

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

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ARTICLE INFO

Article history:

Received 13 February 2017

Received in revised form 18 July 2017

Accepted 18 July 2017

Available online 21 July 2017

Keywords:

Sweet syndrome

Peritonitis

Sterile peritonitis

Recurrent peritonitis

Case report

ABSTRACT

INTRODUCTION: Sweet syndrome (acute febrile neutrophilic dermatosis) is a subset of rare inflammatory disorders, first described by Dr. Robert Douglas Sweet in 1964 (Sweet, 1964). The co-existence of Sweet syndrome and spontaneous recurrent peritonitis has never been previously mentioned in the medical literature.

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.

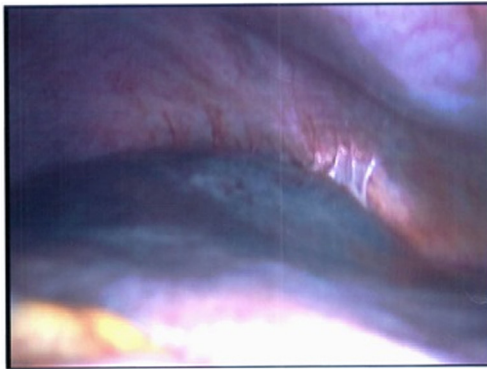
She was diagnosed with Sweet syndrome, at another hospital, in 1998 on skin biopsy after previous recurrent superficial skin abscess requiring surgical management and presence of erythematous nodules. The first episode of abdominal pain in 2000 was when she presented with left iliac fossa pain and pyrexia. 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

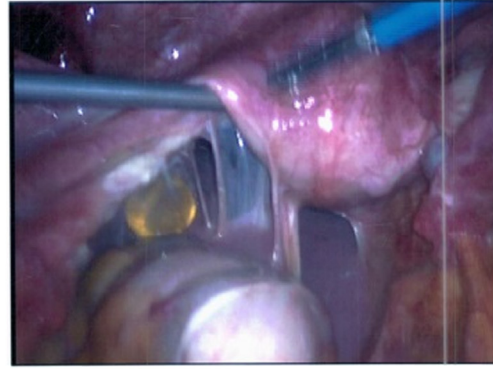
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female

Date of Birth:
Sex:

Still_009



Still_010



Still_011



Still_012

Fig. 1. Intraoperative laparoscopic stack images of the pelvis (June 2014): Each still image demonstrates the presence of pelvic sidewall and organ adhesions. Pus is seen clearest in still.010 as grey turbid fluid inferior to the uterus mobilised with laparoscopic instruments.

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 $17.1 \times 10^9/L$ and neutrophils $15.1 \times 10^9/L$) 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 prob-

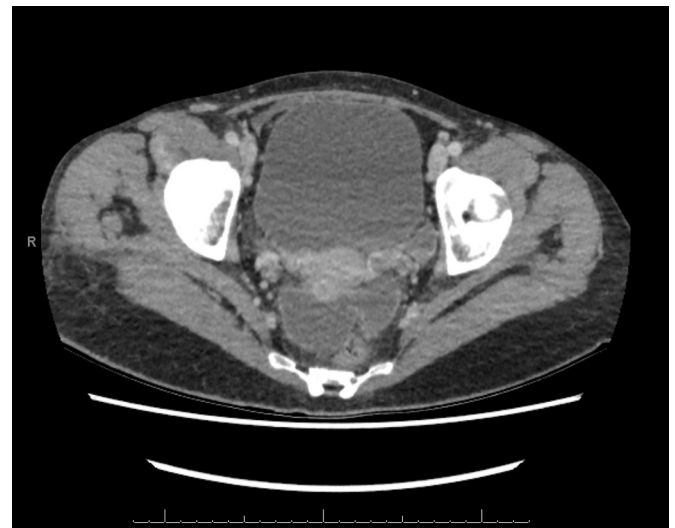


Fig. 2. Transverse plane image of CT scan pelvis: An image showing presence of an organising rectouterine collection with free fluid in the pelvic cavity.

ably 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

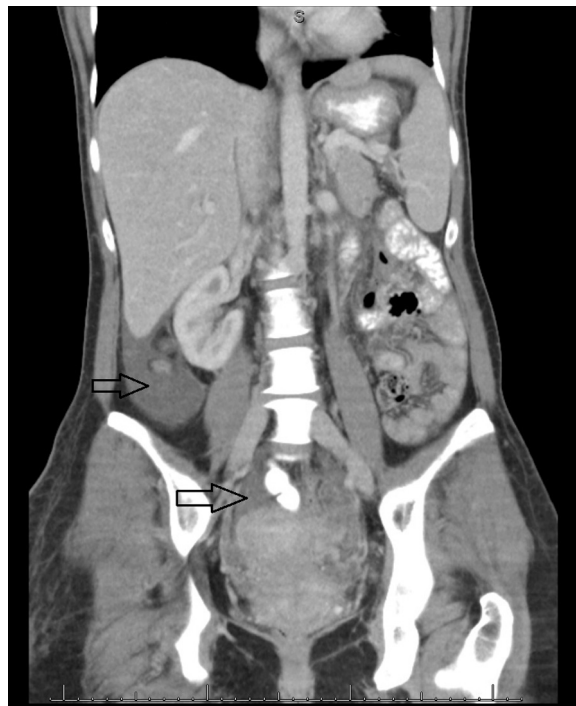


Fig. 3. Coronal plane image of CT scan abdomen and pelvis: The most obvious collections are indicated by arrows (upper: right upper quadrant collection, lower: rectouterine).

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 extra-cutaneous 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

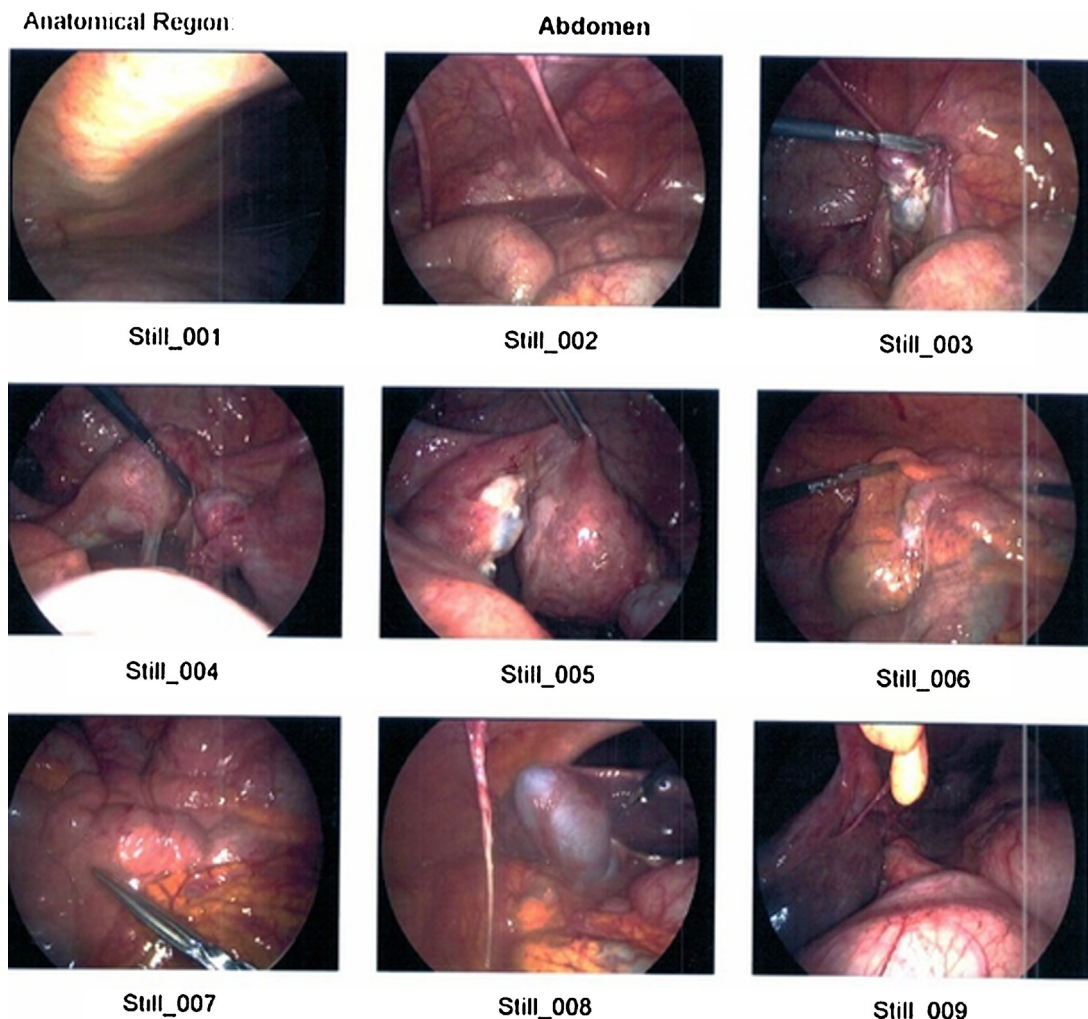


Fig. 4. Intraoperative laparoscopic stack images of the pelvis (January 2015): These images illustrate presence of adhesions (still.012–as the uterus is mobilised) and grey turbid fluid (still.011 – around the inferior aspect of the uterus).

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.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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

Mr. Y. Rajjoub.
Ms. N. Saffaf.

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