

Is it syphilis?

Patient case

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Symtoms

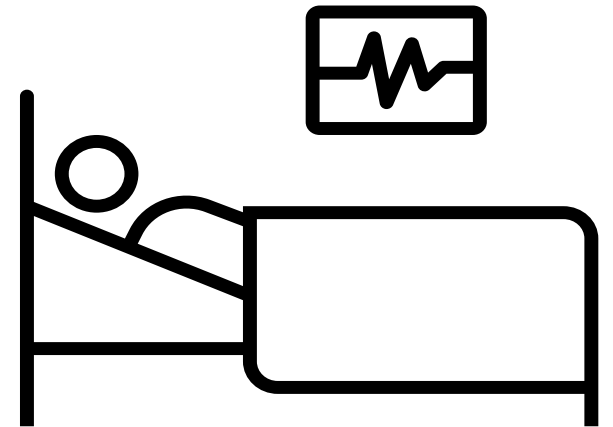
62-year old woman referred to Diagnostic Center Danderyds Sjukhus

- Urtikaria
- Fever
- Lymphadenopathy
- Artraligi
- Fatigue
- Palpable lymphadenopathy in both groins
- Urticarial skin rash
- Otherwise without remark

Symtoms stationary for 3 years with ESR 100 and CPR 100.

Past Medical History:

- Chronic urticaria for several years, treated with Xolair without effect.
- History of unclear seizure episode; treated with Keppra for several years, now discontinued.
- Symptomatic supraventricular and ventricular extrasystoles; extensive cardiac workup unremarkable.



Results

BLOOD

- CRP 198->104->134
- SR- 70-90 since 1 yr back
- Pro-Kalk ua
- Leukocytosis, trombocytosis and normocytic anemia
- S-prot.frak with inflammation
- and elevated polyclonal IgG
- FLC without remark
- ACE without remark

Findings from radiology

CT

- Lymphadenopathy in both groins and in retroperitoneum
- Lymphadenopathy in hilum and lung root
- Otherwise without remark

PET-CT

- Enlarged lymph nodes in the axillary, inguinal, and mediastinal regions, as well as along the aorta and iliac vessels. Markedly increased FDG uptake.
- Diffusely increased FDG uptake in the bone marrow, assessed as inflammation-related.
- Sarcoidosis?



Additional investigations

Lymph Nodes: Overall reactive appearance. Specimen sent to the hematopathology department for re-evaluation.

Hematopathology: In summary, reactive lymph node with no evidence of lymphoma.

Bone Marrow FACS: Normal. Biopsy: Essentially unremarkable; slight increase in macrophages, no granulomas

Consultations

Endocrinologist

Rheumatologist

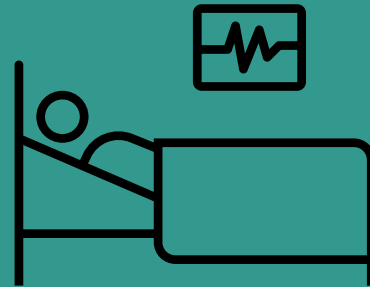
Neurologist

Pulmonologist

Venereologist

Otolaryngologist

Dermatologist



Through Venhälsan, the patient is undergoing treatment for possible neurosyphilis; however, after consultation with a syphilis specialist in Gothenburg, the findings are considered not consistent with neurosyphilis.



Is it Syphilis?

Diagnosis:

Atypical Schnitzler's syndrome.

Treatment initiated with Prednisolone, followed by an IL-1 inhibitor (Kineret) through the Rheumatology Department at Karolinska University Hospital.

Immediate clinical response noted regarding fever, urticaria, joint pain, fatigue, and inflammatory markers.

Currently maintained on Kineret monotherapy. Follow-up imaging shows regression of previous lymphadenopathy.



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Atypical Schnitzler's Syndrome

Atypical Schnitzler's syndrome is a variant of the rare disease Schnitzler's syndrome, characterized by symptoms resembling those of classical Schnitzler's syndrome but lacking the obligatory presence of a monoclonal gammopathy (most often of the IgM type) in the blood.

Instead, it may be associated with a monoclonal gammopathy of the IgG type, or there may be no monoclonal gammopathy at all.

The most common symptoms include recurrent fever, arthralgia, urticarial rash, fatigue, lymphadenopathy, and signs of systemic inflammation.

