

Case Presentation

ESIM 2010, London

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First Admission

- 26-year-old male, primary school teacher, single, nonsmoker
- He was admitted to ER with complaints of
 - ✓ Cough (No sputum)
 - ✓ Fever
- On physical exam:
 - BT:38,1°C, HR:98/min; RR:22/min; BP:118/76
 - Rare rales on auscultation on right lower lung zones
 - Systems otherwise normal

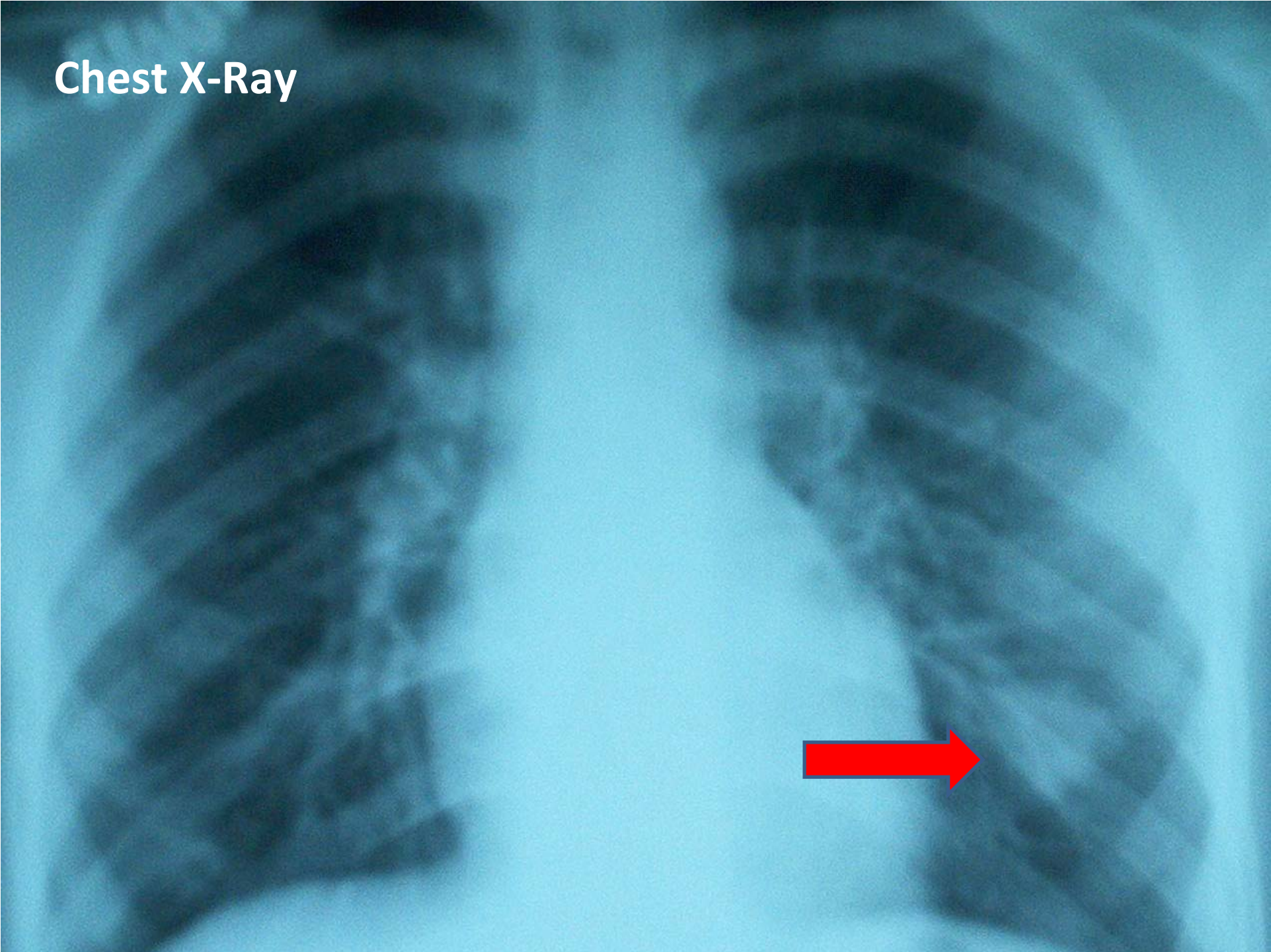
- Lab results

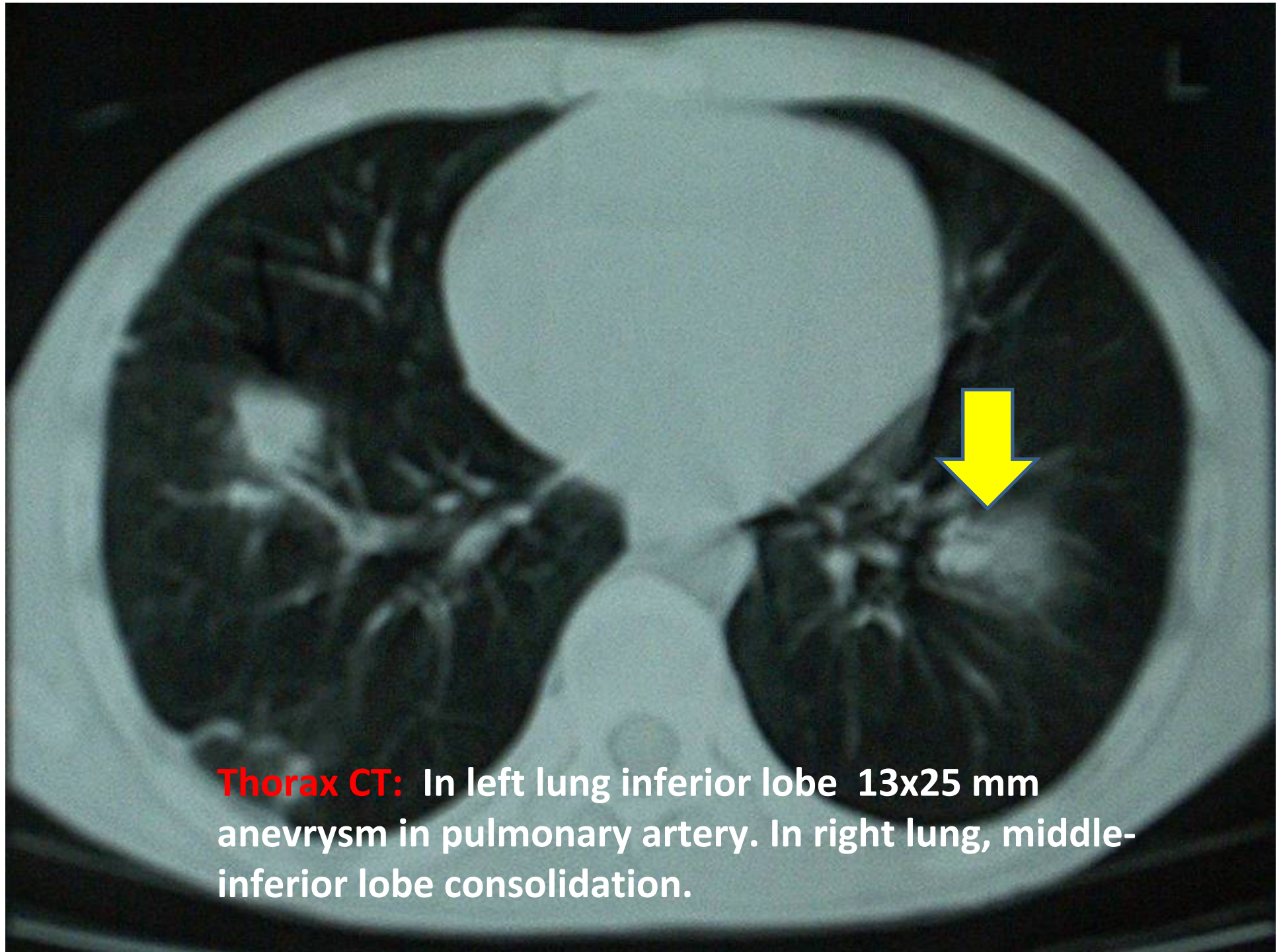
Hb:13,5 g/dl WBC: 9400/mm Trm:194.000/mm

Liver enzymes – Renal function tests normal.

ESR:24mm/h

Chest X-Ray





Thorax CT: In left lung inferior lobe 13x25 mm anevrysm in pulmonary artery. In right lung, middle-inferior lobe consolidation.

- Past medical history

3 years ago he had uveitis and received oral steroids

Two more questions to ask;

Any recurrent oral ulcers?

Any recurrent genital ulcers?

Further questioning revealed that in the last five years he had many episodes of painful oral aphthous lesions. The lesions heal without any treatment. He also had painful genital ulcers that heal with leaving scars.

Behçet's Disease



1937

Triad of

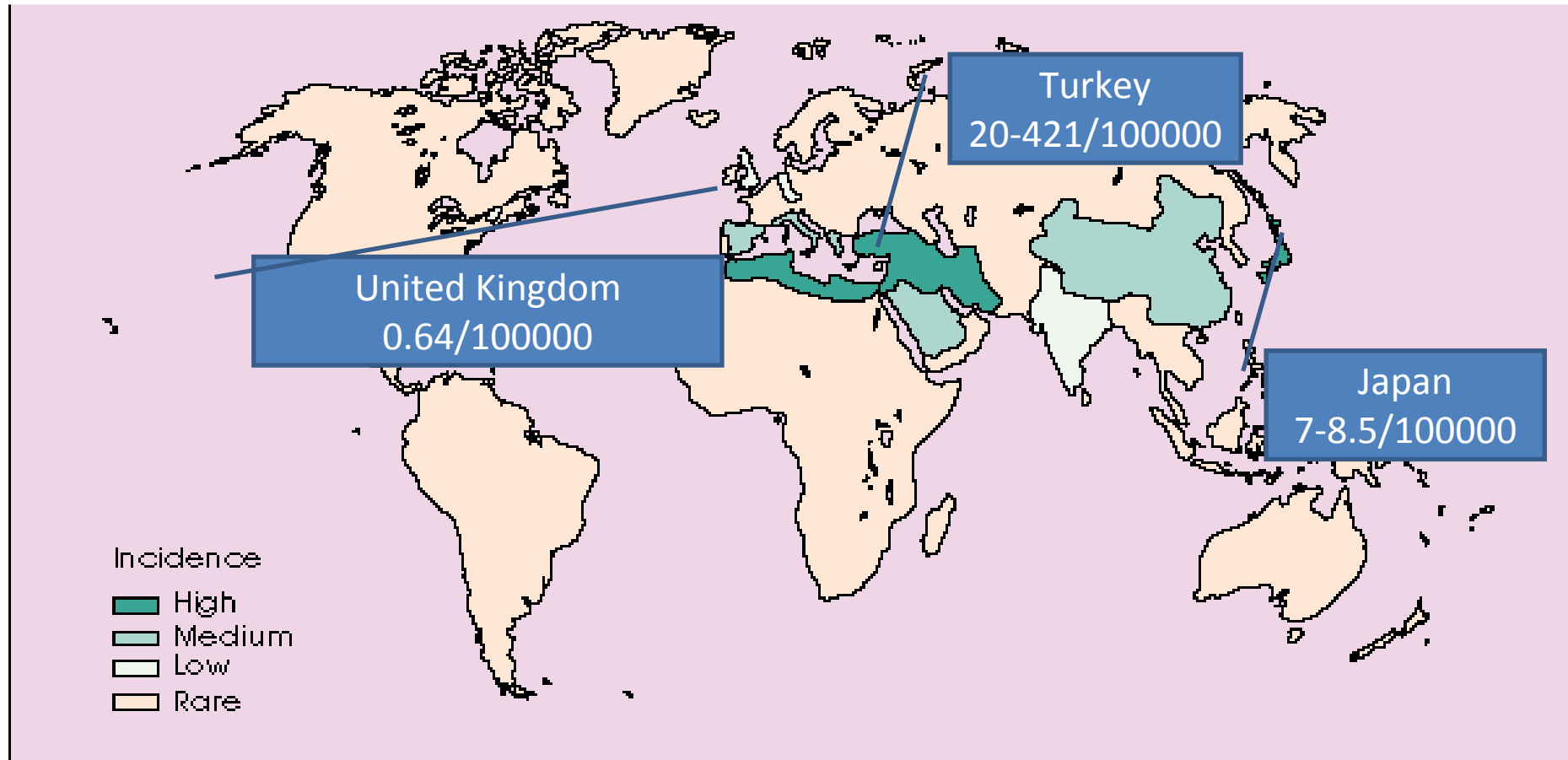
recurrent oral aphthous

lesions

genital ulcers

uveitis

Behçet's Disease



M=F (more severe symptoms in males)

Starts in 2nd-3rd decades

International Study Group Criteria (ISGC-1990)

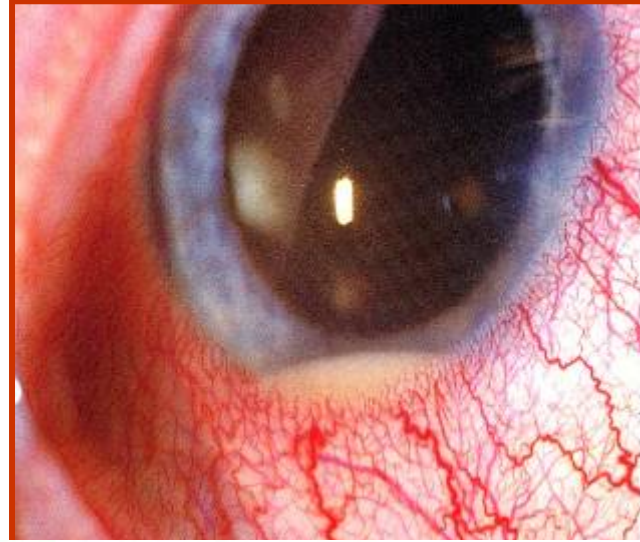
Recurrent oral aphthous lesions;
at least three times a year,
no otherwise reason

+

At least two of these criteria

- Genital ulcer
- Eye involvement
- Cutaneous signs
- Positive pathergy





Hacettepe Univer.
2787291
01.01.1971
AAA
R: 3/1
I: 8/23
FLTR: 30%
MAX OPAC
PLANE: A



- Treatment
 - Antibiotics for possible pneumonia
 - High dose iv steroids and cyclophosphomide
 - Oral steroid maintenance
 - Colchicine 3x1tb
 - ASA
 - Benzatine penicillin G
 - Interferon alpha

In the third month of follow-up , control CT scan was performed, the aneurysm was stable.

He did not adhere to routine controls after, discontinued treatment.

Second Admission

- After 12 months...

- Suffers from dyspnea, hemoptysis

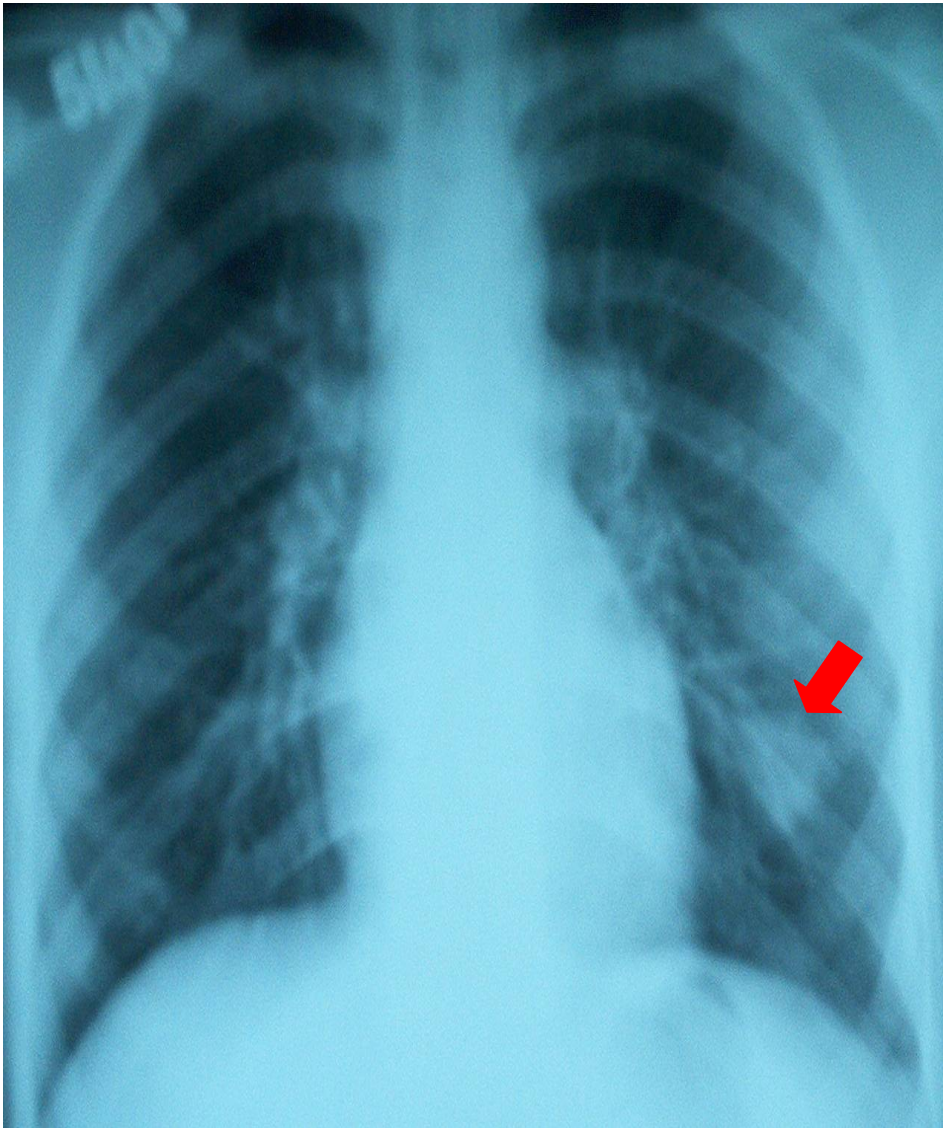
- Lab results:

Hb:11,4g/dl WBC: 9900/mm³ Trm:295.000/mm³

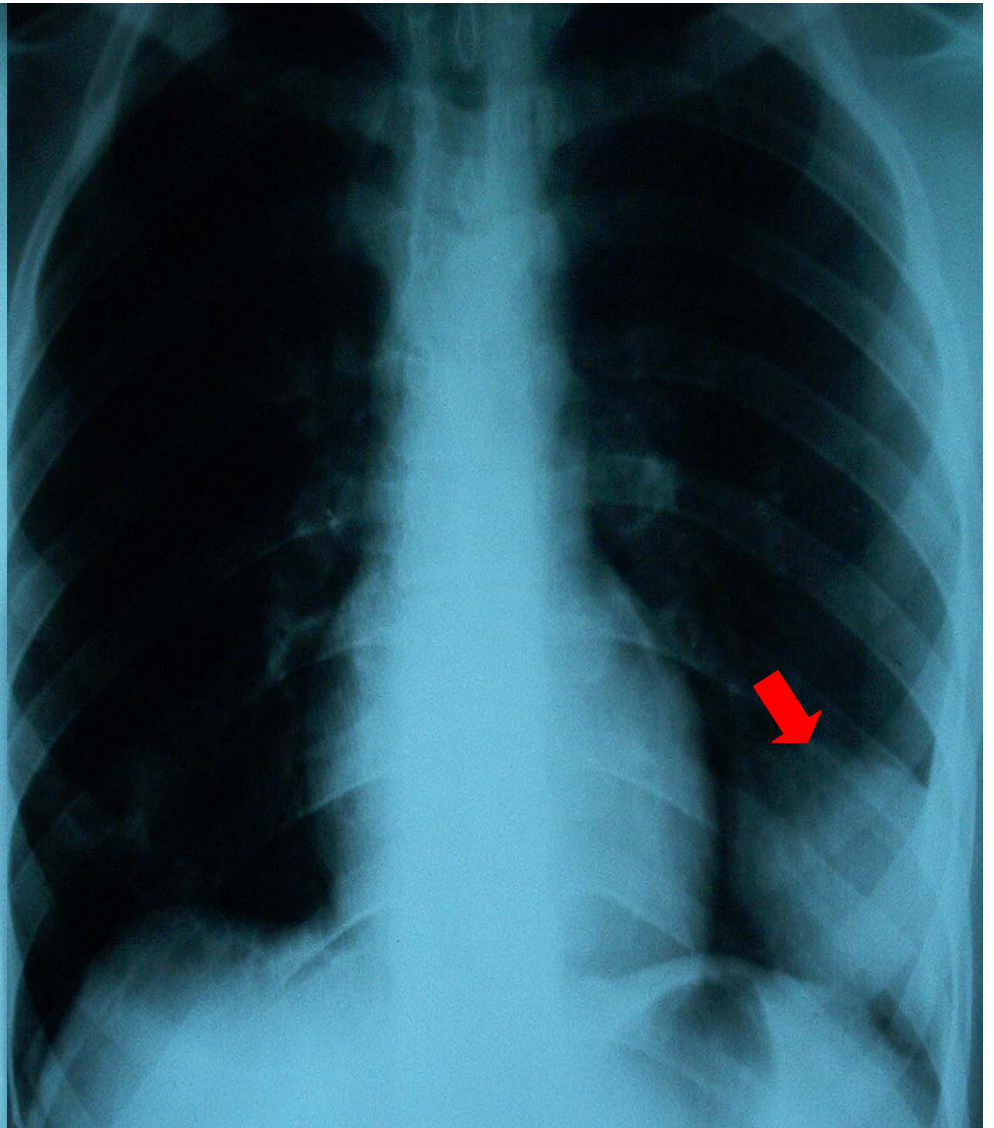
ABG: pH:7.39 pO₂:68mmHg SO₂:95%

Normal Coagulation Parameters

Liver enzymes and renal functions were normal.

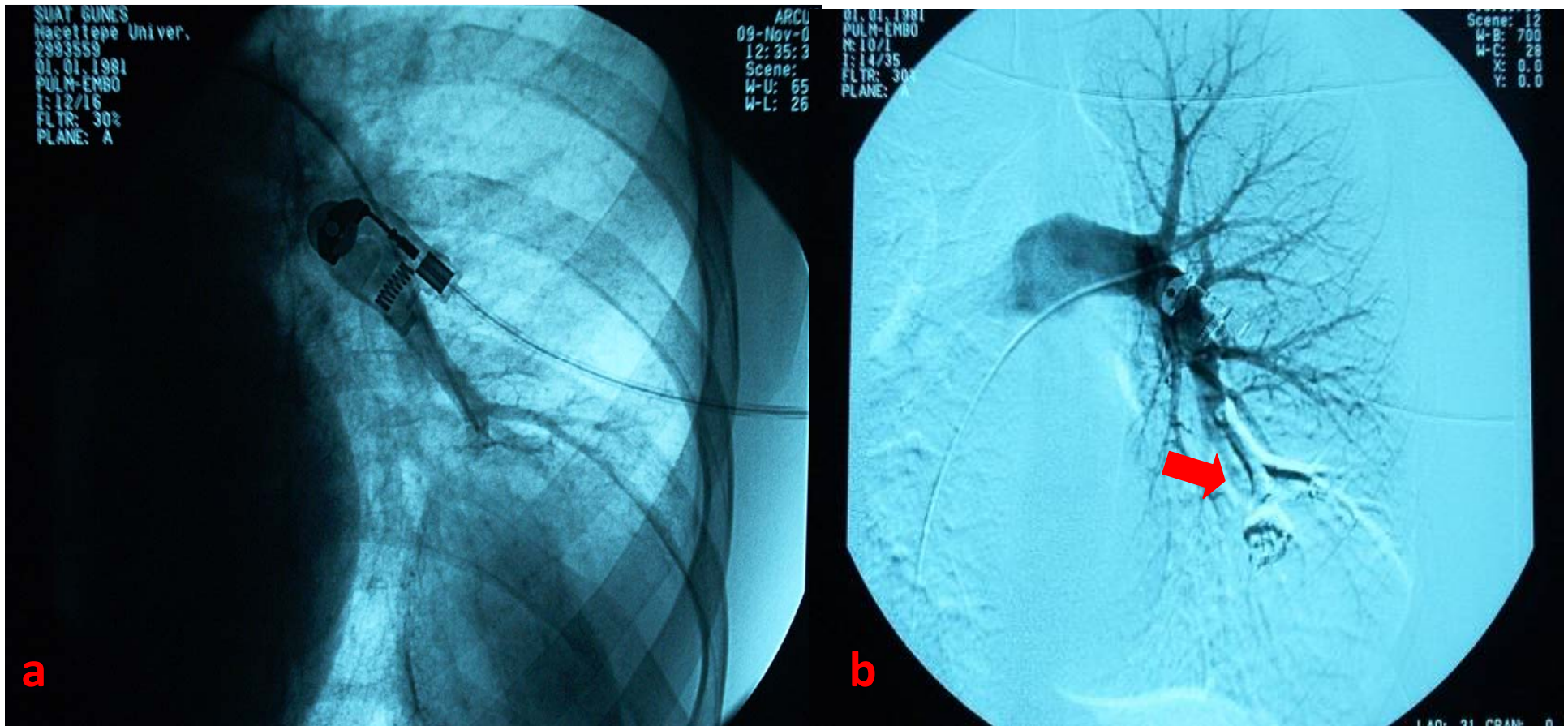


a) Previous Chest X-ray



b) X-ray after bleeding episode

Behcet's Disease



- a) Before Coil embolization
- b) After

- Treatment

High dose steroids and cyclophosphamide

Oral steroid maintenance

Interferon dose was increased

Penicillin G Benzathine

Colchicine and ASA

Classification Of Primary Vasculitis (Chapel Hill Consensus Conference 1992)

Large Vessel Vasculitis

Giant cell (temporal) arteritis

Takayasu arteritis

Medium Size Vessel Vasculitis

Poliarteritis Nodosa

Kawasaki Disease

Small Size Vessel Vasculitis

Wegener Granulomatosis

Churg-Strauss Syndrome

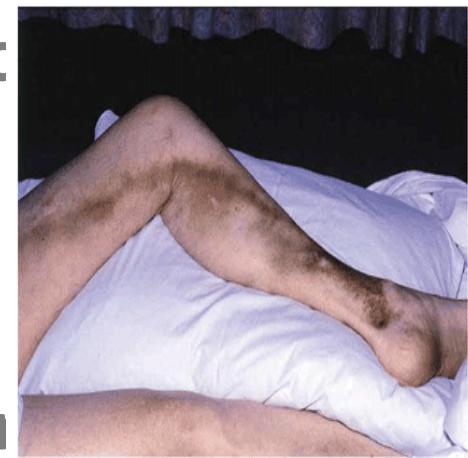
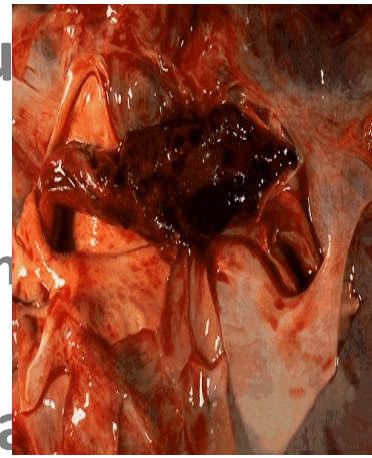
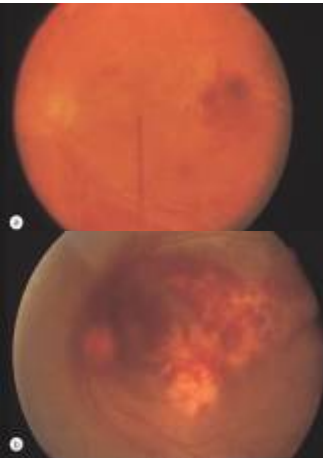
Microscopic Polyangiitis

Henoch-Schönlein Purpura

Essential cryoglobulinemic vasculitis

Cutaneous leucocytoclastic vasculitis

Classification **Behçet Hastalığı** Vasculitis



Involvement of any sized vessels

VEIN > ARTERY

KAWASAKI DISEASE

Small Size Vessel Vasculitis



VASCULITIS IN EVERY LESION

Differential Diagnosis of Large Vessel Vasculitis

- Giant Cell arteritis
- Takayasu arteritis
- Behcet's Disease
- Kawasaki Disease
- Sarcoidosis
- Romatoid arthritis
- Spondiloarhtropaty
- Relapsing polikondiritis
- SLE
- Buerger Disease
- Atherosclerosis
- Marfan syndrome
- Infections
 - Tbc, Syh,
 - Staf, E.Coli, Salmonella
- Norofibromatosis
- Ehler-Danlos syndrome
- SAM
- Fibromuscular displasia
- Trauma/radiation fibrosis

Behçet - Pulmonary Artery Aneurysm

- **1-2 % of patients with Behçet's Disease**
 - Most common region is right inferior lobe.
 - Tends to be multiple.
 - Fusiform, saccular...Sometimes thrombus inside.
- **Rare but lethal! Patients with pulmonary aneurysm: 2 year mortality rate %30**

Park HJ et al Chest 1989

Hamuryudan V et al Br J Rheumatol 1994

Erkan F Opin Pul Med 1999

Seyahi EK et al Medicine 2003

Large Vessel Vasculitis

Table 5 The seven recommendations for the management of large vessel vasculitis with the level of evidence for each statement and the median strength of recommendation as per EULAR operating procedures

Statement	Level of evidence	Median final vote
We recommend a thorough clinical and imaging assessment of the arterial tree when a diagnosis of Takayasu arteritis is suspected	3	C
A temporal artery biopsy should be performed whenever a diagnosis of giant cell arteritis is suspected, but this should not delay the treatment; a contralateral biopsy is not routinely indicated	3	C
We recommend early initiation of high-dose glucocorticoid therapy for induction of remission in large vessel vasculitis	3	C
We recommend that an immunosuppressive agent should be considered for use in large vessel vasculitis as adjunctive therapy	1A for GCA 3 for TAK	B for GCA C for TAK
Monitoring of therapy for large vessel vasculitis should be clinical and supported by measurement of inflammatory markers	3	C
We recommend the use of low-dose aspirin in all patients with giant cell arteritis	3	C
Reconstructive surgery for Takayasu arteritis should be performed in the quiescent phase of disease and should be undertaken at expert centres	3	C

EULAR, European League Against Rheumatism; GCA, giant cell arteritis; TAK, Takayasu arteritis.

Hemoptysis / Radiologic suspicion



Helical Computerized Thoracal Tomography



Pulmonary Artery Anevrysm



Medical Treatment

High dose Steroid+ Cyclophosfamide
+ INF α



No response/Massive hemoptysis



Embolization
NBCA/Coil embolization



Surgery
Lobectomy/
Anevrysm pilication

Take home messages...

- Although Behçet's Disease is rare in western countries, be careful especially for people from eastern countries.
- Consider Behçet's Disease in young patients with;
 - Uveitis, thrombosis in unusual places (Budd-Chiari Syndrome, Portal vein thrombosis, cerebral sinus thrombosis, SVC-IVC syndromes, ...), thrombophlebitis, arterial aneurysms, arterial occlusions, unexplained arthritis.