University of Milan Humanitas Clinical Institute

A frequent symptom for a rare disease

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MEDICAL HISTORY

M. G.

- Caucasian
- Male
- 70 years old
- Retired government employee
- Smokes 20 cigarettes/day since he was 20
- Family history of stroke (father) and lung cancer (brother)
- Six years history of type II diabetes mellitus treated with Metformin

PRESENT HISTORY

In the last 2 months he developed:

- continous fever (his temperature ranged from 37.5° C to 38°C)
- loss of appetite
- 10 Kg weight loss
- malaise and fatigue
- headache mostly in the forehead

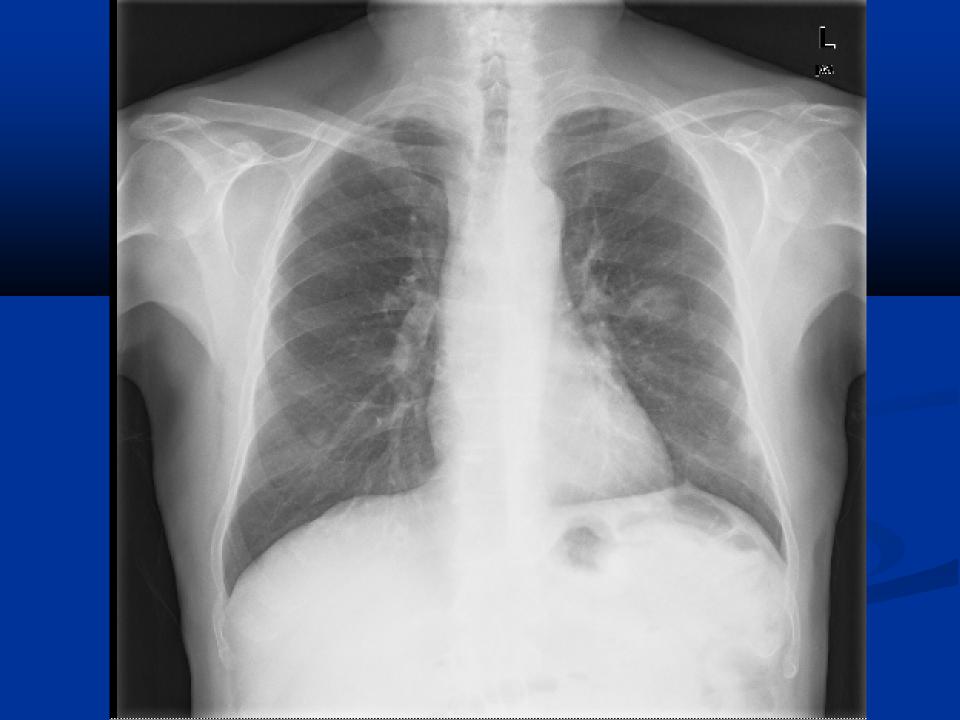
He was treated with amoxicilline/clavulanic acid without any improvement, and he was admitted to our hospital

PHYSICAL EXAMINATION

- Cachectic appearance
- Temperature 37.5° C
- Normal vital signs
- A few rales were heard at the left lung base
- Unremarkable cardiac and abdominal examinations
- Two small, soft, not tender lymph nodes were palpable in the anterior cervical region
- Tenderness on palpation over frontal sinus
- Normal neurological examination

BLOOD TEST

- Hb 8.5 g/dl
- MCV 89
- MCHC 34.8
- WBC 11500/mmc
 NEU 9100/mmc
 LYM 1300/mmc
 MON 800/mmc
 EOS 200/mmc
 BAS 0/mmc
- PTL 288000/mmc
- CRP 26.2 mg/dl (0-1 mg/dl)
- Ferritin 1110 ng/ml
- Transferrine 1.6 g/l
- Iron 10 mcg/dl
- Normal liver and kidney function



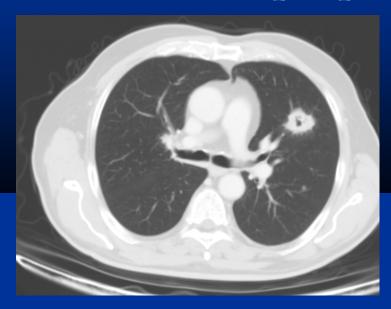
HYPOTHESIS

?

- Cancer
- Metastasis
- Abscess
- Pneumonia/fungal pneumonia
- TB

CHEST CT SCAN AND LUNG ULTRASOUND:

multiple pulmonary nodules









LUNG CANCER?

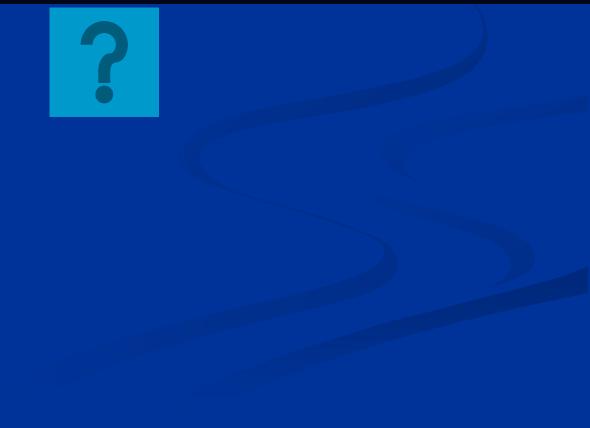
Fibrobronchoscopy and transbronchial biopsy:

tracheobronchial mucosal infiltration

But...

There are not hilar or mediastinal masses at the CT scan

HYPOTHESIS



ENT advice:

nasal ulcers

Facial CT scan:

sinus opacification and sinus polyps thickened turbinates and sinus mucosa



Transbronchial biopsy

Histological findings:

"inflammatory infiltrate composed of neutrophils, lymphocytes, histiocytes, and eosinophils"

Blood test

cANCA positive IgA 380 mg/dl (<350)

Proteinuria

2.5 g/24 hours

CT guided lung biopsy

Histological features:

"parenchymal necrosis, vasculitis, and granulomatous inflammation characterized by an inflammatory infiltrate composed of neutrophils, lymphocytes, histiocytes, and eosinophils"

DIAGNOSIS

Wegener granulomatosis

with upper respiratory tract, tracheal, pulmonary, and renal involvement

Treatment:

PDN 1 mg/Kg/die Cyclophosphamide 2 mg/Kg/die