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# 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 (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.



## 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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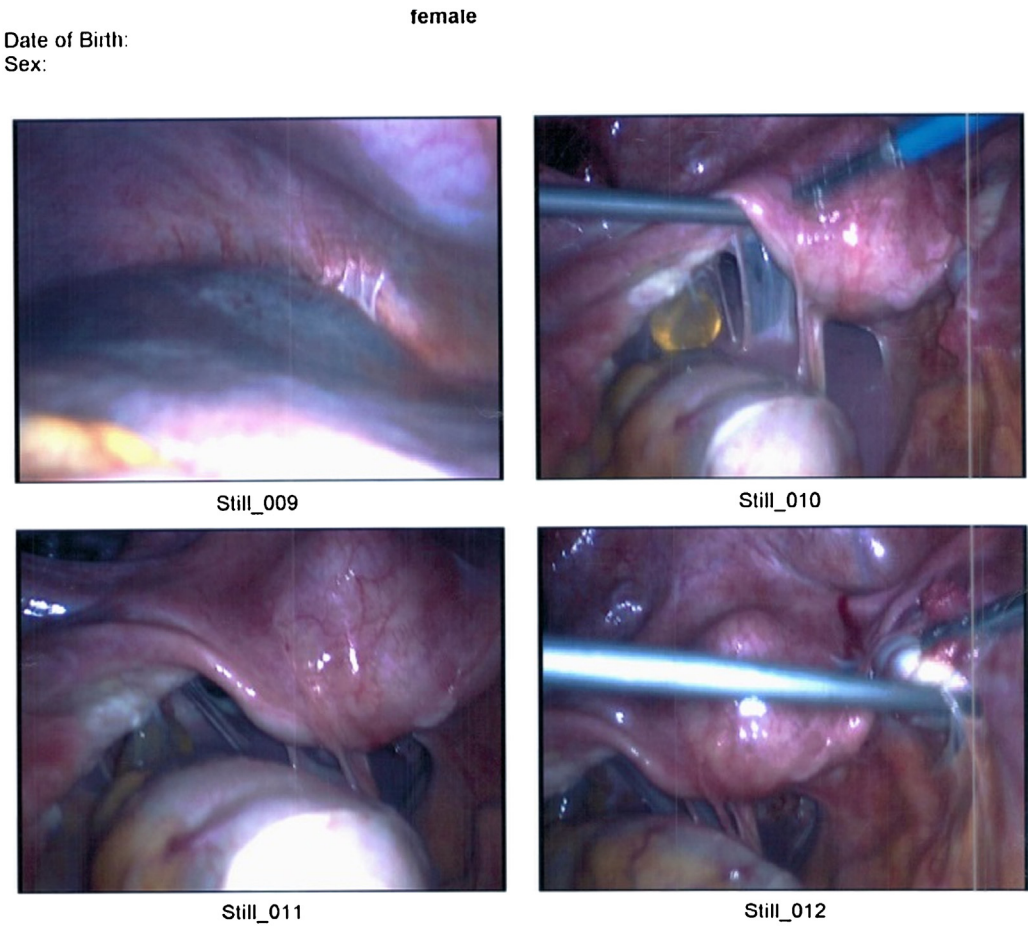


**Fig. 1.** Intraoperative laparoscopic stack images of the pelvis (June 2014): Each still image demonstrates the presence of pelvic sidewall and organ adhesions. Pus is seen clearest in still_010 as grey turbid fluid inferior to the uterus mobilised with laparoscopic instruments.

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 × 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.

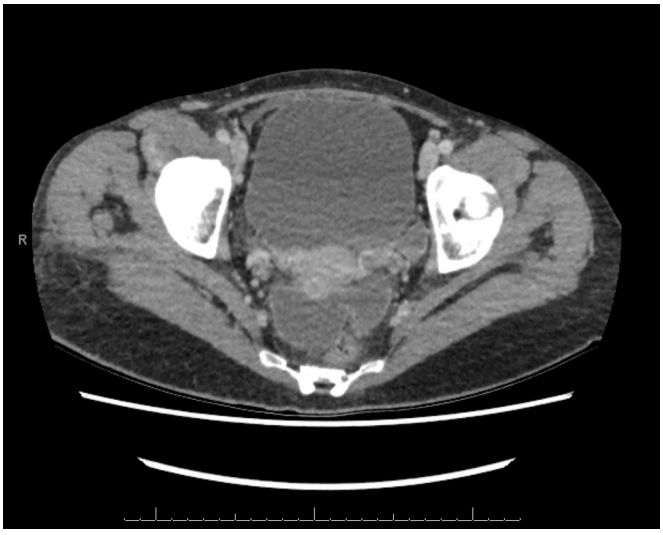


**Fig. 2.** Transverse plane image of CT scan pelvis: An image showing presence of an organising rectouterine collection with free fluid in the pelvic cavity.

Further to this discussion, an ultrasound of the abdomen and pelvis was performed, which reported: Poorly visualised pelvic