Atypical Presentation of Endometriosis

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Introduction

Endometriosis is the presence of endometrial glands and stroma outside of the uterus. It is a benign inflammatory disease that affects generally reproductive age group females. But It can also be seen in premenarchal girls and postmenopausal women. Endometriosis is commonly found in approximately 10% to 15% of women between 25 to 44 years of age and in infertile women, the incidence is almost 25% to 40%.[1] The localization of endometriotic lesions can vary, with the most commonly involved organ is ovaries followed by the posterior broad ligament, the anterior cul-de-sac, the posterior cul-de-sac, and the uterosacral ligament.[2] Endometriotic nodules also affect the intestinal tract and the urinary system like the ureter, the bladder, urethra. Nevertheless, endometriosis is not limited to the pelvis but can damage extra pelvic structures like the pleura, the pericardium, or the central nervous system.[3] The clinical presentation of the disease differs in women and may be unexpected not only in the way presenting but also in the duration. The main clinical manifestations include chronic pelvic pain (dyspareunia, dysmenorrhea, dysuria, dyschezia) and infertility.[4] Here we are presenting a case of 27 years female with dysmenorrhea, infertility, massive haemorrhagic ascites and all preoperative investigations were suggestive of ovarian cancer. Endometriosis associated with the massive haemorrhagic ascites is a very rare presentation and only few cases have been reported yet. Differential diagnosis of women with haemorrhagic ascites includes rupture of haemorrhagic ovarian cyst, spontaneous rupture of spleen, ovarian cancer, hepatoma, tuberculosis, endometriosis, ectopic pregnancy, Meigs syndrome.[5] Other less frequent causes include mesothelioma, Budd Chiari syndrome, perforated duodenal ulcer, primary splenic lymphoma, mesenteric cyst, multiple myeloma, and cirrhosis. Endometriosis is an exceptionally rare cause of haemorrhagic ascites. Predominant theories regarding the pathophysiology of haemorrhagic ascites includes peritoneal irritation from ruptured endometriotic implants, sub diaphragmatic obstruction of lymphatics and retrograde menstruation.[6]

Case summary

My patient Mrs. Sanju Sahu, 27 years female, nulligravida, married for 2 years came with complaints of abdominal distension for 5-6 months and was anxious to conceive. Abdominal distension was insidious in onset and gradually progressive. She had also complaints of severe pain during menses, however her cycles were regular and flow was average. Ultrasonography was suggestive of bulky ovaries with ascites. CECT of abdomen done, suggestive of solid cystic lesion in bilateral adnexa, 4.2 x 2.9 x 3.3 cm on right side and 4.3 x2.4x 3.7 cm in left side with moderate ascites with mild omental caking. Her CA 125 was elevated (1243). CA 19-9, beta HCG, CEA, INHIBIN B were within normal limits. FNAC of ascitic fluid showed cluster of atypical cells which were positive for CK7 and PAX -8, also focal positive for ER, WT1 and p53 and were suggestive of metastatic deposits most likely from ovarian epithelium. PET scan was done suggestive of FDG solid cystic lesions in bilateral adnexa, likely primary neoplastic etiology with omental fat stranding, peritoneal deposit and gross ascites. On the basis of all reports, she was planned for staging laparotomy followed by chemotherapy in suspicion of ovarian malignancy. Before surgery, she was advised for ovum preservation as she was nulligravida. But patient wanted to go directly for treatment of ovarian mass.

Intraoperatively, 1500 ml greenish haemorrhagic ascitic fluid was present. Bilateral fallopian tubes and ovaries were found to be adherent to posterior surfacer of uterus and sigmoid colon. Bilateral tubes were oedematous, right ovary was grossly normal but in left ovary 4x3 cm cystic lesion was noted which was ruptured during dissection. Uterus was grossly normal. Adhesiolysis followed by left oophorectomy was done. Bilateral pelvic and paraaortic lymph nodes dissection, infra colic omentectomy were done. A mass of 5x5 cm noted in sigmoid colon and was adherent to the posterior wall of uterus. Hence, intraoperative Surgeon call was given. On further inspection, multiple mesenteric lymph nodes were found to be enlarged and biopsy was taken. Small bowel and liver micro deposits were also present. A postoperative colonoscopy was advised by surgeon for the mass over sigmoid colon. All samples were sent for histopathology examination. Her postoperative period was uncomplicated.

Histopathology findings of all samples were consistent with multifocal endometriosis involving tubo-ovarian complex, uterovesical fold and omentum. Based on histopathology report, she was put on injection leuprolide for 3 months. Colonoscopy was done in view of sigmoid colon mass which was normal. Repeat ultrasonography in July was normal. Patient is symptomatically better and is currently under follow up.

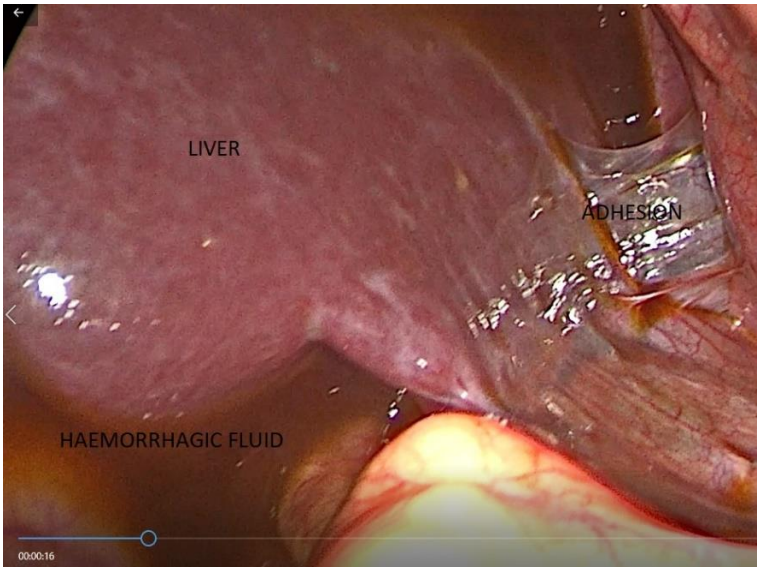

Figure 1: adhesions between liver and diaphragm and haemorrhagic ascites

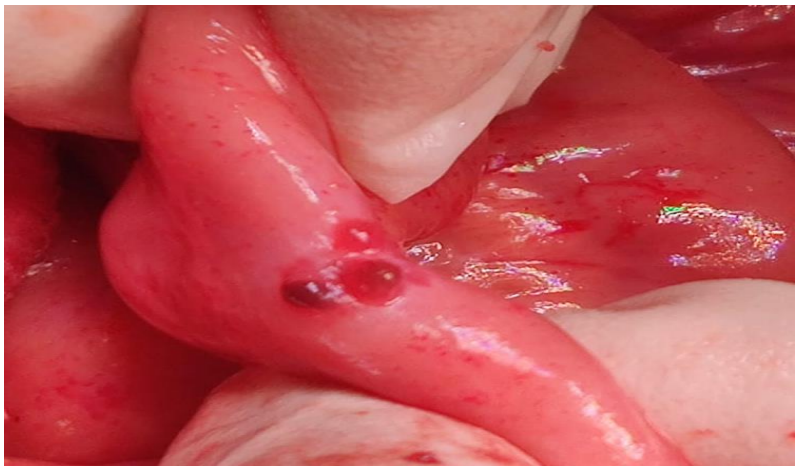

Figure 2: endometriotic spot

Discussion

Endometriosis is defined as the presence of endometrial glands and stroma like lesions outside of the uterus. The lesions can be peritoneal lesions, superficial implants, cysts in the ovary or deep infiltrating disease. Risk factors for endometriosis are nulliparity, early menarche and late menopause, shorter menstrual cycle, heavy menstrual bleeding and outflow tract obstruction. Factors which are decreasing the risk are multiple birth, increased lactation period, late menarche, OCP use and increase consumption of omega fatty acids. There is no definitive aetiology of endometriosis, there are several hypotheses regarding how endometriotic lesions

develop. Sampson's theory of retrograde menstruation is most accepted theory. Other theories are coelomic metaplasia by Meyer and Ivanoff, lymphatic and vascular spread metastatic theory by Halban, mutation of cancer drives genes like ARID1A, K RAS, PIK-3CA, AND PPP2R1A.

The degree of clinical manifestations of the patient is not directly associated with the extent of the disease or the size of endometriotic lesions.[7] The diagnosis of the disease usually delays in an average time of 4 to 11 years from the onset of symptoms[8] because of non-existence of a pathognomonic test or biomarker which detect the disease.

Hemorrhagic ascites should be considered a complication of endometriosis, especially in nulliparous women of childbearing age group with an abdominal distention, a pelvic mass, dysmenorrhea, abdominal pain, weight loss and eventual pleural effusion, suggesting a diagnosis of ovarian malignancy.[9] Other known causes of bloody ascites, especially a malignant process, must be excluded. Preliminary diagnosis of endometriosis is usually done on the basis of clinical history and physical examination. There is uterine or adnexal tenderness, a retroverted fixture, nodulating uterosacral ligament, and any pelvic masses may be present. Women with endometriosis show altered levels of CA-125, cytokines, angiogenic and growth factors compared to normal women, but none of the markers have been proven to be definitive clinical tool for diagnosis of endometriosis. On transvaginal sonography, ovarian cyst may be seen. The frequency of ovarian endometriomas accounts for approximately 17% to 44% in women with endometriosis, are bilateral in 50% of the cases.[10] Gold standard diagnostic tool of endometriosis is laparoscopy. They can appear as red, white, clear vesicular, black “powder-burns’’ or "gunshot’’ lesions.[11] The role of histological confirmation is a bit controversial since macroscopically detected endometriotic lesions cannot always be verified histologically and vice versa.[7]

The treatment of endometriosis is broadly categorized into two main categories, pharmacological and surgical. Currently, there is not a specific drug that could inhibit the progress of the disease.[12] First-line pharmacological therapy proposed for the management of endometriosis consists of non-steroidal anti-inflammatory drugs, progestins, or combined hormonal contraceptives. Combined hormonal contraceptives can be administered either cyclically or continuously, and they exert their effect by inhibiting follicular development, lowering the levels of LH and FSH, and leading to decidualization and atrophy of the ectopic endometrium. Progestins inhibits the ovulation and creating a hypoestrogenic milieu, and by

binding directly to the progesterone receptors in the endometrium, they cause decidualization and atrophy of endometriotic implants. As a second-line medical treatment is gonadotropin-releasing hormone (GnRH) analogues for the suppression of the endometriosis relating symptoms. They act by binding to pituitary receptors and downregulating the pituitary-ovarian axis. Surgical treatment is done for persistent pelvic pain that does not responded to medical management, evaluation of severe symptoms or treatment of anatomic abnormality. During laparoscopy all the endometriotic lesions and adhesions are excised. By the ablation of the endometriotic tissue, the local inflammatory milieu decreases in the pelvic cavity, thus increasing the chances for conception.

Conclusion

Endometriosis is a debilitating disease that impacts the quality of life of adult and adolescent patients. Presentation of endometriosis varies and it can also present with significant internal bleeding. Endometriosis should be considered in the differential diagnosis when bloody ascites and a pelvic mass are present, particularly in young female patients, for which there is no obvious explanation is present.

References

[1] . Talbert LM, Kauma SM. Endometriosis (1990) In: Danforth DN, ScottJR(Eds). Danforth's obstetrics and gynecology. (5thedn), Philadelphia: JB Lippincott.

[2] . Jenkins S, Olive DL, Haney AF. Endometriosis: pathogenetic implications of the anatomic distribution. Obstet Gynecol. 1986 Mar;67(3):335-8. [PubMed]

[3] . Macer ML, Taylor HS. Endometriosis and infertility: a review of the pathogenesis and treatment of endometriosis-associated infertility. Obstet Gynecol Clin North Am. 2012 Dec;39(4):535-49. [PMC free article] [PubMed]

[4] . Parasar P, Ozcan P, Terry KL. Endometriosis: Epidemiology, Diagnosis and Clinical Management. Curr Obstet Gynecol Rep. 2017 Mar;6(1):34-41. [PMC free article] [PubMed]

[5] . Otolorin EO, Ojengbede O, Falase AO (19987) Laparoscopic evaluation of the tubo-peritoneal factor in infertile Nigerian women. Int J Gynecol Obstet 25:47-52.

[6] . Palayekar M, Jenci J, Carlson Jr JA (2007) Recurrent hemorrhagic ascites: A rare presentation of endometriosis. Obstet Gynecol 110:521-522

[7] . Chapron C, Bourret A, Chopin N, Dousset B, Leconte M, Amsellem-Ouazana D, de Ziegler D, Borghese B. Surgery for bladder endometriosis: long-term results and concomitant management of associated posterior deep lesions. Hum Reprod. 2010 Apr;25(4):884-9. [PubMed]

[8] . Agarwal SK, Chapron C, Giudice LC, Laufer MR, Leyland N, Missmer SA, Singh SS, Taylor HS. Clinical diagnosis of endometriosis: a call to action. Am J Obstet Gynecol. 2019 Apr;220(4):354.e1-354.e12. [PubMed]

[9] . Dias CC, Andrade JM, Ferriani RA, Villanova MG, Meirelles RS (2000) Hemorrhagic ascites associated with endometriosis: A case report. J Reprod Med 45:688-690.

[10] . Bastos BB, Fonseca EKUN, Yamauchi FI, Baroni RH. Chocolate cyst with ground glass appearance in endometriomas. Abdom Radiol (NY). 2017 Dec;42(12):2957-2958. [PubMed]

[11] . Kim JH, Han E. Endometriosis and Female Pelvic Pain. Semin Reprod Med. 2018 Mar;36(2):143-151. [PubMed]

[12] . Platteeuw L, D'Hooghe T. Novel agents for the medical treatment of endometriosis. Curr Opin Obstet Gynecol. 2014 Aug;26(4):243-52. [PubMed]