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Case Report

Necrotic pseudoxanthomatous nodules of the ovary and peritoneum presenting as haemorrhagic ascites

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Abstract

A 30-year-old woman presented with a four-week history of abdominal swelling. She had a long history of hormonal treatment for infertility but pregnancy test was negative. On ultrasound she was seen to have a large volume of ascites which was found to be haemorrhagic on insertion of an ascitic drain. Ascitic fluid cytology, culture and tuberculosis PCR were negative. An MRI pelvis confirmed large volume haemorrhagic ascites and showed a left sided ovarian cyst with possible haemorrhage. At laparoscopy the most notable findings were widespread necrotic-looking masses, biopsies of which excluded malignancy but revealed necrotic pseudoxanthoma cells, a rare manifestation of endometriosis. Although the patient initially declined surgery and treatment for her endometriosisin order to try for a pregnancy with in vitro fertilisation her symptoms recurred. She thus proceeded to laparoscopy andremoval of ovarian cysts. She is currently on goserelin with no recurrence of symptoms

Key Words:

Ascites, haemorrhagic, pseudoxanthoma, endometriosis, ovarian cyst, nodule

Introduction

New blood-stained ascites is often caused by malignancy or tuberculosis. Although endometriosis has been previously described to be a rare differential for this clinical presentation, the presence of necrotic pseudoxanthoma cells is a much rarer entity. Thus, early gynaecology team review with expert histological examination is crucial to unequivocally exclude malignancy and provide a definitive diagnosis.

Case Presentation

A 30-year-old Afro-Caribbean woman presented to the emergency department with a four-week history of abdominal swelling. She had no associated pain or fever and appeared systemically well. She had a history of helicobacter pylori gastritis, for which she had received eradication therapy and menorrhagia secondary to fibroids. She was nulliparous with one previous miscarriage. She had undergone ovarian stimulation from the age of 22 to 24 years, and for a three-month period ending six months prior her admission. She was a non-smoker with an insignificant alcohol history. She was originally from the Congo but had lived in the UK for 9 years, working in healthcare. On examination, she had a distended abdomen with shifting dullness but no organomegaly and normal bowel sounds. Other systems examination was unremarkable. Initial full blood count demonstrated Hb 95g/L and blood film showed a microcytic, hypochromic anaemia. She had a mildly raised C-reactive protein at 19mg/L and normal amylase at 83 U/L. She had normal renal profile, liver function, clotting. Urine pregnancy test was negative and serum HCG was < 0.1 U/L. Ultrasound of the abdomen and pelvis showed marked ascites with 1600cc of free fluid. The uterus contained multiple fibroids and the left ovary contained a dominant follicle. There were no abnormalities of the other abdominal or pelvic organs. An ascitic drain was inserted under ultrasound guid-

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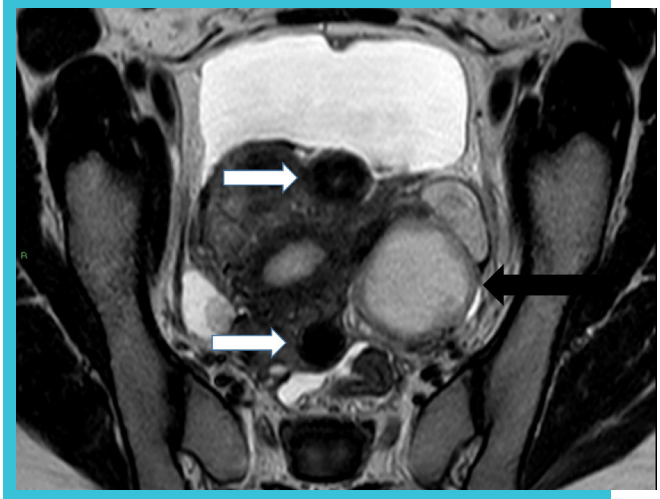
ance and 3 litres of heavily blood stained fluid was drained. Microscopy of the ascitic fluid demonstrated scanty white blood cells with no organisms seen. Tuberculosis culture and PCR were also negative. Fluid protein was 46 g/L and fluid glucose was 1.4 mmol/ L. Cytology did not demonstrate any malignant cells. Serum CA125 level was normal at 29 U/ml as were other tumour markers CEA, CA19-9 and alphafetoprotein. Immunoglobulins and auto-antibody screen were also normal. High vaginal swab demonstrated no bacterial or fungi and chlamydia nucleic acid amplification test (NAAT) was negative.

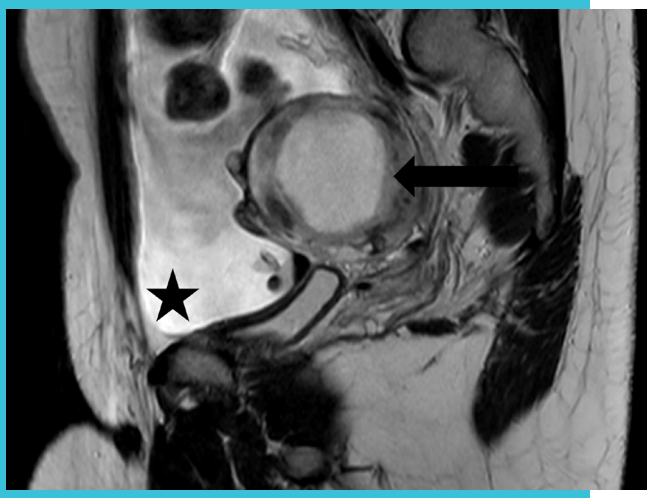
Figure 1.



Axial T2 weighted MRI images of the pelvis. These demonstrate a thick walled left adnexal mass (black arrow) and a fibroid uterus with typical signal intramural fibroids (white arrows).

In view of the blood-stained ascites, a CT thorax, abdomen and pelvis was organised to exclude malignancy. This confirmed large volume ascites, multiple uterine fibroids and a solitary indeterminate 3mm pulmonary nodule but no definite evidence of malignancy. The patient subsequently proceeded to have an FDG-PET which reported the lung nodule to be non-FDG avid, a small amount of non-specific heterogenous uptake was seen within the fibroids but no significant FDG-avid pathology to account for the ascites. She proceeded to an MRI pelvis (Figures 1 and 2) which again demonstrated a large quantity of peritoneal haemorrhage, fibroid uterus and a thick-walled cyst in the left ovary. The right adnexa contained a thickened serpiginous area of soft tissue possibly representing a thickened right fallopian tube.

Figure 2.



Sagittal T2 weighted MRI images of the pelvis. There is free intraperitoneal fluid with layering signal in keeping with blood (black star) and a thick walled left adnexal mass (black arrow).

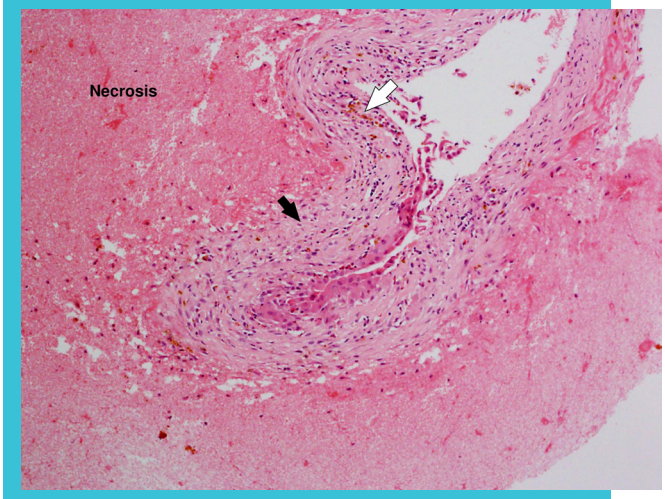
In order to obtain tissue diagnosis the patient was referred to the gynaecology team who performed a diagnostic laparoscopy. The most notable findings at the time of surgery were widespread necrotic-looking masses; the left adenexa was adherent to the pelvic side wall with necrotic material surrounding the Fallopian tube and ovary, superficial necrotic tissue was also seen over the right tube with necrotic tissue around the right broad ligament. Further, necrotic peritoneal tissue was seen free in the abdomen together with hepatic adhesions. The uterus was bulky with multiple fibroids but the endometrium appeared normal. Biopsies taken at laparoscopy revealed extensive necrosis and old haemorrhage involving the left and right adnexae, with no overt features of malignancy, but the cause of the necrosis was initially not apparent; thus making it difficult to make a long-term treatment recommendation.

To elucidate the cause of necrosis further, a CD10 stain was requested which showed strong positivity supporting the possibility of endometriosis and a further CD68 stain demonstrated strong positivity indicating the presence of necrotic pseudoxanthoma cells (Figure 3, 4). This rare entity is thought to represent an unusual manifestation of endometriosis which can be found in the peritoneum and ovaries. The patient was asymptomatic following laparoscopy and was therefore discharged from hospital. At follow-up three months later, her abdomen had become more distended and ultrasound of the abdomen and pelvis showed reaccumulation of ascites. Four litres of blood stained fluid was drained by paracentesis but the patient declined further intervention. The patient presented acutely six weeks later to another centre with abdominal pain and distension. MRI abdomen and pelvis demonstrated similar findings to previously and she again underwent paracentesis. A laparoscopy was performed with removal of bilateral ovarian cysts and biopsies from peritoneum and cysts were reported as endometriosis. She was started on a course of goserelin and remains asymptomatic at 2 months.

Discussion

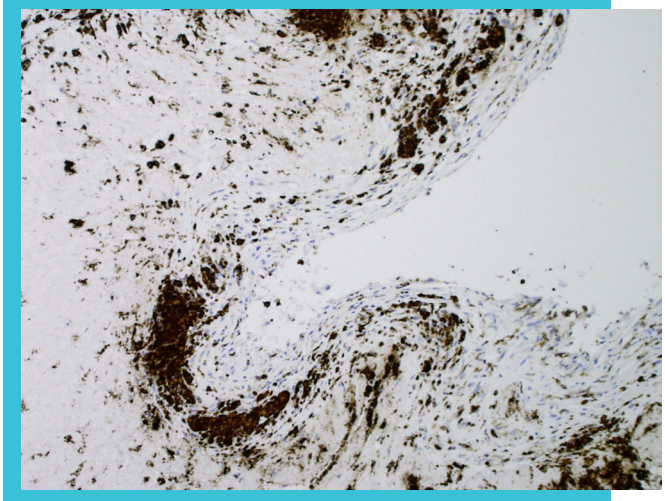
As this patient was systemically well with no detectable malignancy, tuberculosis or liver disease the abnormal appearances of the pelvic organs on MRI led us to suspect a benign gynaecological cause. Endometriosis generally involves the peritoneum, ovaries and the recto-vaginal septum but can affect other anatomical sites, for example the gastrointestinal tract, leading to symptoms such as bloody stools. However, the concurrent diagnosis of endometriosis and ascites is relatively rare and thus biopsies are required to confirm the diagnosis but also to exclude malignancy [1]. The finding of necrotic pseudoxanthomatous nodules at biopsy is a rarer entity with only a handful of cases having been previously reported. This disease entity was initially described in 1988 by Clement et al. who reported four cases of women who underwent surgery for uterine leiomyomas (n=2), uterine adenocarcinoma (n=1) and an enlarging abdominal mass (n=1) [2]. At the time of surgery all four were found to have nodules on the peritoneum or free within the peritoneal cavity.

Figure 3.



X20 magnification photomicrograph of biopsy from necrotic area in left adnexa. Large numbers of pseudoxanthoma cells (black arrow) are present beneath the epithelium. Some of these contain haemosiderin (white arrow). Large areas of necrosis are present on the left of the image.

Figure 4.



X20 magnification photomicrograph of biopsy from necrotic area in left adnexa demonstrating positive immunohistochemistry for CD68 on pseudoxanthoma cells

Histology of the ovaries and peritoneum demonstrated granulomas with central necrosis containing numerous necrotic histiocytes (pseudoxanthoma cells). Although endometriotic glands and stroma were not identified in the majority of the nodules the authors felt that this was likely to be an unusual end stage presentation of endometriosis with typical ovarian endometriosis having been found in all cases. All four patients were treated with definitive surgery–total abdominal hysterectomy, bilateral salpingo-oopherectomy and where indicated omentectomy with removal of the nodules with no evidence of recurrence of symptoms with follow up of 13 years, 8 months, 4 months and 4 years. Carey et al. reported the case of a 39-year-old woman presenting with an acute abdomen [3]. At laparotomy she was found to have a large ruptured ovarian tumour with large volume old blood in the abdomen. Histology of the ovarian mass revealed a grade 1 endometrioid cystadenocarcinoma and accompanying endometriosis. Interestingly, seven weeks later, when the patient was reoperated for staging laparotomy and completion of surgery she had developed new peritoneal nodules which were necrotic and contained numerous pseudoxanthoma cells. There was no obvious evidence of endometriosis in the pelvis or remaining ovary. The authors concluded that the appearance of the nodules may have been a non-specific pronounced tissue reaction to endometriotic cyst contents or blood.

Finally, Agarwal et al. report two cases of women presenting to gynaecology outpatients with infertility [4]. In the first case, a 24-year-old lady underwent laparacoscopy which showed multiple adhesions between the pelvic organs and nodules over the ovarian surface which were biopsied. In the second case, a 35-year-old lady underwent laparotomy which again showed multiple pelvic adhesions with a left endometriotic cyst and nodules on the left fallopian tube, both of which were sent for biopsy. Histology from the nodules in both cases showed granulomas with central necrosis and surrounding pseudoxanthoma cells and hyalinized connective tissue. The endometriotic cyst in the second case contained endometriotic glands and stroma. Our case further supports the association between endometriosis and pseudoxanthomatous nodules, though it remains unclear whether they form in association with the endometriosis itself or as a non-specific reaction to blood in the peritoneal cavity over a long period. This case has similarities to those previously reported, including the patient's complaint of infertility and the finding of haemorrhagic material in the abdomen. However, this case highlights the fact that pseudoxanthomatous nodules at laparoscopy may be the only indicator of endometriosis as initial biopsies may be non-diagnostic. The presence of pseudoxanthomatous nodules within the biopsies of patients presenting with large volume blood stained ascites is incredibly rare but needs to be recognised as a clinical entity to allow a definitive diagnosis to be made and thus the correct treatment to be recommended.

Acknowledgement
None

Declaration of Interest
None

References

1. Gungor T, Kanat-Pektas M, Ozat M, Zayifoglu Karaca M. A systematic review: endometriosis presenting with ascites. Arch Gynecol Obstet. 2011;283(3):513-8.
2. Clement PB, Young RH, Scully RE. Necrotic pseudoxanthomatous nodules of ovary and peritoneum in endometriosis. Am J Surg Pathol. 1988;12(5):390-7.
3. Carey M, Kirk ME. Necrotic pseudoxanthomatous nodules of the omentum and peritoneum: a peculiar reaction to endometriotic cyst contents. Obstet Gynecol. 1993;82(4 Pt 2 Suppl):650-2.
4. Agarwal K, Verma D, Kashyap A, Rautela A. Necrotic Pseudoxanthomatous Nodule: An Unusual and Less Known Manifestation of Endometriosis. World Journal of Pathology [Internet]. 2014; 4.