



An a«MAZE»ing case...

Abire Allaoui, Morocco

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Patient introduction

- Mrs A. F
- 63 years old
- No personal history
- Husband treated for pulmonary tuberculosis
- Family history:
 - Sister: SLE
 - Brother: Hypothyroidism (Origin?)
 - Daughter: Grave's disease

Story of the illness

2 Years

Poor general
condition

Weight loss

4 Months

Prolonged fever

Chronic
Diarrhea/Chronic
cough

- Hospitalisation: Infectious department
- First Hypothesis? First steps workup?

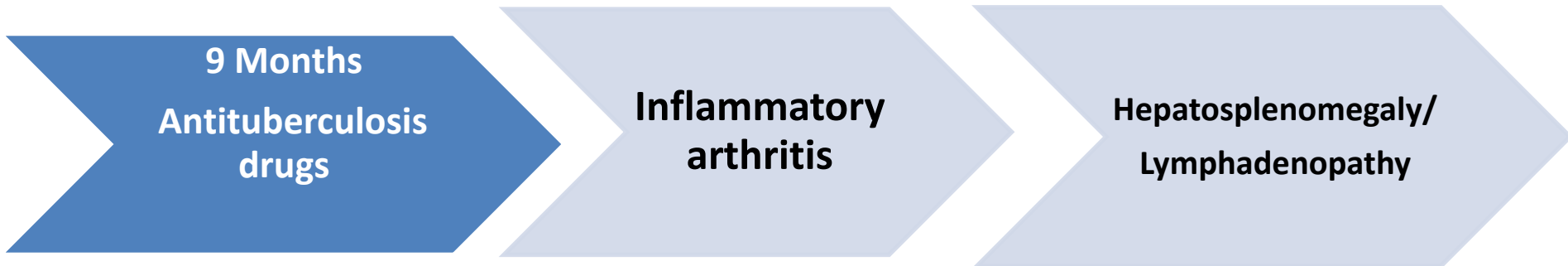
Lab tests

- Anemia 10 g/dl lymphopenia
- Inflammatory syndrom: ESR 73mm↑ CRP 68mg/l↑
- TST: negative
- HIV: negative
- Blood and stool cultures: negative
- Radio Chest/CT scan of the chest: micronodular and interstitial infiltrates, Bronchiectasies
- 3 BK expectorations: negative
- GI endoscopy : Nodular gastritis and colitis
Isolated Giardiasis



Next step??

Clinical course



- No improvement, New signs...
- Transfer to internal medicine department

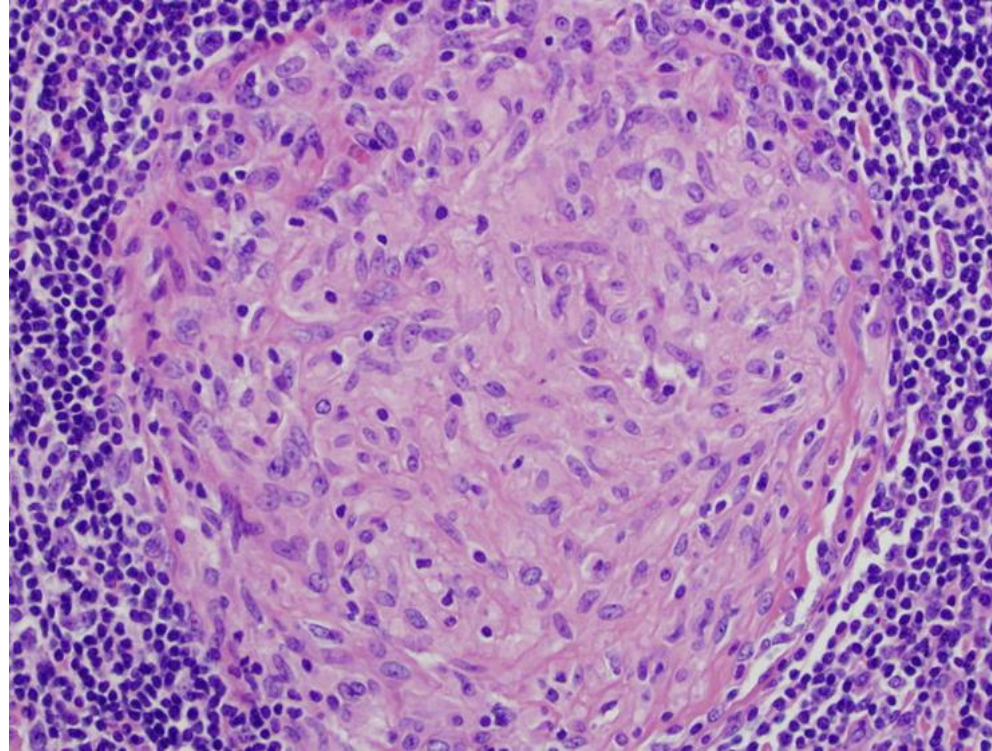
An autoimmune syndrome??

- Iron Balance, coombs (-): inflammatory anemia
- ANA (-), ANCA (-)
- Renal exploration negative

- Node biopsy

Non-necrotizing granuloma

- Ca^{2+} , Ph: normal
- ACE ↑
- Bone marrow examination: normal



Systemic sarcoidosis??Lymphoma?

Reanamnesis

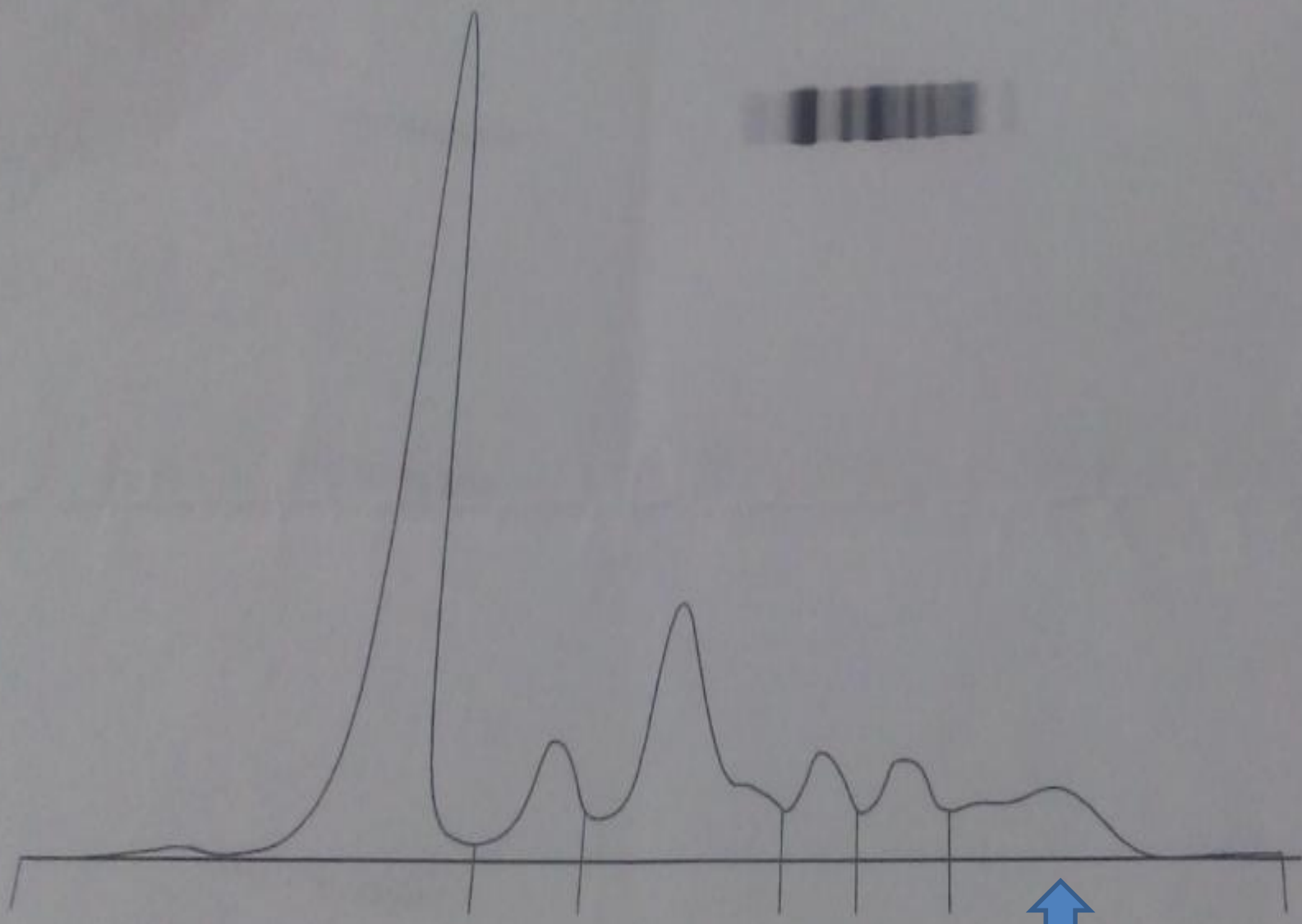
- Eye pain+ deminished eye vision since 2 weeks



Ophthalmologist examination: **Uveitis**

Electrophorèse des protéines sériques

sur Capillaires -sebia-



Further investigation...

- Serum levels of Ig : IgG 1.1↓ IgA 0.3 ↓ IgM 0.4↓
- Flow cytometric study: CD19 ↓↓: 18/μl
CD4:662 CD8:290
- Primary immunodeficiency?

Treatment and evolution

- Corticosteroids+ perfusion immunoglobuline
- Good results: quality of life
 - Gain weight
 - No fever

Take home messages

- No matter the age
- Unexplained prolonged signs: Why not PID?

Prolonged fever

Bronchiectasies

Chronic diarrhea /Giardiasis

Sarcoidosis like patterns



Protein Electrophoresis

Causes of hypogammaglobulinemia..More frequent

- Nephrotic syndrome
- Hemopathy
- Protein loss enteropathy
- Immunosuppressive treatment...

But hypoalbuminemia+++

Causes of granulomatosis

(1) Infections

Fungi

Histoplasma
Coccidioides
Blastomyces
Sporothrix
Aspergillus
Cryptococcus

Protozoa

Toxoplasma
Leishmania

Metazoa

Toxoplasma
Schistosoma

Spirochaetes

T pallidum
T carateum
T pertenuis

Mycobacteria

M tuberculosis
M leprae
M kansasii
M marinum
M avian
BCG vaccine

Bacteria

Brucella
Yersinia

(2) Vasculitis

Wegener's

Necrotising sarcoidal
Churg-Strauss
Lymphomatoid
Polyarteritis nodosa
Bronchocentric
Giant cell arteritis
Systemic lupus erythematosus

(3) Immunological aberrations

Sarcoidosis
Crohn's disease
Primary biliary cirrhosis
Hepatic granulomatous disease
Langerhan's granulomatosis
Orofacial granulomatosis
Peyronie's disease
Blau's syndrome
Hypogammaglobulinaemia
Histiocytosis X
Immune complex disease

(4) Leucocyte oxidase defects

Chronic granulomatous disease of childhood and adults

(5) Hypersensitivity Pneumonitis

Farmers' lung
Bird fanciers'
Mushroom workers'
Suberosis (cork dust)
Bagassosis
Maple bark strippers'
Paprika splitters'
Coffee bean
Spatlese lung

(6) Chemicals

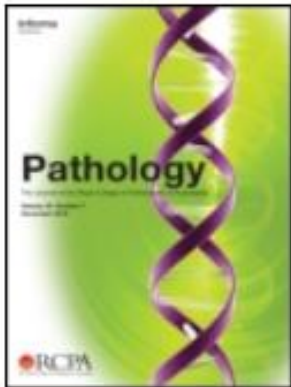
Beryllium
Zirconium
Silica
Starch
Talc

(7) Neoplasia

Carcinoma
Reticulosis
Pinealoma
Dysgerminoma
Seminoma
Reticulum cell sarcoma
Malignant nasal granuloma

(8) Miscellaneous infections

Whipple's disease
Cat scratch
Lymphogranuloma



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Recent onset granulomatous common variable immunodeficiency in an 88-year-old woman

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**Thank you for
your
attention**

