A rare case report describing the relation between sweet syndrome and spontaneous recurrent peritonitis

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ABSTRACT

INTRODUCTION: Sweet syndrome (acute febrile neutrophilic dermatosis) is a subset of rare inflammatory disorders, first described by Dr. Robert Douglas Sweet in 1964 [1]. It is characterised by the sudden appearance of painful cutaneous lesions (papules, nodules and plaques) which are both oedematous and erythematous [2,3]. Although most commonly it is accompanied by fevers and leucocytosis, there may also be a variety of extra-cutaneous manifestations involving the eye, musculoskeletal system and internal organs [2,4]. After excluding malignancy and drug exposure, most cases are described as “idiopathic” or “classical-type”, with a peak incidence between the ages of 30–60 [2].

Although the aetiology of this condition is not clear, it has been loosely hypothesised that infection can precipitate exacerbations of Sweet syndrome [5]. There are no previously documented reports describing an association between Sweet syndrome and spontaneous recurrent peritonitis.

PRESENTATION OF CASE: We report a case of a 37-year old female patient with known idiopathic Sweet syndrome, diagnosed on skin biopsy, who presented with multiple episodes of spontaneous peritonitis. Investigation through abdominal laparoscopy showed large amounts of free pus in the abdomen without bacterial isolation.

DISCUSSION: Differential diagnoses, investigations and management of suspected spontaneous peritonitis are discussed. It was suspected that her Sweet syndrome had caused a rare form of previously undescribed recurrent sterile peritonitis.

CONCLUSION: This case illustrates the importance of careful evaluation of patients with known inflammatory disorders, such as Sweet syndrome. It also demonstrates the need to have a multidisciplinary approach, by collaboration between the disciplines of medicine, surgery, microbiology and radiology.

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1. Introduction

Sweet syndrome (acute febrile neutrophilic dermatosis) is a subset of rare inflammatory disorders, first described by Dr. Robert Douglas Sweet in 1964 [1]. It is characterised by the sudden appearance of painful cutaneous lesions (papules, nodules and plaques) which are both oedematous and erythematous [2,3]. Although most commonly it is accompanied by fevers and leucocytosis, there may also be a variety of extra-cutaneous manifestations involving the eye, musculoskeletal system and internal organs [2,4]. After excluding malignancy and drug exposure, most cases are described as “idiopathic” or “classical-type”, with a peak incidence between the ages of 30–60 [2].

Although the aetiology of this condition is not clear, it has been loosely hypothesised that infection can precipitate exacerbations of Sweet syndrome [5]. There are no previously documented reports describing an association between Sweet syndrome and spontaneous recurrent peritonitis.

2. Case presentation

A 37-year-old lady presented to hospital with a 48-h history of right sided abdominal pain and nausea, with no vomiting and previously normal bowel movements. She had no urinary symptoms or vaginal discharge and her last menstrual period (LMP) was 8 days previously. The patient also reported to have never been sexually active.

She was diagnosed with Sweet syndrome, at another hospital, in 1998 on skin biopsy after previous recurrent superficial skin abscess requiring surgical management and presence of erythematous nodules. The first episode of abdominal pain in 2000 was when she presented with left iliac fossa pain and pyrexia. On this occasion she had an initial period of conservative management followed by an abdominal laparoscopy which, showed a large volume of frank pus in the pelvis and right para-colic gutter, associated with dense pelvic adhesions. Owing to the complexity of the case the operation was converted to open surgery for examination and lavage, with the appendix (although grossly normal) also removed. The pus sent for analysis had no bacterial growth, which was initially presumed due to pre-operative antibiotic use.

The patient underwent a further elective abdominal laparoscopy in 2014 for ongoing lower abdominal pains. This showed a large amount of pus in the pelvis and abdomen (Fig. 1), including a large right sub-phrenic abscess. There were extensive adhesions...
between the uterus and posterior pelvic wall with bowel adhered to the pelvic side wall. Adhesiolysis and peritoneal lavage were carried out and the patient was started on antibiotics with a presumed diagnosis of pelvic inflammatory disease (PID). However, as the patient had never been sexually active a suspicion was raised that this could have been related to her pre-existing Sweet syndrome. Of note, she was started on an acute prescription in the one month period prior to her latest admission, for an acute relapse of Sweet syndrome.

On this admission the patient presented febrile, with a temperature of 38.5 °C, tachycardia and low blood pressure. Abdominal examination revealed a soft but tender right iliac fossa, with features of peritonitis. Blood tests showed raised white cell count (WCC 17.1 × 10/L and neutrophils 15.1 × 10/L) and CRP (218 ng/L); renal and liver function and amylase were normal. Abdominal and chest x-rays showed no abnormalities. Bloods cultures were negative after 5 days on incubation.

CT Abdo-Pelvis with contrast (Figs. 2 and 3) which showed the presence of adnexal collections. The largest organising collections are on either side of the midline between the rectum and uterus (Figs. 2 and 3). The collection on the right was 5 × 4 cm in maximum transverse dimension and the left 4.5 × 2.5 cm (Fig. 3). There was associated small to moderate ascites mostly in the right paracolic gutter and around the liver. An incidental duplex left kidney. There was no free intraperitoneal gas. And no visible bowel pathology. The images were discussed in MDT and with gynaecology. The consensus was made that these low pelvic collections were probably secondary to pelvic inflammatory disease or of tube-ovarian origin.

Further to this discussion, an ultrasound of the abdomen and pelvis was performed, which reported: Poorly visualised pelvic...
structures transabdominally and that a trans-vaginal scan was not appropriate at this time as the patient had never been sexually active. Overall, the anteverted uterus contained one 10 mm intramural fibroid but was otherwise unremarkable with no adnexal masses and unremarkable ovaries. A large amount of free fluid was present within the pelvis both anterior to the uterus and in the cul-de-sac. A small amount of free fluid was also identified within Morrison’s pouch.

A gastroenterology opinion was sought, who suggested that the findings appeared to be consistent with a pelvic infection, exacerbated by a Sweet syndrome attack, with no features to suggest inflammatory bowel disease. A gynaecological review confirmed her last menstrual period (LMP) had been 8 days prior to admission and that she patient had never been sexually active. A cervical smear in the previous year was reported normal. Speculum examination showed no masses and only mild cervical excitation. At this stage, she was diagnosed with suspected PID of unknown cause and continued on a triple therapy of intravenous antibiotics (amoxicillin, metronidazole and gentamicin), with a subsequent MRI pelvis.

As she did not improve with conservative management a decision was made to undertake a further abdominal laparoscopy.

The findings showed (Fig. 4): Free blood stained, purulent fluid in the sub-diaphragmatic, sub-hepatic spaces and in the right paracolic gutter. There were features of reactive inflammation on the ovaries. The small bowel looked normal and there was only mild diverticulosis of the proximal sigmoid colon. The mid to distal sigmoid had a long stricture. The decision was made to wash out and leave a drain in the pelvis.

The patient made an uneventful recovery and was discharged from hospital with a drain in-situ which was later removed, and a course of oral antibiotics. Culture and sensitivity from abdominal fluid showed: copious white cells and pus cells but, no bacterial growth (aerobic or anaerobic) and no yeasts. Out-patient flexible sigmoidoscopy which showed no significant findings requiring further treatment.

3. Discussion

Spontaneous bacterial peritonitis (SBP) is defined as having infected ascitic fluid without evidence of any surgical intra-abdominal pathology [6]. This condition usually happens in patients with advanced cirrhosis and should be suspected in patients with cirrhosis who develop suggestive symptoms like pyrexia, abdominal pain and tenderness.

It is believed that one of the early stages in developing SBP is the overgrowth in bowel flora and the extra intestinal dissemination of specific organisms which are often coliforms. Altered bowel motility and increased intestinal permeability are associated factors [7,8]. A diagnosis of SBP is confirmed if the neutrophils count in the ascitic fluid is >250 cells/mm³, culture results are positive, and secondary causes of peritonitis have been excluded [10]. Other conditions which can give similar presentation include Tuberculous peritonitis [11] and malignancy-related ascites [12]. The mainstay of treatment is antibiotics for these patients.

In contrast, our case shows a young woman who seemingly develops recurrent episodes of peritonitis with no organism involved. It can be hypothesised that this may be an extracutaneous manifestation of Sweet syndrome, where neutrophils may infiltrate the peritoneal cavity causing widespread reaction and development of pus and adhesion formation. It could be further hypothesised that in the presence of steroids this increased the translocation of white cells and macrophages into the peritoneum creating the acute abdominal crisis which she experienced.

A literature search of EMBASE, MEDLINE, UpToDate and Clinical Key reported no similar published cases in the medical literature of sterile recurrent peritonitis related to any inflammatory disorder. This appears to be the first documented case highlighting a potential relationship between idiopathic Sweet syndrome and recurrent peritonitis. The main learning point is that patients with inflammatory conditions such as this should be investigated thoroughly to rule out any common causes for bacterial peritonitis (e.g. PID, appendicitis, diverticulitis). Presently we do not know whether
Sweet syndrome has any real relationship to the development of bacterial peritonitis. However, we have illustrated here that even in the absence of antibiotics, Sweet syndrome can be associated with a florid intra-abdominal disorder and ultimately the development of recurrent spontaneous peritonitic episodes.

Conflict of interest

There is no conflict of interest in this case to be declared.

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Ethical approval

No ethical approval was required for this case report.

Consent

Written consent was obtained from the patient for the publication of patient-specific information pertaining to this case report and accompanying images.

Authors contribution

Mr. Yaseen Rajjoub and Ms. Nadia Saffaf (First authors) Contributed to: study design, data interpretation, writing paper.
Mr. A. Goodman
Contributed to: supervision of project, proof reading.

Guarantors

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References

[1] R.D. Sweet, An acute febrile neutrophilic dermatosis, Br. J. Dermatol. 76 (1964) 349.
[2] P.R. Cohen, Sweet’s syndrome—a comprehensive review of an acute febrile neutrophilic dermatosis, Orphanet J. Rare Dis. 2 (2007) 34.
[3] P.R. Cohen, H. Hongsmann, R. Kurzrock, et al., Acute febrile neutrophilic dermatosis (Sweet syndrome), in: L.A. Goldsmith, S.I. Katz, B.A. Gilchrest (Eds.), Fitzpatrick’s Dermatology in General Medicine, vol. 1, 8th ed., McGraw Hill, 2012, pg. 362.
[4] S.L. Moschella, M. Davis, Neutrophilic dermatoses, in: J.L. Bologna, J.L. Jorizzo, R.P. Rapini (Eds.), Dermatology, vol. 1, Elsevier, 2008, pg. 379.
[5] Susanne S. Voelter-Mahlknecht, Bullous variant of Sweet’s syndrome, Int. J. Dermatol. 11 (2005) 901-905.
[6] B.A. Agba, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, the SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, Int. J. Surg. 34 (2016) 180-186.
[7] Conn: spontaneous peritonitis and bacteremia in laennec’s cirrhosis caused by enteric organisms, a relatively common but rarely recognized syndrome, Ann. Intern. Med. 60 (April (4)) (1964) 568–580.
[8] A.M. Madrid, F. Cumsille, C. Defilippi, Altered small bowel motility in patients with liver cirrhosis depends on severity of liver disease, Dig. Dis. Sci. 42 (April (4)) (1997) 738–742.
[10] J.G. McHutchison, B.A. Runyon, Spontaneous bacterial peritonitis, in: C.M. Surawicz, R.L. Owen (Eds.), Gastrointestinal and Hepatic Infections, WB Saunders Company, Philadelphia, 1994, p. 455.
[11] E.A. Akrivladis, B.A. Runyon, Gastroenterology 98 (1990) 127.
[12] D.J. Hillebrand, B.A. Runyon, W.G. Yasmineh, G.P. Rynders, Ascitic fluid adenosine deaminase insensitivity in detecting tuberculous peritonitis in the United States, Hepatology 24 (December (6)) (1996) 1408–1412.