Case Report

Campylobacter gastroenteritis associated with Sweet’s syndrome

Sumita Pai,1 Ed Rytina,2 Jane Sterling,3 J. A. Karas1 and S. H. Aliyu1

1Clinical Microbiology & Public Health Laboratory, Addenbrooke’s Hospital, Cambridge CB2 0QQ, UK
2Department of Histopathology, Addenbrooke’s Hospital, Cambridge CB2 0QQ, UK
3Department of Dermatology, Addenbrooke’s Hospital, Cambridge CB2 0QQ, UK

Sweet’s syndrome or acute febrile neutrophilic dermatosis has been associated with underlying infection, malignancy, inflammatory disease and certain medications. The infection agents associated with this include Streptococcus species, Yersinia species, Chlamydia species, Salmonella species and Helicobacter pylori. We report a case of Sweet’s syndrome in a 73-year-old woman following a 2 week course of severe gastroenteritis caused by Campylobacter species. Histological examination of skin lesions showed marked inflammatory infiltrate throughout the dermis, composed of neutrophils and histiocytes. The patient was successfully treated with topical and systemic steroids. To date, this is the first case of Sweet’s syndrome to be reported linked to Campylobacter species to our knowledge.

Case report

A 73-year-old woman with type II diabetes and a history of Duke’s colon cancer type A resected 16 years earlier presented with a 3 day history of bilateral conjunctival suffusion, arthralgia and discrete painful erythematous lesions initially involving her fingers, progressing over the next 24 h to involve her elbows, shins and trunk. The rash was preceded by a 2 week history of severe progressively worsening abdominal pain and watery non-bloody diarrhoea due to Campylobacter species. She was started on ciprofloxacin by her general practitioner 3 days after the onset of rash. Prior to this she was not on any medication.

On examination, she had multiple, well-defined, raised violaceous lesions on her extremities, trunk, arms and legs (Fig. 1). Her blood inflammatory markers were raised with a C-reactive protein of 139 mg l−1, erythrocyte sedimentation rate of 91 mm h−1, white cell count of 10.1 × 109 l−1 and neutrophils 8.69 × 109 l−1. An autoimmune screen (anti-nuclear antibody) was negative and she had normal complement levels. A surveillance colonoscopy done about 8 weeks earlier had been reported as unremarkable with no evidence of active malignancy or inflammatory bowel disease.

A skin biopsy showed marked inflammatory infiltrate throughout the dermis, composed of neutrophils and histiocytes with abundant nuclear dust plus perivascular lymphocytes but no vascular damage (Fig. 2). This was consistent with a resolving neutrophilic dermatosis with late histiocytic infiltration (Sweet’s syndrome).

Following the results of the biopsy, the patient was started on prednisolone (60 mg daily) and then subsequently discharged on a tapering prednisolone regimen with topical 0.05 % clobetasol propionate. Her gastroenteritis symptoms settled 2 days after admission and the ciprofloxacin was discontinued. At follow-up 4 weeks later, the cutaneous lesions had almost completely resolved. Two serial abdominal CT scans performed at 2 and 7 months after discharge showed incidental pancreatic cysts with no evidence of bowel cancer recurrence.

Discussion

Sweet’s syndrome or acute febrile neutrophilic dermatosis was first described by Dr Robert Sweet in 1964. It is an unusual abnormal hypersensitivity response characterized by an abrupt onset of widespread asymmetrical painful erythematous plaques or nodules typically involving the hands, arms, upper trunk and face accompanied by fever and neutrophilia. Skin biopsy demonstrates a characteristic dense perivascular and interstitial neutrophilic infiltrate without primary leukocytoclastic vasculitis. The disease predominantly affects women aged 30–50 years and older men (50–90 years old) with a female-to-male ratio of 2–3 : 1.

Several conditions have been noted to be associated or occur coincidentally with Sweet’s syndrome (Kim & Choe, 2010; O’Brien, 2005). These include haematological malignancies (especially acute myeloid leukaemia), solid tumours (breast, gastrointestinal and genitourinary),
inflammatory bowel disease, pregnancy and medications (in particular granulocyte colony-stimulating factor, all-trans-retinoic acid and vaccines) (Neoh et al., 2007; Kürkçüoğlu & Aksoy, 1997; Rubegni et al., 2001). Although quinolone usage has been linked to Sweet’s syndrome, our patient was started on ciprofloxacin 3 days after the onset of lesions.

Classical or idiopathic Sweet’s syndrome usually occurs after an infectious process and accounts for more than half of all cases (Vano-Galvan et al., 2010). In addition to the skin, neutrophilic infiltration of other organs such as bone, central nervous system, eyes and kidney is well described. Ocular involvement (i.e. conjunctivitis, periorbital and orbital inflammation) is variable in classical Sweet’s syndrome and uncommon in malignancy-associated and drug-induced forms of Sweet’s syndrome (Cohen, 2007). The ocular inflammation in Sweet’s syndrome occurs at the same time as, or a few days after, the appearance of the skin lesions (Gottlieb et al., 2008). Classical Sweet’s syndrome has been linked to respiratory tract (Streptococcus, Chlamydia) and gastrointestinal tract (Salmonella, Helicobacter pylori and Yersinia) pathogens (Kürkçüoğlu & Aksoy, 1997; Rubegni et al., 2001; Vano-Galvan et al., 2010). To our knowledge, this is the first case of Campylobacter-associated Sweet’s syndrome to be reported in the literature.

Prednisolone is the mainstay of treatment for Sweet’s syndrome, and in most cases a dose of 40–80 mg per day is extremely and rapidly effective. Recurrence of Sweet’s syndrome is reported in 25–30% of cases despite an initial excellent response. It generally develops as the steroid is being tapered (Gottlieb et al., 2008).

Campylobacter species are comma-shaped, Gram-negative, motile, non-sporing organisms. Acute Campylobacter enteritis is frequently self-limiting with gradual resolution of symptoms over several days. Approximately 10–20% of patients seek medical attention due to lack to resolution of symptoms or development of uncommon sequelae. The latter include Guillain–Barré syndrome, hepatitis, interstitial nephritis and reactive arthritis (Allos & Blaser, 2010). The only other post-infectious skin-related condition described with Campylobacter species is erythema nodosum (Ellis et al., 1982).

In conclusion, we report what we believe to be the first case of Sweet’s syndrome linked to Campylobacter infection and treated successfully with topical and systemic steroids. Sweet’s syndrome should be included among the list of post-infectious complications of Campylobacter gastro-enteritis.

References


