

Unusual association of diseases/symptoms

Sweet's syndrome in a patient with rheumatoid arthritis, Sjögren's syndrome and lymph node tuberculosis

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Summary

Sweet's syndrome (SS) is an acute neutrophilic dermatosis characterised by abrupt onset of fever, leukocytosis and cutaneous eruption, with dermal neutrophilia on skin biopsy. Most cases are idiopathic but SS can be associated with various affections, especially neoplastic, inflammatory and infectious diseases. The authors report the case of an SS occurring in a patient with a known rheumatoid arthritis associated with a secondary Sjögren's syndrome, with incidental finding of concurrent lymph node tuberculosis. In case of SS, an associated disease (malignant, inflammatory or infectious diseases) must imperatively be searched for, knowing that two or more of these affections can coexist.

BACKGROUND

Sweet's syndrome (SS) is the most common neutrophilic dermatosis (ND). It is most often idiopathic but may be associated with a wide range of diseases, especially malignant conditions, infectious diseases or inflammatory diseases.

CASE PRESENTATION

We report the case of a 28-year-old woman with a 2-year history of seropositive and destructive rheumatoid arthritis

(RA) associated with a secondary Sjögren's syndrome, who was treated by methotrexate (15 mg/week) and low-dose prednisone (5 mg/day). She presented with a widespread cutaneous eruption that involved the face, trunk and four limbs, associated with fever, arthralgia, bilateral conjunctivitis and deterioration of general condition. This symptomatology occurs 5 days after a nasopharyngitis. Clinical examination objectified the fever at 38.5°C and demonstrated well-demarcated erythematous papules, with centrifugal extension involving the face, arm, back



Figure 1 Lesions of the arm: well-demarcated erythematous papules with centrifugal extension.

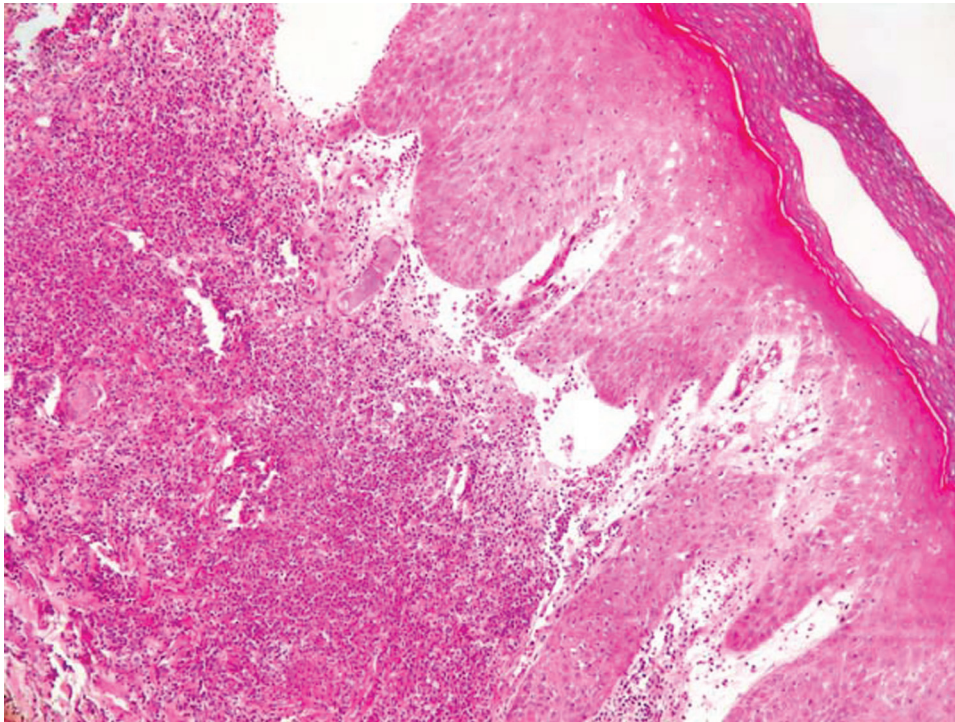


Figure 2 Skin biopsy H&E ×100: dense extensive neutrophilic infiltrate throughout the dermis, with prominent oedema in the upper dermis.

of the upper limbs, palms and knees (figure 1). Some of the papules were arranged in an annular pattern or became confluent to form plaques especially on the forehead and ears.

INVESTIGATIONS

Laboratory examinations found increased erythrocyte sedimentation rate (116 mm at the first hour), increased C reactive protein rate (120 mg/l) and leukocytosis with a neutrophil count of 8640 cells/mm³. A skin biopsy revealed a dense extensive neutrophilic infiltrate throughout the dermis with prominent oedema, mild endothelial swelling but no vasculitis was noted (figure 2).

SS diagnosis has been retained according to the criteria developed by Su and Liu and modified by Von den Driesch.¹ In addition to the two major criteria (abrupt onset of typical skin lesions and neutrophilic infiltration of the dermis), the patient had three minor criteria (fever and elevation in the erythrocyte sedimentation rate, C reactive protein level and neutrophil count). During follow-up, the systematic clinical examination also showed mobile and painless cervical lymph nodes. Cervical ultrasound revealed multiple cervical lymph nodes, the largest one measuring 5 cm long axis. The lymph node aspiration cytology was inconclusive. A tuberculin reaction was positive and the sputum test for *Mycobacterium tuberculosis* was negative. Chest x-ray was normal. A surgical adenectomy has also been performed and histology confirmed the diagnosis of lymph node tuberculosis.

TREATMENT

An antitubercular therapy was initiated (2 months quadruple therapy on isoniazide, rifampicin, ethambutol and

Learning points

- ▶ In case of SS, we must first search for a malignant condition (haematological or solid neoplasm) or more rarely an inflammatory disease (eg, RA, primary or secondary Sjögren's syndrome, systemic lupus erythematosus) or infectious disease (eg, tuberculosis).
- ▶ Two or more of these conditions can coexist.

pyrazoline, followed by 6-month bitherapy on isoniazide and rifampicin) with a favourable evolution.

OUTCOME AND FOLLOW-UP

No recurrence of SS was noted during 2-year follow-up.

DISCUSSION

ND and especially SS are frequently associated with various affections, including inflammatory and infectious diseases. The association of SS to RA is classical, recently a new form of ND, rheumatoid ND, has even been reported in patients with severe RA.² Also, several cases of SS associated with Sjögren's syndrome were reported in the literature.^{3 4} Some cases have also been described during or subsequently to an infectious disease.⁵ The association of SS with tuberculosis is not common, only few cases have been described in literature.⁶⁻⁸ In our case, SS occurs in a patient with a known RA associated with a secondary Sjögren's syndrome, with incidental finding of concurrent lymph node tuberculosis; this association has not been described previously to our knowledge.

Competing interests None.

Patient consent Obtained.

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