

What's this weird mush around the aorta ?

28th Course : Allergology and Immunology Update – January 24, 2026 – Interlaken

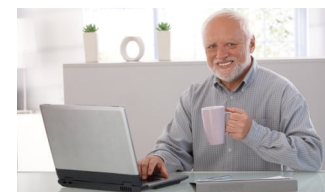
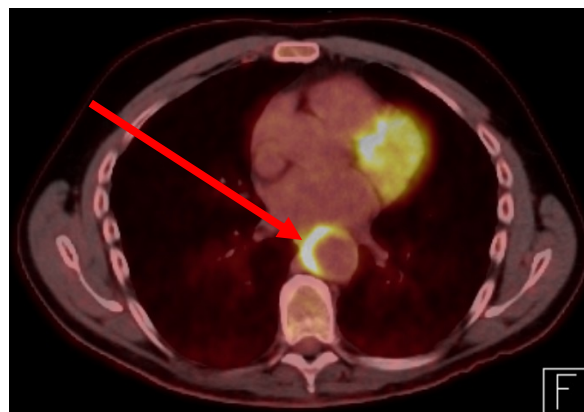
Case Presentations 3 : Clinical Immunology

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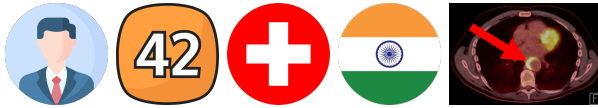
Case presentation : Periaortic lesion



Any ideas ?

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Case presentation



Social history :

- Born in Switzerland
- Parents of Indian origin
- Computer scientist
- Travels for work a lot
- No pets

Substance use history :

- Smoking : 10 pack-years, 
- Alcohol : **X**
- Drugs : **X**

Allergies :

- Aspirin : Skin reaction, no work-up

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Case presentation : Past medical history



PMH :

- Seminoma, left testicle (2023)
 - 2.2023 : Radical orchiectomy R0. Pure seminoma. Follow-up.
 - 3.2024 : Two left para-aortic retroperitoneal LN on abdominal CT.
 - 4.2024 : Combined radiochemotherapy (Carboplatin, 30 Gy).
- Chronic lower back pain (2000)
- Recurrent cystic skin lesions (2011)
- Warthin's tumor, right parotid gland (2024)
- Migraine (date unknown)

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Case presentation : Family history



FMH :

- Lung cancer in father
- Psoriasis in sister, brother, nephews

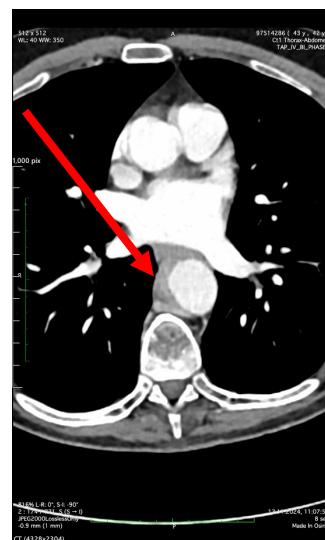
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Case presentation : History of present illness



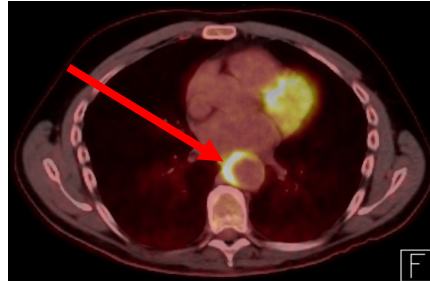
HPI :

- 12.11.2024 : Thoraco-abdominal CT
 - Not typical for inflammatory involvement because hemicircumferential



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Case presentation : History of present illness



HPI :

- 25.11.2024 : PET/CT
 - SUVmax 11.2
 - No other detectable sites of vascular involvement
 - Complete regression of the two hypermetabolic LN
 - **A neoplastic cause seems unlikely**
 - Differential : IgG4-RD? Vasculitis? CTD? Other?

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Case presentation : Review of systems

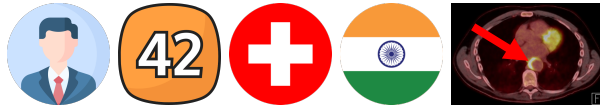


ROS :

- Patient **paucisymptomatic**
 - No general symptoms
 - Cardiac/respiratory : occasional **chest tightness**
 - GI symptoms : **recurrent oral aphthosis**
 - Skin : **cystic lesions** on cheeks, behind ears
 - Joints : **chronic lower back pain**, induced by cold, after exercise. No morning stiffness. Diagnosis of Scheuerman disease in 2004.
 - No urinary, neurological, ocular or ENT symptoms

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Case presentation : Physical exam



PE :

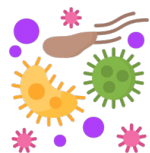
- Appears well, alert and oriented, vital signs stable and within normal limits
- Regular heart rate and rhythm, no murmurs
- Lungs clear to auscultation bilaterally
- Abdomen soft, non-tender, non-distended
- Normal peripheral pulses
- Normal neurologic exam
- No pain on lumbar spine percussion
- **Cystic scarring in cervical region**

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Work-up



- ESR 35 mm/h
- Normal creatinine
- No cytopenia
- ALP 1.5x ULN
- CRP 12 mg/L
- Normal SPEP
- SAA 21.9 mg/L



- Negative blood cultures (aerobic, anaerobic)
- QuantiFERON negative
- Brucella, Coxiella, Syphilis negative
- HIV, HBV, HCV negative



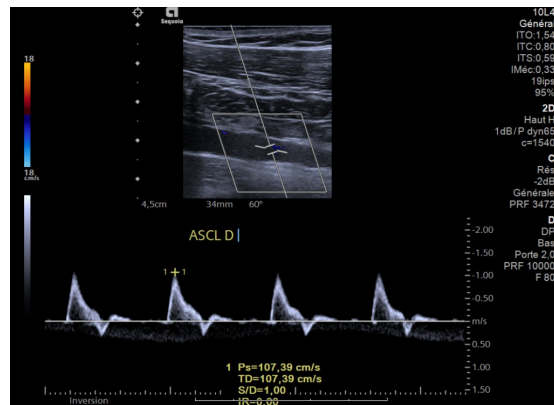
- ANA, anti-ENA, ANCA, RF, anti-CCP negative
- IgG4 0.53 g/L
- Plasmablasts 3550 cells/mL
- Serum immunofixation normal
- C3 normal, C4 0.38 (N<0.34)

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Work-up : Radiology



- **Doppler US of temporal, common carotid, and axillary arteries (1.2025):**
 - No wall thickening or sign of vasculitis



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Work-up : Tissue sampling

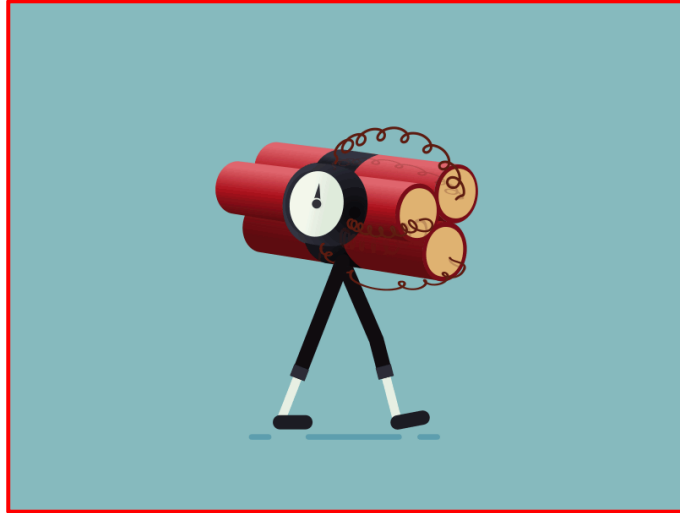


- **US-guided transesophageal periaortic fine-needle aspiration (3.2025):**
 - Cylindrical cells, squamous cells, lymphocytes, and blood
 - No cells suspected of malignancy

Representative of the lesion ?

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6 months go by...



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Case presentation : History of present illness

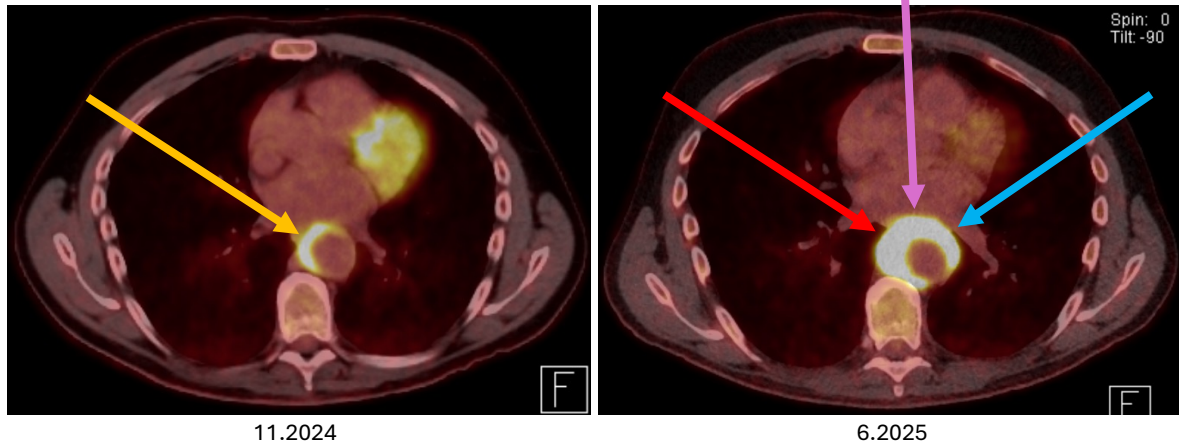


HPI :

- Patient **really symptomatic**
 - **Fever** (39.5°C), chills, **weight loss** 8 kg in 6 weeks
 - Loss of appetite, fatigue, **night sweats**
 - Oppressive **chest pain** in the evenings
 - Rest unchanged

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Work-up : Repeat PET/CT



11.2024

6.2025

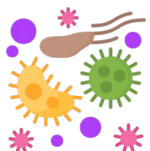
- Mediastinal and retrocrural LN suggesting 2^{ary} malignant lesion
- Primary inflammatory process unlikely

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Work-up : Standard labs



- ESR 88 mm/h
- Inflammatory anemia Hb 90 g/L
- No leukocytosis
- Thrombocytosis 428 G/L
- CRP 123 mg/L
- No renal involvement
- ALP 1.5x ULN, GGT 2x ULN
- Ferritin 700 ug/L
- Urinalysis unremarkable
- Repeat blood cultures negative (aerobic, anaerobic, mycobacteria)



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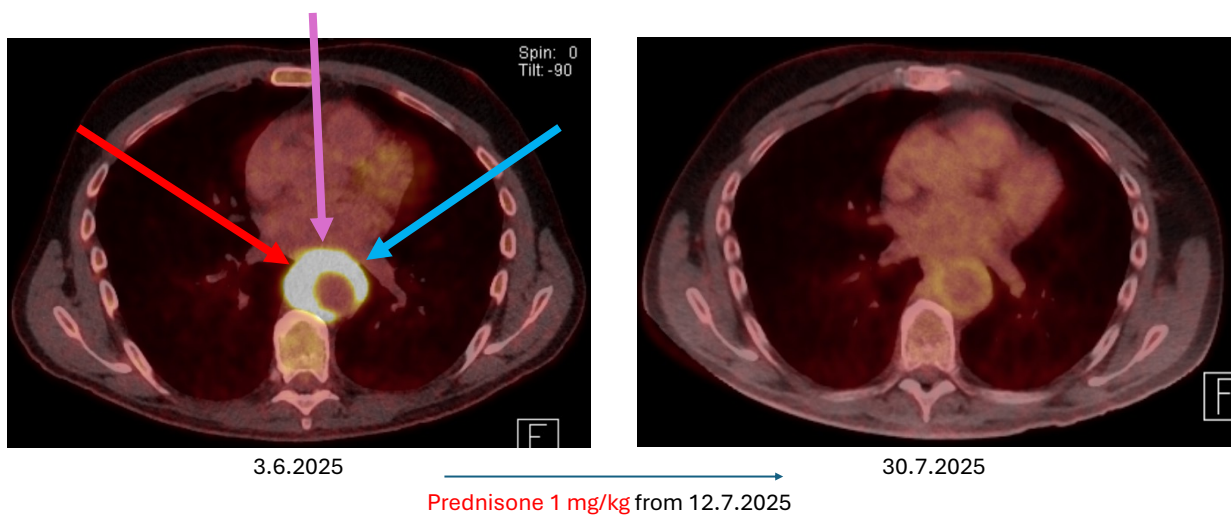
Work-up : Periaortic biopsy



- **Thoracoscopic periaortic biopsy (7.2025) :**
 - Myofibroblastic proliferation without high-grade atypia
 - Prominent inflammation
 - No evidence of seminoma
 - No evidence of lymphoma
 - ALK negative inflammatory myofibroblastic tumor ?

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Treatment



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What is the diagnosis ?

- 1 Steroid-sensitive periaortic myofibroblastic inflammatory tumor
- 2 Erdheim-Chester disease with superinfection by Brucella
- 3 Spondyloarthritis-associated periaortitis
- 4 Tertiary syphilis with falsely negative TPHA

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If we go back to the history...



- Chronic lower back pain (2000)
 - Family history of psoriasis
 - Pain controlled by **ecofenac**
 - Morning stiffness 2-3 hours
 - No uveitis, psoriasis, dactylitis, colitis
 - HLA-B27 negative

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10.06.2014

Docteur
Vincent FREIBURGHIAUS
Rue Laurent-Péroud 1
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Anterior spondylitis D12-L3 Bilateral sacroiliitis Parasyndesmophytes

CS

M. KANANI Aun Né le 23.12.1981 (32 ans)

IRM LOMBAIRE ET DES ARTICULATIONS SACRO-ILIAQUES du 06.06.2014

Prescripteur : Dr Vincent FREIBURGHIAUS

Indications : Suite de cas 20.09.14.

Technique : T1 T2 sagittal colonne STIR sagittal coupes myélographiques T2 axial SI STIR COR DP FS ax et T1 cor.

Description : Colonne : Présence d'un œdème osseux de la partie antéro-supérieure des vertèbres L2 et L3, et de la partie inférieure de la vertèbre L1 et de la partie supérieure de la vertèbre D12. Les disques par contre se présentent normalement. Pas d'argument parlant en faveur d'une discopathie. Présence de para-syndesmophytes antérieurs de L1 à L3. Discrete arthrose postérieure en L4-L5 et L5-S1. Au niveau des articulations SI on constate à droite une image de sclérose de l'os sous-chondral au versant du sacrum de 7 mm et de 18 mm à gauche en pondération T1. Cette image correspond à une infiltration graisseuse. Pas d'érosion visible.

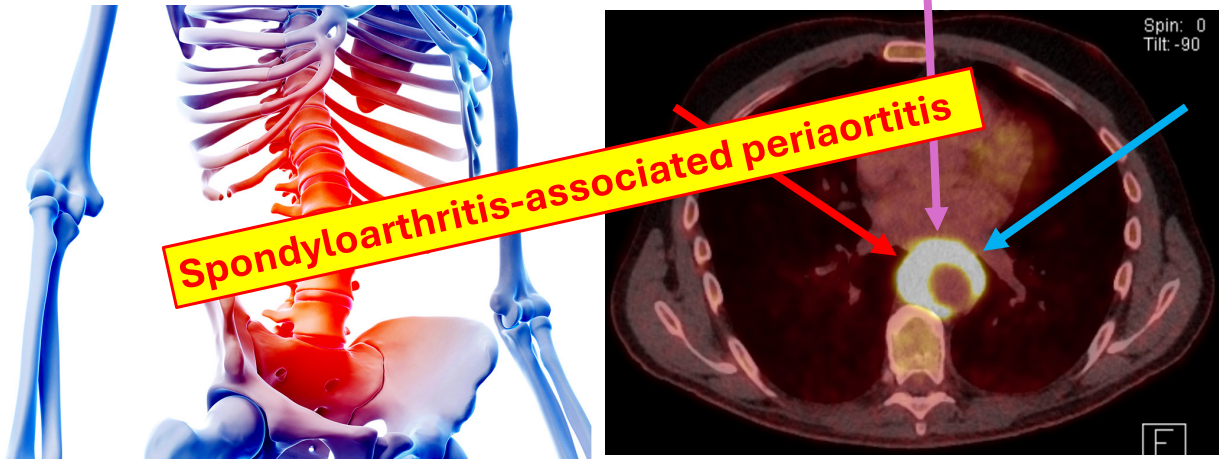
Conclusions : Examen mettant en évidence une spondylite antérieure de D12 à L3 et les 1ers signes d'une sacro-iliite ddc à prédominance gauche. Présence de para-syndesmophytes déjà visibles sur l'examen précédent. Le tout pourrait parler en faveur d'une spondylite et sacro-iliite dans le cadre d'un psoriasis.

Avec mes salutations les meilleures.

Docteur Roger AEBI
Signature électronique

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Final diagnosis



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1. Inflammatory myofibroblastic tumor

- **Rare mesenchymal neoplasm**
 - [myo]fibroblasts, inflammatory infiltrate
- Children, young adults
- Lungs, abdomen, retroperitoneum
- 50-80% kinase gene fusions (**ALK gene +++**)
- **Surgery + ALK inhibitors** in ALK-positive cases



Choi. Cancers. 2025

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2. Aortitis and periaortitis

- **Aortitis**
 - Aortic wall inflammation (GCA, TAK)
- **Periaortitis**
 - Inflammation extends to periaortic tissue (IgG4, IRF)
- Nonspecific symptoms or vascular complications
- **Histopathology = gold standard**
- Management depends on etiology
- Monitoring essential (relapse, progression)

Table 1. Classification of Aortitis and Periaortitis

Noninfectious aortitis	
Large-vessel vasculitides	
Giant cell arteritis	
Takayasu arteritis	
HLA B27-associated spondyloarthropathies ←	
Ankylosing spondylitis	
Reiter syndrome	
Immunoglobulin G4-related aortitis	
Variable-vessel vasculitides	
Behcet's disease	
Cogan's syndrome	
Relapsing polychondritis	
Rheumatoid arthritis	
Systemic lupus erythematosus	
ANCA-associated	
Wegener arteritis	
Polyarteritis nodosa	
Sarcoidosis	
Neoplastic ← lymphoma +++, metastatic lung or esophageal carcinoma	
Other	
Idiopathic aortitis	
Inflammatory aortic aneurysm	
Idiopathic retroperitoneal fibrosis (periaortitis)	
Radiation-induced aortitis	
Infectious aortitis	
Bacterial (e.g., Salmonella or Staphylococcus)	
Luetic (syphilis)	
Mycobacterial (e.g., Mycobacterium tuberculosis or Nocardia)	
Viral (e.g., hepatitis B, hepatitis C, and human immunodeficiency virus)	

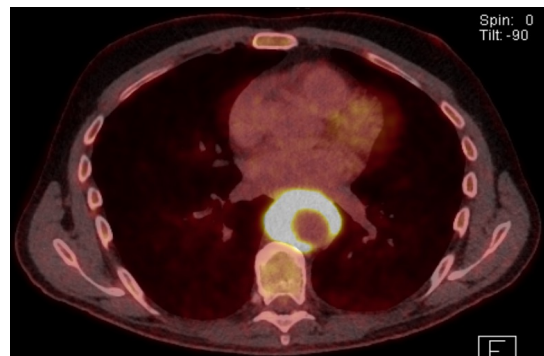
ANCA: anti-neutrophil cytoplasmic antibody.

Kadian-Dodov et al. JACC. 2022
Hernandez Mateo et al. Poster. ECR 2018

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3. Spondyloarthritis-associated periaortitis

- **Rare extra-articular manifestation**
- Aortic valvulitis, AVB, inflammatory aneurysm
- Asymptomatic or compressive effects
- Periaortic thickening and fibrosis, **inflammation**
- **Corticosteroids** +/- additional IMT (+/- surgery)



Palazzi et al. Joint Bone Spine. 2011

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