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Case Series

Rare and atypical presentations of endometriosis causing diagnostic dilemma and management challenges: an experience at a tertiary center

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ABSTRACT

Endometriosis is a disease that most of the times is diagnosed by its common presenting sign and symptoms, but can have varied presentations and may come as a surprise during surgery. We here present a case series of unusual presentations of endometriosis, which created diagnostic dilemma and challenges in the management of the cases at this tertiary center and share our experience. Endometriosis must be kept as a differential diagnosis whenever patient presents with acute pelvic or abdominal pain with adnexal masses and things are not fitting into straight forward diagnosis.

Keywords: Endometriosis, Infertility, Atypical, Massive hemoperitoneum, Adnexal mass

INTRODUCTION

Endometriosis is a very common disease with which women presents to gynaecologists. Its pathogenesis is related to estrogen hormone. It occurs mainly among women of young age. The estimated rate of prevalence is 5-10%.¹ Most of the times we are able to diagnose the disease by its common presentations but still the presentation can be so varied, it can put a very experienced clinician to test.

CASE SERIES

Case 1

A 42-year-old post cesarean status lady reported to a peripheral hospital with acute severe lower abdominal pain, right adnexal mass and hemoperitoneum. Her urine pregnancy test was negative. Preoperative imaging showed bilateral adnexal masses of around 4-5 cm with moderate hemoperitoneum. There were no retroperitoneal lymph nodes. Her CA125 was 138 u/ml, rest tumor markers were not sent. She was taken for emergency

laparotomy there. Intraoperatively she was found to have around 500 ml of hemoperitoneum with right adnexal mass densely adherent to uterus and large bowel and considered inoperable there. So hemoperitoneum was drained and a biopsy was taken from right ovarian cyst and abdomen was closed after putting intra-abdominal drain. Patient was managed conservatively and discharged few days later. She reported to our center with breathlessness after two weeks of discharge. On evaluation she was found to have hepatosplenomegaly, right adnexal mass, with large pelvic collection of size 11×10×10.6cm, with bilateral pleural effusion. Her ascitic fluid, pleural fluid, FNAC from adnexal mass came negative for malignancy as well as tuberculosis. Her tumor markers for epithelial and germ cell ovarian tumor except CA125 were normal. Patient was counselled for all possible differential diagnosis.

She underwent exploratory laparotomy with total abdominal hysterectomy with bilateral salpingo oophorectomy with evacuation of right pelvic hematoma with excision of multiple peritoneal nodules. (Figure 1). To our surprise frozen section showed benign hemorrhagic

cyst. Final histopathology (HPE) also showed endometriosis. Patient recovery was uneventful.

In this case the presence of haemoperitoneum, adnexal mass, dense adhesion with uterus, pleural effusion, hepatosplenomegaly with raised CA125 raised high suspicion for tuberculosis and ovarian malignancy but the diagnosis of endometriosis came as a surprise to us.

Case 2

A 32-year-old nulligravida reported to our center for pain lower abdomen and infertility of five-year duration. She had a history of laparoscopic ovarian cystectomy in 2017 for adnexal mass and also, she was given antitubercular treatment (ATT) for one-year duration till December 2021 elsewhere. On evaluation she was found to have large cystic left adnexal mass with CA125- 120 u/ml, CA19.9-44 u/ml, LDH- 820u/ml. Her CEA, AFP, β HCG were normal.

In this case the young lady had adnexal mass with raised epithelial and germ cell tumour markers with past history of adnexal mass with ATT taken. The differential diagnosis of extrapulmonary tuberculosis and ovarian malignancy was considered. After proper counselling for possible diagnosis, she underwent laparotomy with left ovarian cystectomy. Intraoperatively there were dense adhesions seen between mass, omentum and large bowel. During adhesiolysis the cyst got ruptured and purulent material was drained (Figure 2). There were multiple straw coloured fluid filled inclusion cyst seen. The frozen section and final HPE of cyst wall showed endometriosis and aspirate was sterile. Patient recovered well. The presence of purulent aspirate from the cyst during adhesiolysis was a surprise. The final histopathology was again endometriosis and the aspirate was sterile.

Case 3

24-year-old nulligravida lady with HBsAg positive status was evaluated initially at another center for distention of abdomen, loss of appetite with primary infertility. Clinically she was found to have shifting dullness. CT imaging showed normal uterus with left ovarian cyst of size 5x4.2x3 cm with no pelvic and mesenteric lymphadenopathy. Her CA125 was 38u/ml, CA19.9 was 45 u/ml and CEA was 2.7ng/ml. Her germ cell tumor markers were normal.

Ascitic fluid was negative for malignancy and tuberculosis. She was started on ATT at that center which she took for two months and then she stopped herself as there was no improvement. Then she reported to our center. After evaluation and counselling for possible differential diagnosis, she underwent laparoscopy with left ovarian cystectomy and drainage of approximately 3.5 liters of hemoperitoneum (Figure 3). Frozen section and final HPE of the ovarian cyst showed endometriosis and

haemorrhagic ascitic fluid was negative for the malignancy.

In this case the young lady had primary infertility, ascites, adnexal mass with near normal epithelial ovarian tumour markers. Here the presence of massive haemorrhagic ascites was a surprise. As this was a rare presentation of endometriosis so a search of literature was done and we could find only few case reports of endometriosis with massive hemoperitoneum.^{2,3}

Post-surgery patient recovered and conceived within two months and delivered at this center few months back.

Case 4

A 40-year-old para 2 status lady presented with pain lower abdomen of four to five months duration and large cystic right adnexal mass. Her CA125 was 283 u/ml. Her CA19.9 was 39 u/ml. All her other tumour markers including germ cell markers were within normal range. She never had any dysmenorrhea and dyspareunia. Her computer tomography scan (CT scan) showed large adnexal mass of size 14.6x13.8x9.2 cm, however there were no involvement of omentum or ascites and pelvic nodes. After counselling she underwent exploratory laparotomy with frozen section and proceed. Her frozen section of right adnexal mass showed possibility of hemorrhagic cyst with few areas showing atypical cells with high possibility of underlying malignancy.

She underwent complete primary surgical staging with total abdominal hysterectomy, left salpingo-oophorectomy, infracolic omentectomy, multiple peritoneal biopsies and bilateral pelvic lymph node dissection. Her final HPE came as a surprise and showed endometriosis. Patient recovered well and was kept under follow up for two years and she was asymptomatic.

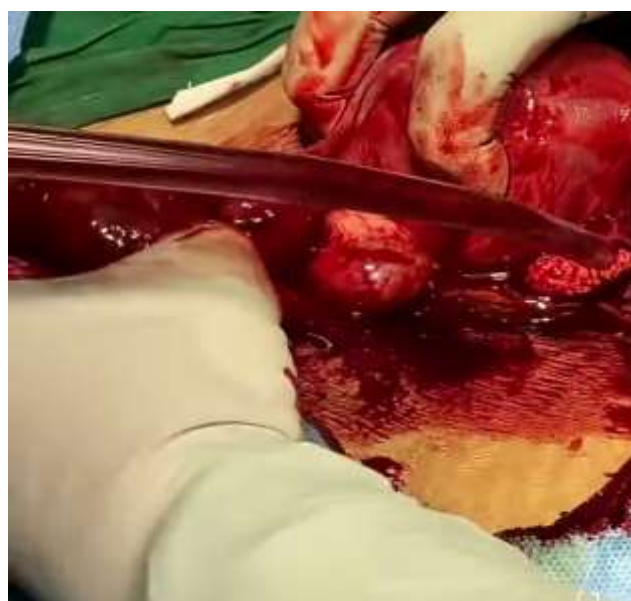


Figure 1: Haemoperitoneum found intraoperatively.

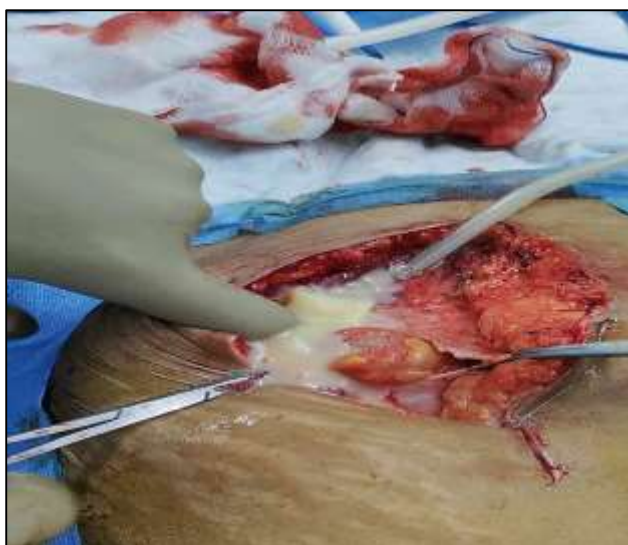


Figure 2: Pus like material drained from the cyst.



Figure 3: Haemoperitoneum seen intraoperatively.

DISCUSSION

Endometriosis is presence of endometrial tissue outside the cavity of uterus. The most common sites are ovaries, pouch of Douglas and pelvic peritoneum. Women with endometriosis may be asymptomatic or may have a spectrum of presentation ranging from infertility, pelvic pain, and pelvic masses. The most common symptoms with which a lady presents to gynaecology outpatients department are dyspareunia, dysmenorrhea and pelvic pain and abdomino-pelvic masses. Diagnosis may be challenging when endometriosis presents atypically with multisystem involvement.⁴ Though it is a disease of benign nature but due to its ability to infiltrate adjacent organs and cause dense adhesions, it distorts the anatomy and hence can have atypical presentation. Also, pelvic endometriomas which present as adnexal masses may

confuse with ovarian malignancy due to raised tumour markers like CA125.

Endometriosis may present as tuboovarian abscess if the adnexal mass gets infected. This happens more so in infertile patients as these patients have multiple risk factors like pelvic inflammatory disease (PID), egg retrieval, uterine instrumentation, procedures like hysterosalpingography, hysteroscopy etc.⁵ Recently bacterial implantation theory has also been suggested for this.⁶

Endometriosis presenting with massive ascites is very rare. Only few case reports are available in literature.^{2,3} Multiple explanations are suggested for presence of massive ascites with endometriosis. One explanation is the constant release of cells and chemicals from a ruptured endometrioma leading to pelvic inflammation.⁷ Another possible mechanism is peritoneal irritation due to endometriosis.

Sampson was the first to describe criteria for the diagnosis of a malignancy due to endometriosis.⁸ He suggested the presence of endometriosis near the tumour with excluding the possibility of invasion from other tissues.

In various studies the rate of malignant transformation is around 1-2%.⁹ Atypical endometriosis is a precursive lesion for endometriosis related epithelial ovarian cancer. Endometrioid and clear cell ovarian adenocarcinoma is found to have been associated with endometriosis.^{10,11} There is very high incidence of atypical endometriosis in endometriosis associated ovarian carcinoma. Atypical endometriosis is taken as a phase of transition from benign endometriosis to malignancy due to associated chronic inflammation and oxidative stress.

CONCLUSION

The ability to invade tissues and association for dense adhesions makes the clinical presentation so varied. Hence one can get surprises while managing the cases with atypical presentations of endometriosis. To conclude one must keep endometriosis as possible diagnosis while dealing with adnexal masses especially when the presentation is atypical.

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REFERENCES

1. Won S, Cho YJ, Lee N, Kim M, Kim MK, Jung YW, Yun BS et al. Atypical endometriosis is related to a higher recurrence rate. *Eur J Obstetr Gynecol Reproduct Biol.* 2020;254:44-51.
2. Togami S, Kobayashi H, Haruyama M, Orita Y, Kamio M, Douchi T. A very rare case of endometriosis presenting with massive

- hemoperitoneum. *J Minimally Invasive Gynecol.* 2015;S1553-4650(15)00241-1.
3. Kim BH, Park SN, Kim BR. Endometriosis-induced massive hemoperitoneum misdiagnosed as ruptured ectopic pregnancy: a case. *J Med Case Rep.* 2020;14(1):160.
 4. Bennett GL, Slywotzky CM, Cantera M, Hecht EM. Unusual manifestations and complications of endometriosis-spectrum of imaging findings: pictorial review. *AJR Am J Roentgenol.* 2010;194(6):WS34-46.
 5. Gao Y, Qu P, Zhou Y, Ding W. Risk factors for the development of tubo-ovarian abscesses in women with ovarian endometriosis: a retrospective matched case-control study. *BMC Women's Health.* 2021;21:43.
 6. Khan KN, Akira F, Koichi H, Michio K, Masahiro N, Shinji F et al. Bacterial contamination hypothesis: a new concept in endometriosis. *Reprod Med Biol.* 2018;17(2):125-33.
 7. Obajimi G and Awolude O. Endometriotic ascites: A very rare presentation of pelvic endometriosis *Ann Ibd. Pg. Med.* 2019;17(2):190-92.
 8. Sampson J. Endometrial carcinoma of the ovary arising in endometrial tissue in that organ. *Arch Surg.* 1925;10:1-72.
 9. Taniguchi F. New knowledge and insights about the malignant transformation of endometriosis. *J Obstet Gynaecol Res.* 2017;43(7):1093-100.
 10. Kobayashi H. Screening, epidemiology, molecular biology, and treatment strategies for endometriosis-associated ovarian cancer. *Reprod Med Biol.* 2010;9(1):17-22.
 11. Kurman RJ, Shih IM. The dualistic model of ovarian carcinogenesis: revisited, revised, and expanded. *Am J Pathol.* 2016;186(4):733-47.

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