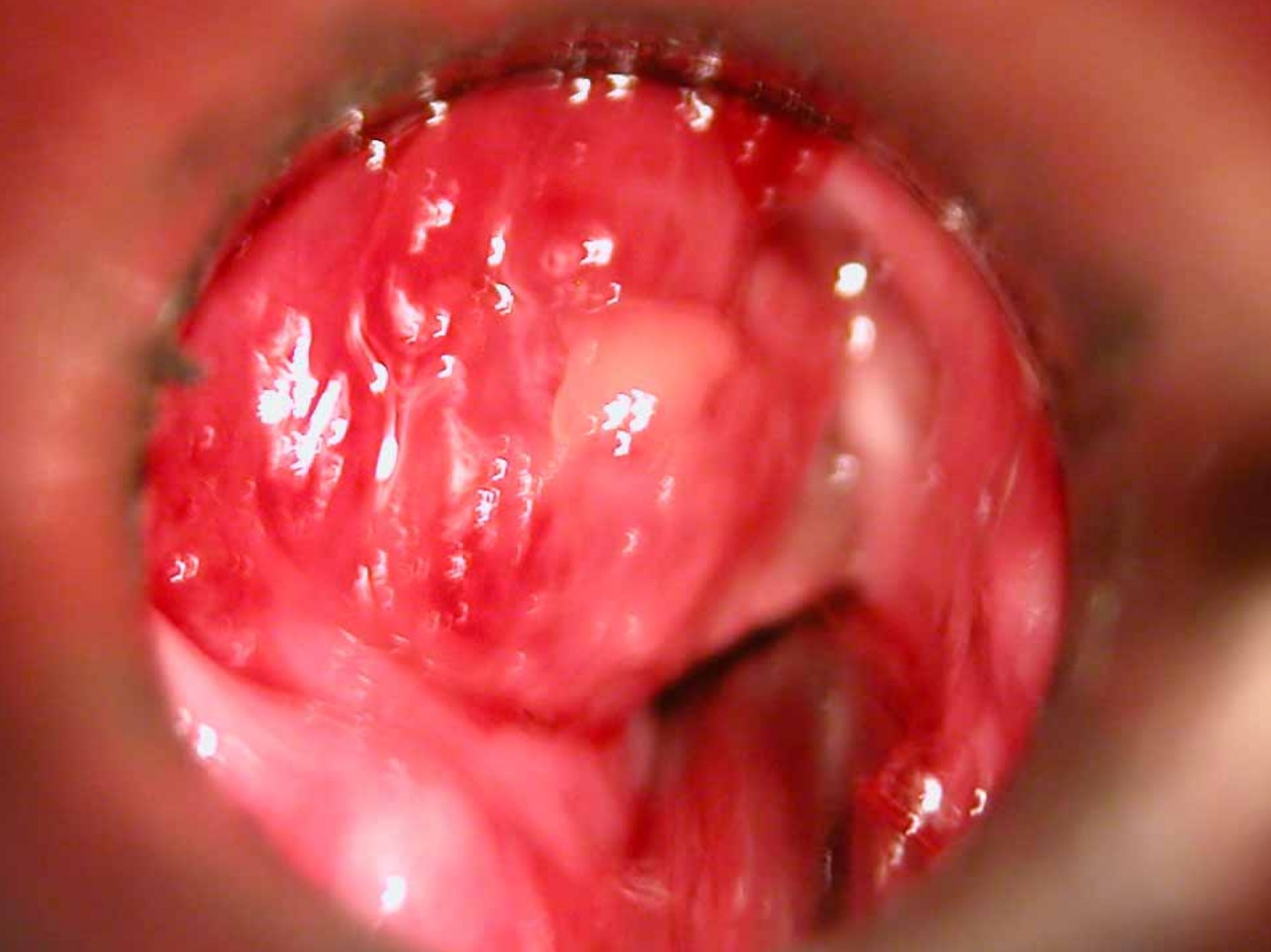
Initial Treatment

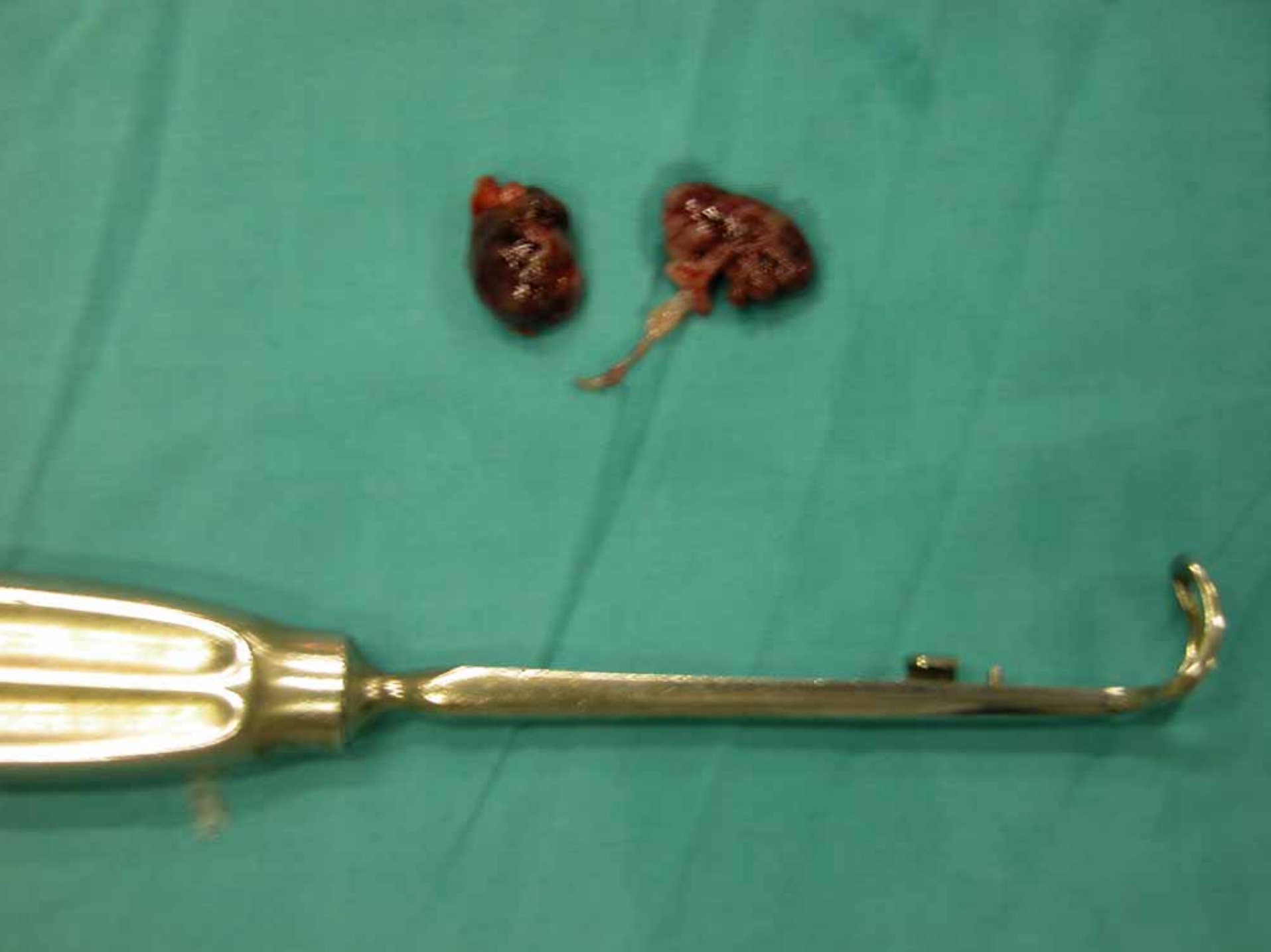
- AntiTB Rx
- Antibiotics: Zinnat
- Steroids: Prednisone
- Biopsy/ EUA:
    - Pyogenic Granuloma - Continue treatment

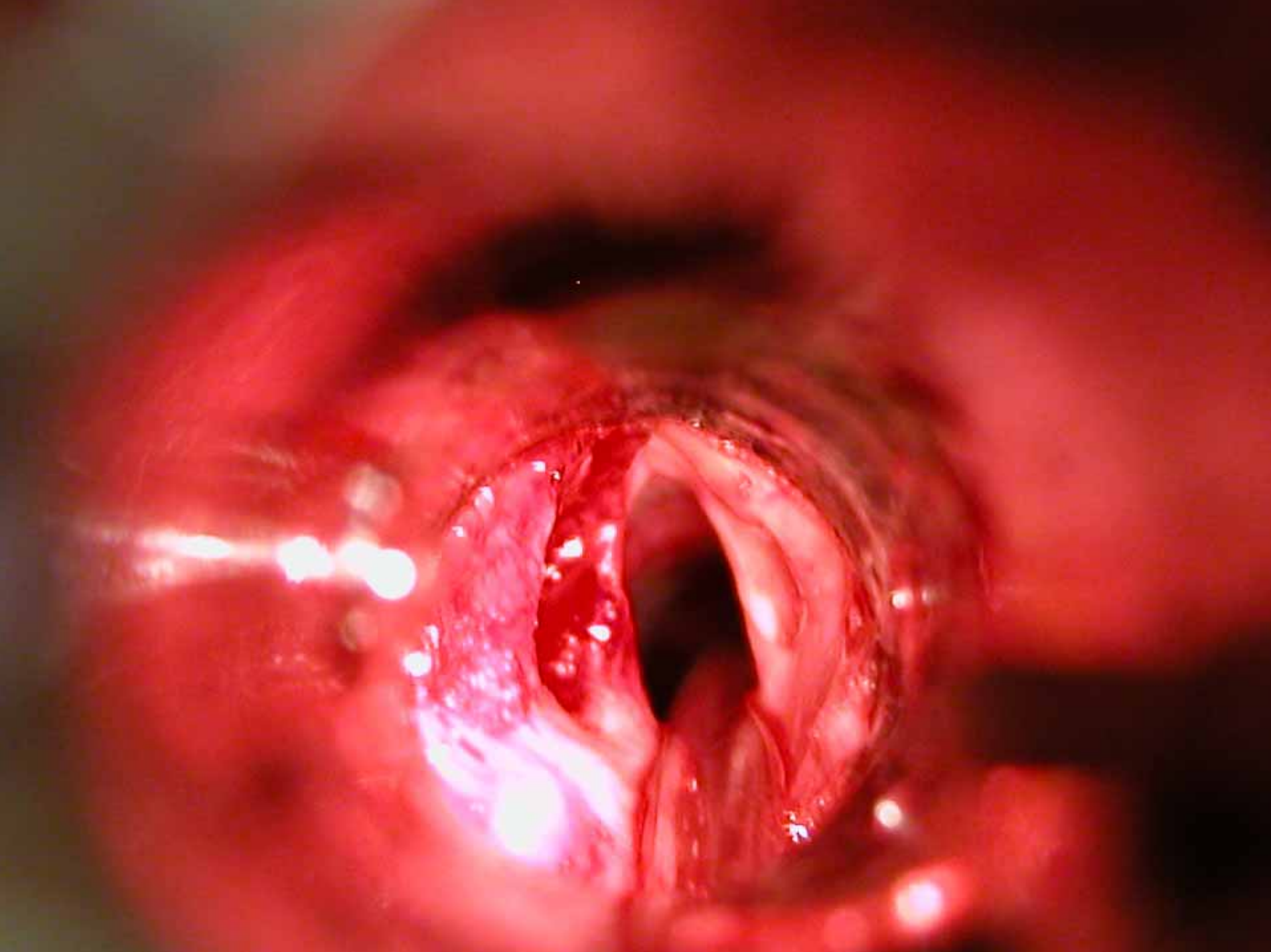
Further Management

- Extubate day 4
  - Stridor +
  - Nocte sleep apnoea
- Rescope ENT: Nasal mass (dark)
  Supraglottic mass
- Plan surgery
- CD4+ 232 (9%) /CD8+ 1301 (52%)
  CD4:CD8 0.17:1



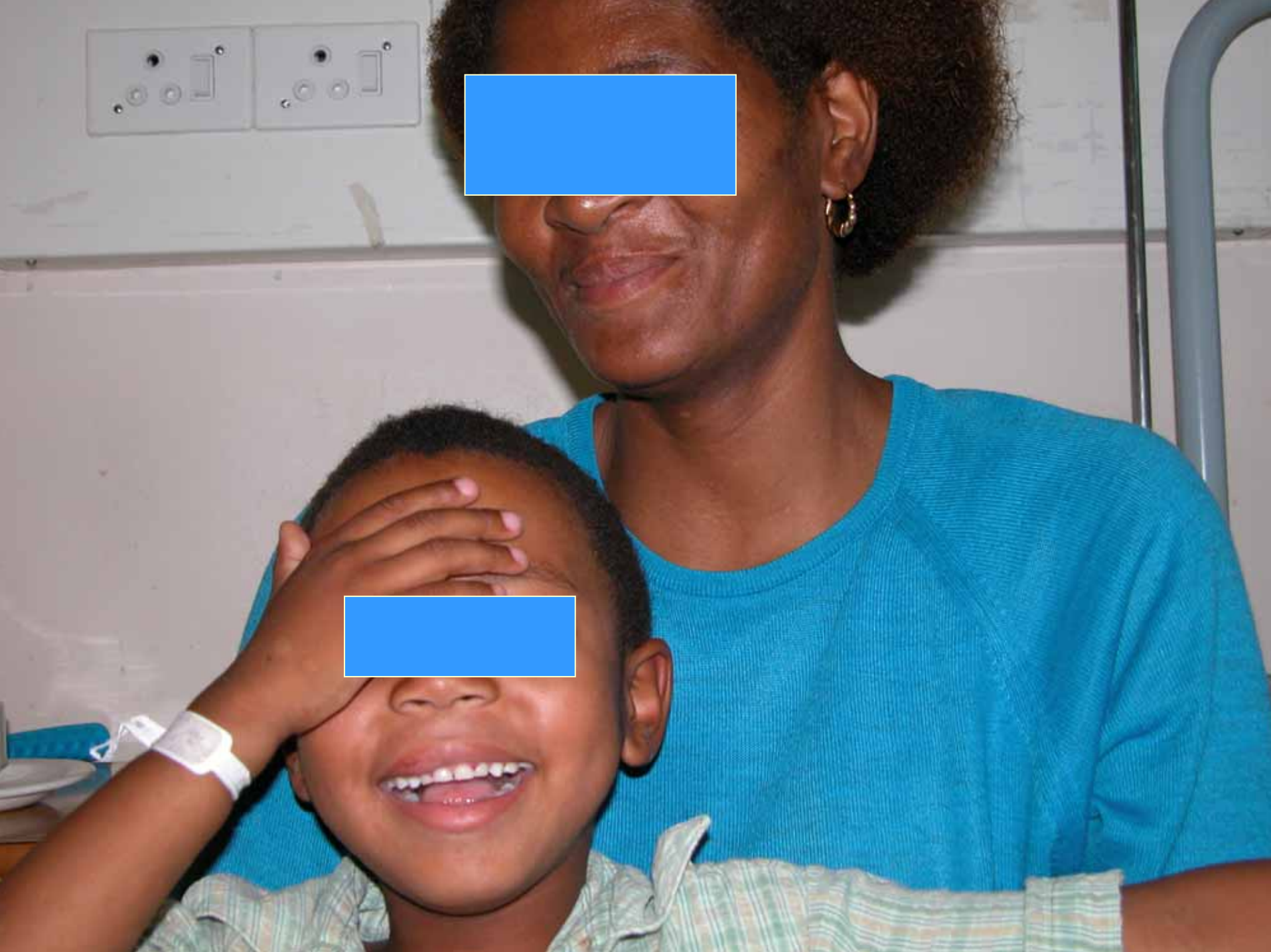






Surgery / Management

- Adenoidectomy : bleed
  : 2x 2x3cm purple
- Excise supraglottic mass : bleed
  : 1x 1.5 cm
  : ventricle
  - Adrenaline topical
  - Mitomycin C 5 min
- Histology / Diagnosis ???
  - histo/ immunohistochemistry



Histology / Microbiology

- AFB Nil
- MCS Haemophilus
- HHV 8
- **Kaposi's Sarcoma**
- Rx: Zinnat/ Predinosone

Management

- HAART
- Chemotherapy / Oncology
  - Vinkristine 1mg
  - 4 x Vinkristine 1 weekly
- Prednisone (taper)
- F/U ENT rescope 2/52 to E. Cape

Kaposi's Sarcoma

- Clustered cases 1982 L.A. / AIDS
- Past: tumour elderly men, Mediterranean
- KSHV= Human herpesvirus-8
  - co-factor

Pathology

- Mesenchymal cells that form blood vessels + proliferate
- Surround by spindle cells
- Extravasated RBC = pigment (hemosiderin)

- HIV: infect CD4 / T cells } interleukins
  } cytokines
- HHV-8: infect B-cells } growth factor

Clinical

- Pigmented macular-papular lesion
- Solitary nodule / Clusters
- Surround / distal oedema
  - Lymph infiltration
  - Local extravasation (VEGF)

Clinical manifestations

- Mucocutaneous

- GIT: endoscopic
  seldom hemorrhage +++

- URT: sinus/ pharynx/ larynx
  compromise / dysphagia

- Disfigure: ears / nose
  ulcer / 2° infection
  nil bone

Clinical manifestation

- Pulmonary:
  - Pleural- effusion
  - Bronchial- cough/ hemoptysis: bronchoscopy/ Bx
  - Parenchyma- resp. failure
    - CXR: Reticulonodular
    - CT: Peribr. Cuffing
  - Thallium scan (+) vs gallium (-)

Differential

Hematoma
Bacillary angiomatosis
Pyogenic granuloma
Pityriasis rosea
Secondary syphilis
Hemangioendothelioma
Lichen planus
Purpura
Angiosarcoma
Basal cell Ca
Melanoma
Sarcoid plagues
Nevi
Prurigo nodularis

Special investigations

- HIV/ CD4:
  - 1 of the 3 AIDS defining tumour
    - KS
    - Lymphoma
    - Cervix Ca
- HHV-8: 95% of KS
- Punch biopsy / excision biopsy
  (CDC clinician Dx, but recommend)

Kaposi's Sarcoma

- **CLASSIFICATION:**
  - Classic / European
    - 1872
    - Elderly men, Jews / Mediteranean
    - "tumor die with, not from"
  - Epidemic / AIDS associated
    - 30% develop lymphoma
  - Endemic / Africa
    - young black / aggressive
  - Organ transplant
    - (Imm. Suppression)

Staging of AIDS-Related KS
GOOD RISK

- Immune — CD4 > 200
- Illness — NO opp. Infx
- Tumour (T) — small
  ltd. : LN
  skin
  oral / palate

Staging of AIDS-Related KS
POOR RISK

- Immune — Opp. Infx
  B symptoms
  HIV related: NHL, wasting S
- Illness — CD4 < 200
- Tumour — Large
  Oral
  GIT / pulmonary
  Oedema / ulcer

Treatment

<u>Local / Single lesions</u>:

- Cryotherapy
- Surgical removal
- Vinblastine local: repeated injections
  recur
- Radiotherapy: resistance to chemo
  slow growth
  poor outcome

Treatment

- **<u>Systemic / Multiple lesions</u>**: CHEMOTHERAPY
  - Indications: oedema / lymph infiltration
    visceral
    lung
    cosmetics
  - Up to 80% response: BUT repeat/ incurable
  - Triple agent:
    - » Vinblastine
    - » 9-cis retinoic acid
    - » Paclitaxel
    - » IFNα (CD4 > 100)
    - » Kayelex }
    - » Taxol } USA only

Treatment / Complete

- Investigate GIT / Lungs
- HIV: opportunistic organisms
- HHV8: HAART (gancyclovir decr. %)
- Chemo vs Radio etc.