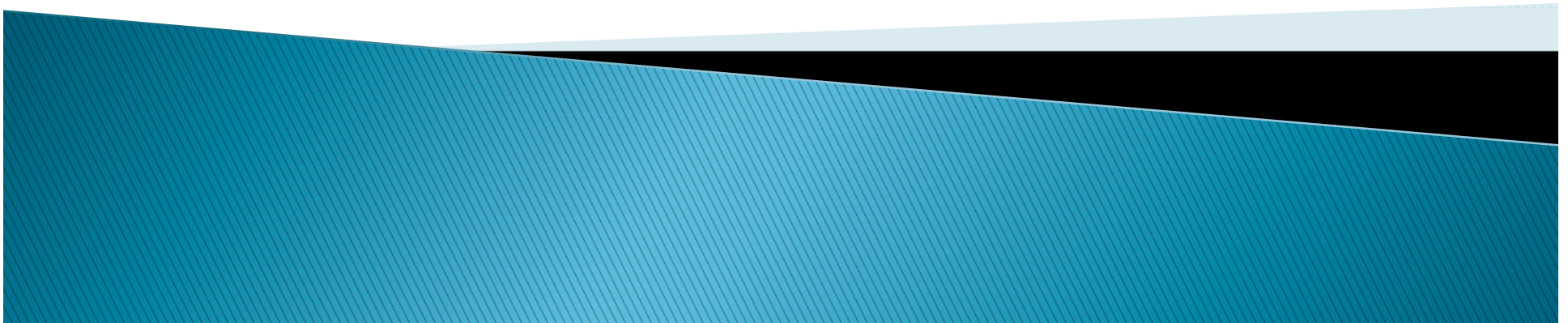


# 41 years old female patient with Raynaud's syndrome and lymphadenopathy

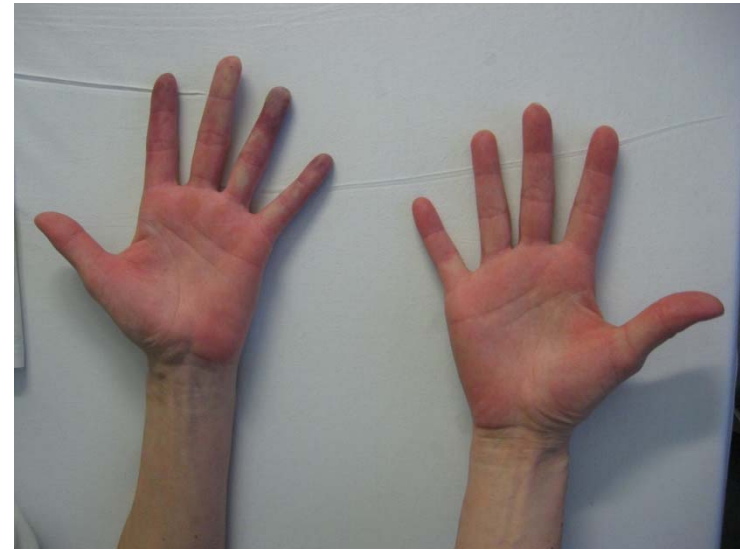
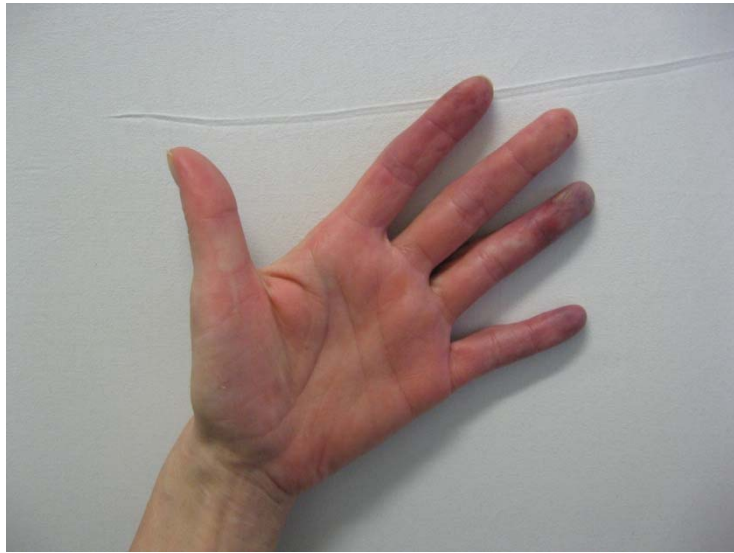
Johannes Hilpert (MD)

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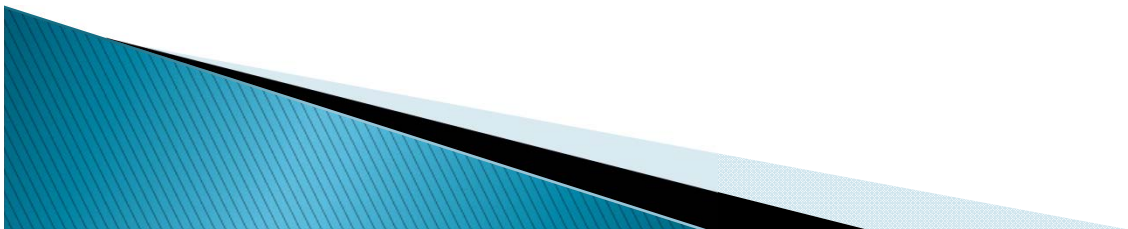


# Acra of the patient



# Initial Presentation

- ▶ 41 year old female patient, BMI 18kg/m<sup>2</sup>
- ▶ Interfering cramps and ataxia
- ▶ Disseminated paresthesia
- ▶ Raynaud's syndrome left Hand
- ▶ Lymphadenopathy
- ▶ Severe depression and anorexia
- ▶ Cerebral stroke in 2008
- ▶ Prehospital diagnostics:
  - Cerebral MRI: postischemic area a. media
  - Abdominal CT: no pathologies
  - Excluded MELAS syndrome



# Initial laboratory diagnostics

- ▶ Screening for vasculitis (prehospital): negative
- ▶ Leucocytosis ( $19.300 \times 10^9/L$ )
- ▶ Severe eosinophilia (60%)
- ▶ ESR 61 / 83 mm, CRP 13,3mg/dl
- ▶ Polyclonal gammopathy (IgG: 3040mg/L)
- ▶ Urine status: negative



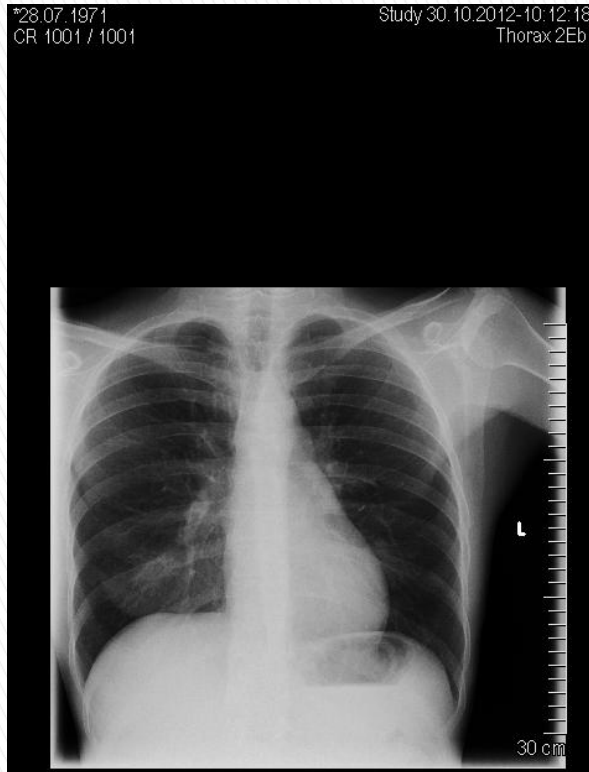
# Hematological findings

- ▶ Bone marrow biopsy:
  - Thrombo-, Erythropoiesis intact
  - Massive eosinophilia, „Hiatus leucämicus“
  - No proliferation of myeloblasts
- ▶ FACS analysis:
  - No signs for leucemia  
(CD 117: 0,5 %; CD 34: 0,2%)
- ▶ Cytogenetics:
  - No signs of acute leucemia or B-NHL

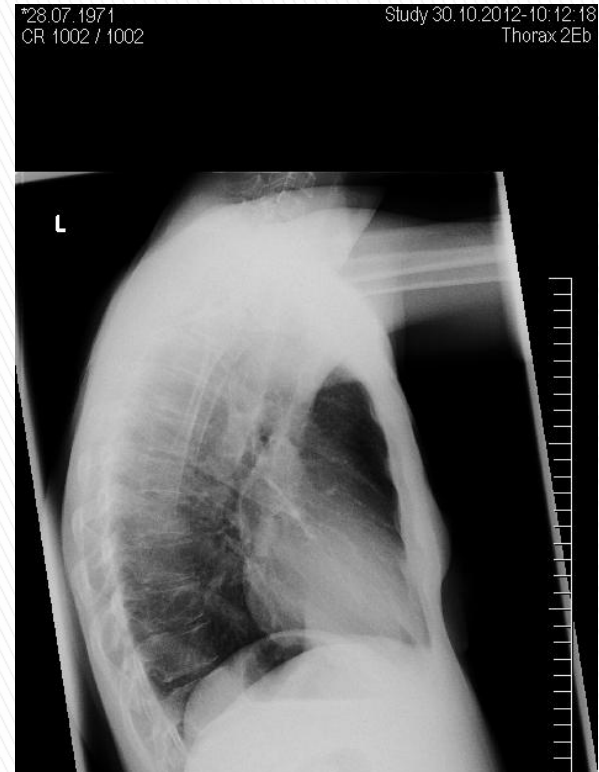




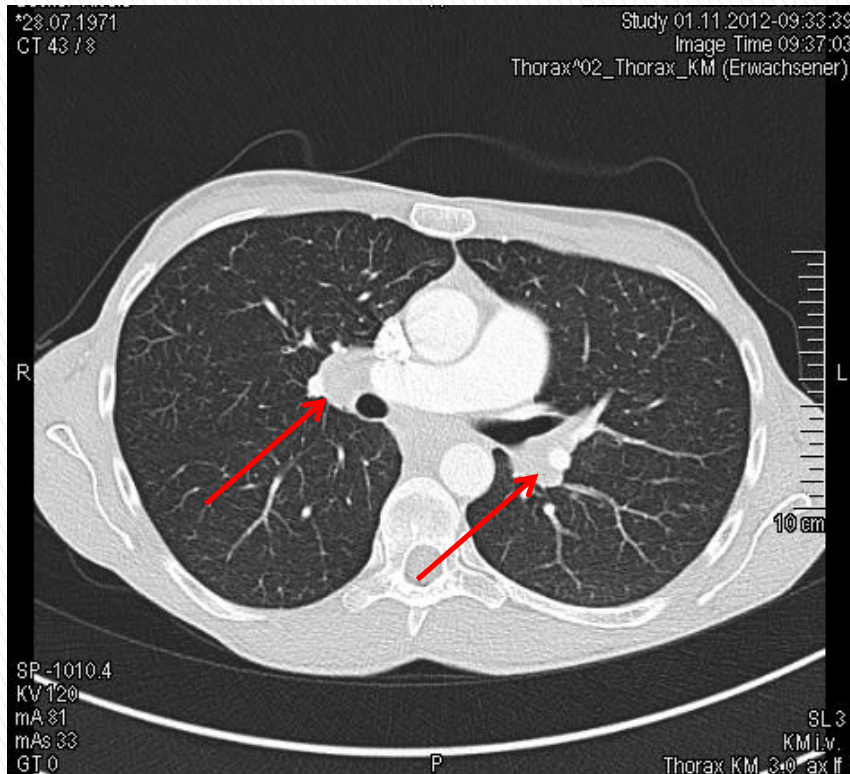
# X-ray of the chest



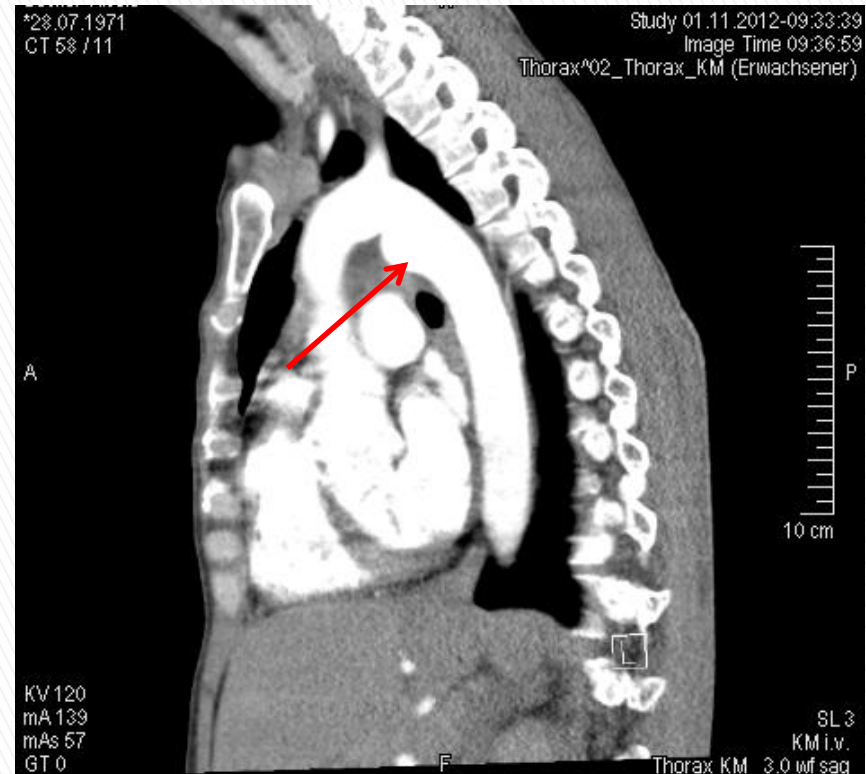
emphysema



# CT of the chest

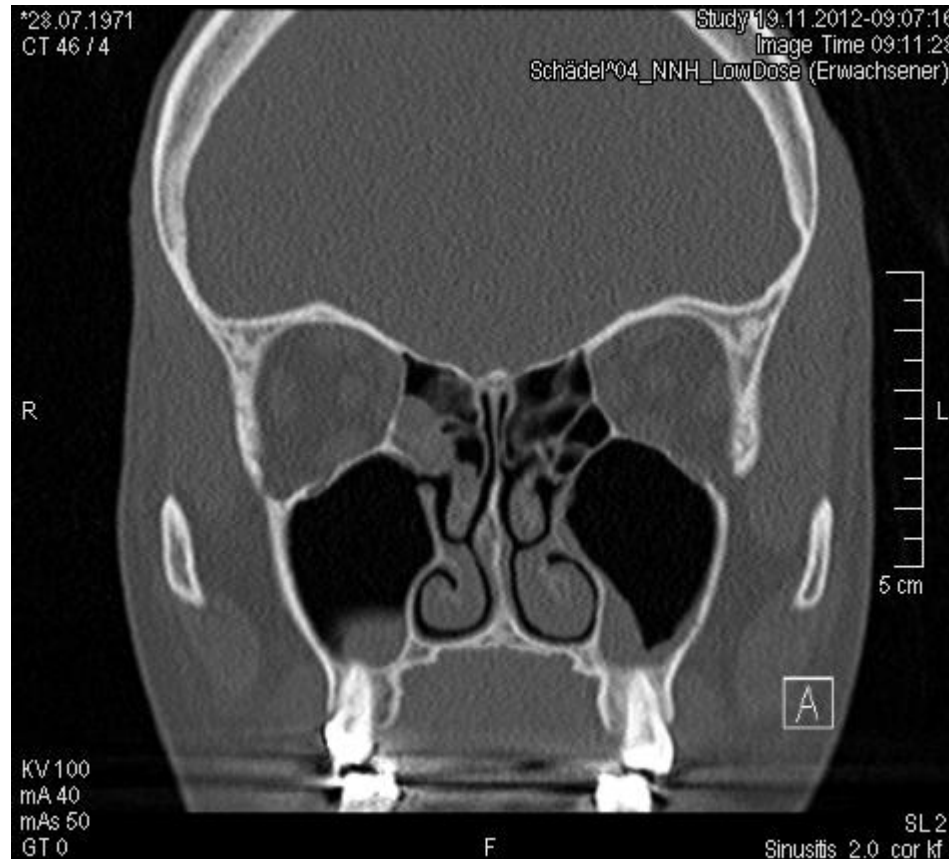


Bihilary lymphadenopathy  
up to 19mm



Aorta ascendens  
aneurysm 38mm

# Sinusitis maxillaris and ethmoidalis





# Diagnostics (2)

- ▶ Echocardiography:
  - Hypertrophic cardiomyopathy (septal hypertrophy)
  - Prolaps of the mitral valve
- ▶ Bodyplethysmography
  - FEV1 2.22l  $\triangleq$  74% compared to reference group
  - With bronchospasmolysis: FEV1 3,02l  $\triangleq$  100%
- ▶ Abdominal ultrasound
  - 3 hemangiomas of the liver



# Vasculitis vs. Leucemia or HES

- ▶ Raynaud's syndrome
- ▶ Disseminated lymphadenopathia
- ▶ Obstructive ventilation disorder
- ▶ Recurrent infections of respiratory tract
- ▶ Chronic sinusitis
- ▶ Aneurysm of the aorta
- ▶ Cardiomyopathy
- ▶ Stroke in history

- ▶ Raynaud's syndrome
- ▶ Disseminated lymphadenopathia
- ▶ Bone marrow with „hiatus leucemicus“ and massive eosinophilia
- ▶ 60% Eosinophils in peripheral blood

Vasculitis ??

HES or Leucemia ??

# Clinical course in hospital

- ▶ Progredient pain in upper extremities
- ▶ few affect of high dosis opiates, NSAR and amlodipine
- ▶ Increasing leucocytosis and eosinophilia
- ▶ Progression of raynaud's syndrome, starting in right hand, too
- ▶ Loss of appetite
- ▶ reduction in general condition
- ▶ Progressive symptoms of depression



# Further diagnostics

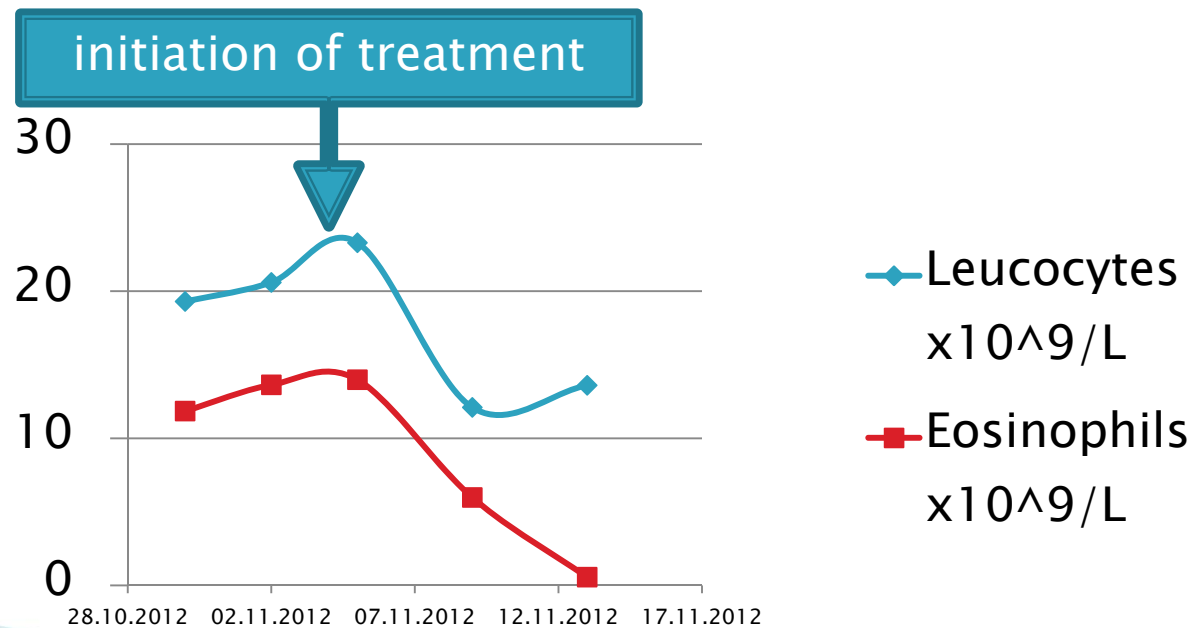
- ▶ Second vasculitis screening
  - Non typical cANCA pattern 1 / 40 (ref. Titer:  $- < 10$ )
  - ELISA negative in all common tests
- ▶ IgE titer 3862 kU/l (reference  $< 100$  kU/l)





# Therapy

- ▶ High dose glucocorticoid-therapy
  - Prednisone 1 mg/kg BW
- ▶ Continued application of NSAR, opiates and amlodipin
- ▶ Calcium & Vit.D3 for bone protection



# Clinical course (2)

- ▶ Rapid fall of total and eosinophilic leucocytes in peripheral blood within days
- ▶ Regredient raynaud's syndrome but mummification of D.IV left hand
- ▶ Intermittent plexus-anesthesia for pain relief
- ▶ Lower doses of opiates
- ▶ Stabilisation and demission to rehabilitation centre 6 weeks after start of glucocorticoid treatment (at demission: 15mg prednisone/d)



# D IV at demission date



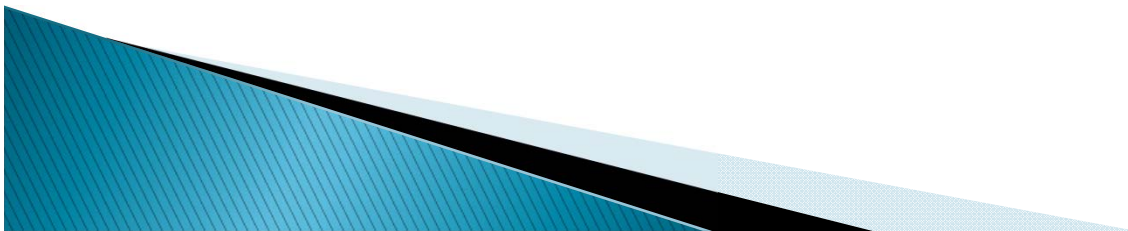
Beginning necrosis



Mumification

# Diagnosis

- ▶ Vasculitis Churg Strauss





# Points of discussion

- ▶ Differential diagnosis ?
- ▶ Start of glucocorticoid treatment ?
- ▶ Application of Calcium and Cholecalciferol ?
- ▶ Other immunosuppressive agents such as MTX or cyclophosphamide ?
- ▶ Biopsy ?
- ▶ Underlying organic disease as cause of severe depression ?

