Unusual association of diseases/symptoms

Sweet's syndrome in a patient with Wegener's granulomatosis and ESRD

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Summary

A female patient in her late 60s with end stage renal disease secondary to Wegener's granulomatosis presented with a 2-day history of fever, nausea, somnolence and purple coloured exudative plaques on her forehead and index finger. Herpes zoster was the initial diagnosis and acyclovir was prescribed. Twenty-four hours later, her level of consciousness (LOC) had declined to a Glasgow coma scale of 10/15 and she started exhibiting myoclonic jerks. The lesions progressed to both sides of the face and acyclovir was discontinued as it was felt to be contributing to the myoclonic jerks. CT scan of the brain and lumbar puncture revealed no abnormalities to explain diminished LOC. Septic screen was negative, and an EEG showed no ictal activity. Skin biopsy revealed neutrophilic dermal infiltrates, leading to the diagnosis of Sweet's syndrome and prednisone (1 mg/kg) was initiated. The patient's condition improved over 12 h and she was discharged 5 days later. She died 6 weeks later from her cardiac disease.

BACKGROUND

This case represents an uncommon association of Sweet's syndrome in a patient with Wegener's granulomatosis.

CASE PRESENTATION

The patient presented with 2-day history of somnolence, nausea/vomiting and progressive purple coloured exudative plaques (figure 1) limited to the facial region and left index finger. She had a history of Wegener's granulomatosis, end stage renal disease (ESRD), hypertension, dyslipidaemia and frequent urinary tract infections. Her family and social history were non-contributory.

INVESTIGATIONS

- ▶ Complete blood count: leukocytosis
- ▶ Renal panel: elevated blood urea nitrogen/creatinine (ESRD)

Figure 1 Lesions on the face.

- ▶ Blood culture: no growth
- ▶ Urine culture: no growth
- ▶ Skin biopsy (histology): neutrophilic infiltration of dermis
- ▶ Skin biopsy (microbiology): negative for fungal stains, tuberculosis and bacterial cultures
- ▶ EEG: mild slowing over the right temporal region. No ictal activities or epileptiform discharges were identified.
- ▶ CT head: age-related changes. CT chest, abdomen, pelvis: unremarkable.

DIFFERENTIAL DIAGNOSIS

- ▶ Sweet's syndrome
- ▶ Leukaemia cutis
- ▶ Erythema multiforme
- ▶ Erythema nodosum
- ▶ Cicatricial pemphigoid
- ▶ Bullous pemphigoid.

TREATMENT

Prednisone 30 mg was given intravenously twice daily for 2 days, 60 mg orally once daily for 1 week (1 mg/kg), and then tapered by 10 mg per week until discontinuation.

OUTCOME AND FOLLOW-UP

There was complete resolution of cutaneous and neurologic symptoms without milia, scarring or neurologic deficits.

DISCUSSION

Sweet's syndrome is characterised by the abrupt onset of tender erythematous plaques or nodules, fever, neutrophilia, elevated erythrocyte sedimentation rate (ESR)/C reactive protein (CRP) and a sudden response to

corticosteroid therapy. Histopathologically plaques show an accumulation of neutrophils in the upper dermis.¹

Sweet's syndrome may be divided into three categories: idiopathic (classical), malignancy associated and drug induced.¹ Classical Sweet's syndrome occurs more commonly in women and is often associated with an upper respiratory tract or gastrointestinal infection, inflammatory bowel disease or pregnancy. It has also been reported in association with a wide variety of other infectious and autoimmune conditions.¹ Malignancy associated Sweet's syndrome typically arises in the setting of haematological malignancies; most commonly acute myelogenous leukaemia (AML). The syndrome has also been associated with other neoplasms including gastrointestinal, breast and genitourinary carcinomas.^{1 2} Numerous medications have been associated with Sweet's syndrome, with granulocyte-colony stimulating factor (G-CSF) being the medication most common therapeutic associated with the drug induced form.^{1 3}

Treatment of Sweet's syndrome consists of systemic corticosteroids. Other first line agents include potassium iodide and colchicine.¹ In classical Sweet's syndrome there is typically a prompt response to therapy.

Several hundred cases of Sweet's syndrome have been reported in the literature since it was initially described in 1964. It is our understanding that it has never, however, been reported in a patient with Wegner's granulomatosis.

Learning points

- ▶ Sweet's syndrome is a rare condition characterised by the abrupt onset of erythematous plaques or nodules, neutrophilia, pyrexia and elevated ESR/CRP.
- ▶ It is categorised as malignancy associated (most commonly haematological – AML), drug induced (most commonly G-CSF) or may be idiopathic.
- ▶ Idiopathic forms are often associated with underlying infectious or autoimmune disorders and pregnancy.
- ▶ Treatment generally consists of systemic corticosteroids with a prompt response.

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Competing interests None.

Patient consent Not obtained.

REFERENCES

1. **Cohen PR.** Sweet's syndrome—a comprehensive review of an acute febrile neutrophilic dermatosis. *Orphanet J Rare Dis* 2007;**2**:34.
2. **Cohen PR,** Kurzrock R. Sweet's syndrome and cancer. *Clin Dermatol* 1993;**11**:149–57.
3. **Walker DC,** Cohen PR. Trimethoprim-sulfamethoxazole-associated acute febrile neutrophilic dermatosis: case report and review of drug-induced Sweet's syndrome. *J Am Acad Dermatol* 1996;**34**(5 Pt 2):918–23.

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