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Adenomyosis Externa, An Unique Uterine Wall Endometrioma – A Rare Case Report

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Abstract :
Endometriosis is a chronic inflammatory disease defined as the presence of endometrium-like tissue outside the uterine cavity. Endometriomas are cystic lesions that occur from the disease process of endometriosis most commonly found in the ovaries. Isolated Uterine endometrioma has never been reported so far. Here we present an unique case of endometrioma, arising from serosal surface of uterus. This is the first case ever reported with the 17cm isolated endometrioma arising from the posterior wall of the uterus.

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Introduction

Endometriosis is a chronic inflammatory disease defined as the presence of endometrium-like tissue outside the uterine cavity. The endometriotic tissue is estrogen-dependent, hence it is found commonly in women during their reproductive age group, but the symptoms of endometriosis and its management can extend beyond menopause (2). The exact incidence of endometriosis is unknown; however, it is said to range from 2 to 10% of female population but up to 50% in infertile women and 2 to 4% postmenopausal (2). Symptomatic individuals present with dysmenorrhea, dyspareunia and infertility which affects not only the patients but their partners as well.

Endometriotic lesions can be classified as ovarian, extraovarian, or mixed. They are predominantly located in the ovaries (96.4%), the soft-tissue (2.8%), gastrointestinal tract (0.3%), and urinary tract (0.2%)(4). Endometriotic lesions which are located outside the pelvis is said to occur due to dissemination of endometrial cells or tissue through lymphatics and blood vessels. This includes the gastrointestinal tract, urinary tract, soft tissues, chest and incisional sites (2). Diagnosis and management of this disease is extremely difficult and it is both emotionally and physically crippling for the patient and the treating physician.

Endometriomas are cystic lesions that occur from the disease process of endometriosis most commonly found in the ovaries. Isolated Uterine endometrioma has never been reported so far. Here we present an unique case of endometrioma, arising from serosal surface of uterus. This is the first case ever reported with the 17cm endometrioma arising from the posterior wall of the uterus.

Case :

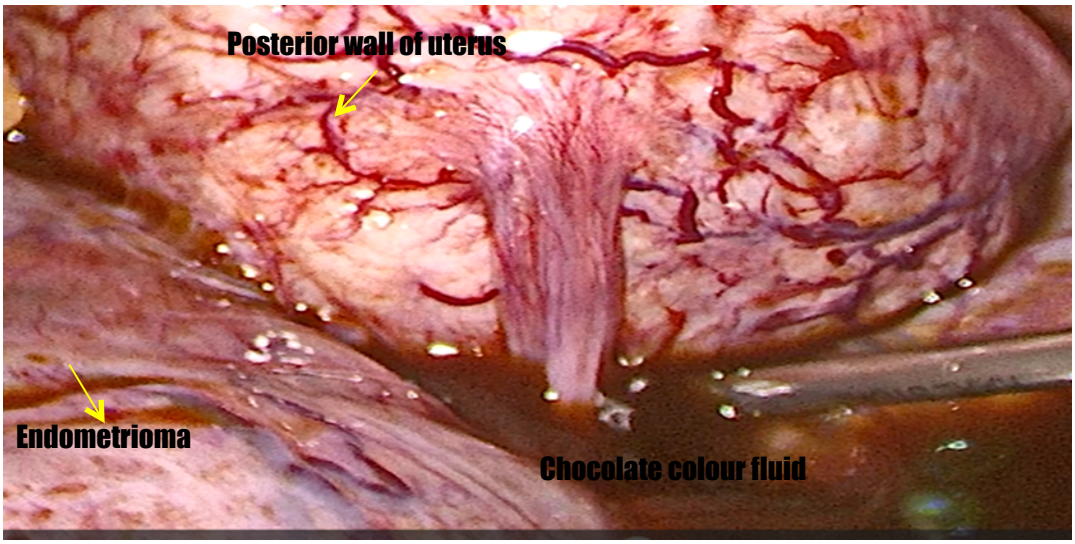
A 39 years old single, nulligravid was referred to the hospital with the diagnosis of large ovarian endometrioma. She presented with complaints of lower abdominal pain specifically more on the left side for two months. She gave history of heavy menstrual bleeding for two years associated with severe congestive dysmenorrhea for the past six months and her menstrual cycles were irregular. She was evaluated with ultrasound and MRI pelvis and was diagnosed as complex ovarian cyst most likely endometrioma arising from the left ovary of about 17 cm, with an elevated CA - 125 of 254. MRI pelvis revealed bulky uterus, with posterior myometrium diffusely heterogenous suggestive of diffuse adenomyosis mostly involving the posterior myometrium. MRI also revealed a large hyperintense cystic mass of size 17 x 15 x 16 cm seen in midline in the pelvis, extending superiorly into the abdominal cavity displacing the bowel loops. A deep solid soft tissue nodule of size 2.7x1.5cm was seen within the lesion which was not showing any evidence of diffuse restriction or contrast enhancement. Left ovary was not seen separately - findings were suggestive of large left ovarian

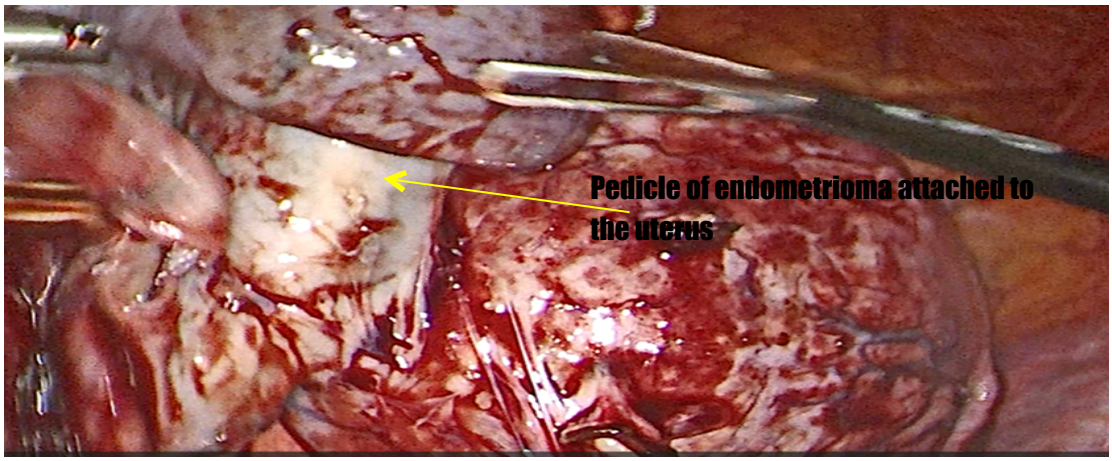
endometrioma. She was referred to us in view of large left adnexal cyst (endometrioma) for further management.

On examination, her vitals were stable and abdominal examination revealed an infraumbilical mass of size corresponding to 18 weeks, more towards the left side with tenderness and absent guarding or rigidity. The most likely diagnosis of endometrioma and possibility of malignancy in view of large size was explained to the patient. She was advised to undergo laparoscopic removal of lesion with possible staging laparotomy.

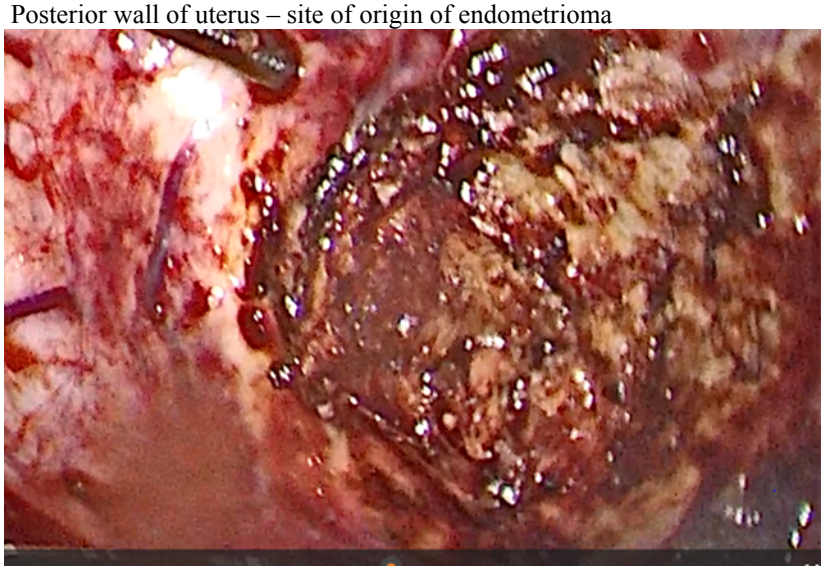
Patient underwent laparoscopy excision of lesion, intraoperatively we found a large thick walled cystic lesion measuring in size of 17x12x15 cm , adherent to the omentum on most of its surfaces and the bowel ,the origin of the lesion being obscure due to the large size and adhesions . Hence the contents of the cyst (chocolate colored fluid) was aspirated and after shrinking of the lesion ,proceeded with adhesiolysis to separate the lesion from the bowel and omentum . The origin of the lesion was identified to be the posterior wall of the uterus with a wide based pedicle of size 2.5 to 3cm. Uterus appeared bulky with diffuse adenomyosis, highly vascular, the endometrioma did not have any connection to the tubes or ovary. The right tube and ovary were normal , left tube and ovary were obscured by the adhesions, post adhesiolysis left ovary and tube visualized and found to be normal . There was no other endometriotic deposits in the pelvis. The endometrioma was removed in toto by coagulation and cutting the pedicle and the cyst retrieved through endo-bag.

Intraoperative images of posterior wall endometrioma



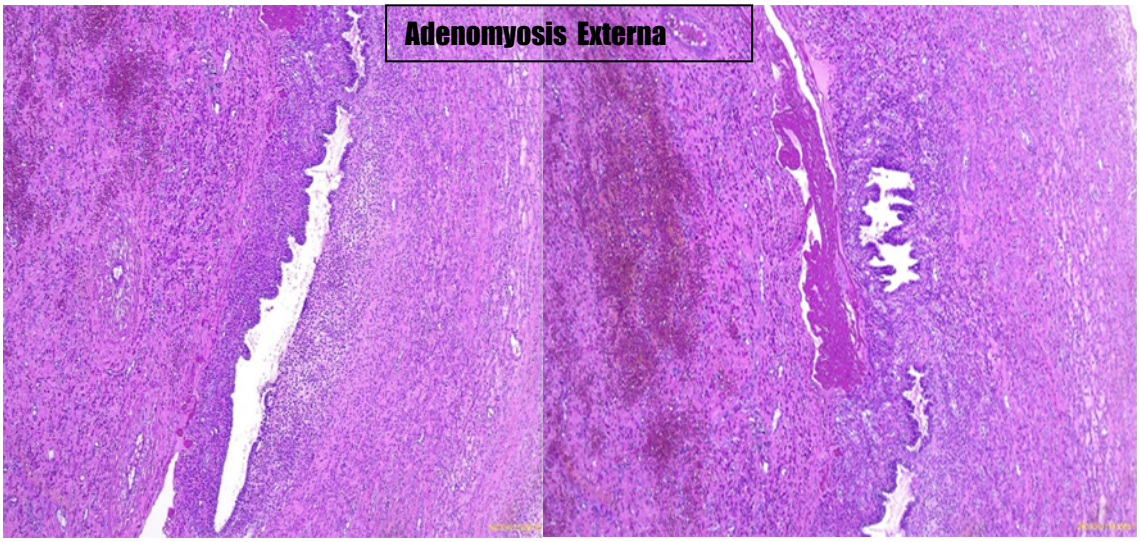


Posterior wall of uterus – site of origin of endometrioma



Histopathology

Histopathology findings reveals cystic areas lined by endometrial lining cells with few glands at places , underlying stromal cells and hemosiderophages in some areas suggestive of adenomyotic cyst thus confirming the diagnosis of endometrioma.



Post operatively she was advised for prolonged medical management and offered GnRH analogues and dinogest. She opted for dinogest only and is currently on regular follow up and is responding well to medical management .

Discussion:

Endometriosis is a chronic inflammatory disease which is caused by the presence of ectopic endometrial tissue which react to changes in the ovarian steroid hormones such as oestrogen and progesterone as expressed by proliferation, differentiation, and bleeding(2). Endometriomas are the most common form of endometriosis on the ovary, 17 to 44% of women diagnosed with endometriosis will have an ultrasound finding of endometrioma(5). These lesions are known as chocolate cysts due to the presence of thick dark brown fluid contained in them. Endometriomas are said to be caused when the seeding of ectopic endometrial tissue occurs, most often on the ovary, which bleeds causing a hematoma(5).

The risk factors leading to endometriosis could be nulliparity, early menarche (typically before 11 to 13 years old), late menopause, short menstrual cycles (less than 27 days), heavy menstrual bleeding, Mullerian anomalies, height greater than 68 inches, low body mass index (BMI), consumption of high amounts of trans

unsaturated fat, exposure to diethylstilbestrol in utero(5) . The etiology of endometriosis is unknown, still under research . The well known and accepted theory for developing endometriosis is retrograde menstruation. This theory says that endometriosis develops from the endometrial tissue travelling in a retrograde manner through the fallopian tubes and into the pelvis during a woman's natural menstrual cycle. This tissue then travels and gets imbibed in different areas, creating the endometriotic lesions. Some of these lesions may seed in an ovary and begins to form an endometrioma. Other two theories that have been suggested include the theory of metaplasia, in this theory it is said that extrauterine cells undergo metaplasia and gets differentiated into endometrial cells. Yet another significant theory is that viable endometrial cells get seeded via the hematogenous and lymphatic spread. But none of the theory is proven (4).

Endometriosis can take one of the three forms, depending on the clinical presentation and management: peritoneal or superficial endometriosis, ovarian endometrioma (OMA), or deep infiltrating endometriosis (DIE)(5). DIE is the most aggressive form, which affects 20% of women who suffer from endometriosis . DIE is defined as the presence of endometriotic lesions over 5 mm in depth under the peritoneal surface; others define it as a pathologic entity, which is called "adenomyosis externa". The 5 mm definition allows the understanding of lightly deeper classic lesions (type I). It would be more suitable to define DIE as adenomyosis externa with unique lesions (infrequently two or three) that are large (mainly >1 cm in diameter), and are reported as type II and type III lesions. Deep infiltrating endometriosis (DIE) is the most aggressive of the three types that constitute endometriosis (8). DIE is characterized by higher expression of invasive mechanisms (caused by matrix metalloproteinases and activins) and of neuro angiogenesis genes (nerve growth factor, vascular endothelial growth factor) compared with superficial and ovarian endometriosis. Due to various complexities of the disease it makes it more challenging for the clinician and the patient. It can affect the pelvic organs , affecting the anatomy and functions of vital organs, affecting the patient's quality of life. In the absence of other types of endometriosis, the isolated presence of DIE was only observed in 6.5% of cases. Although it may be considered a separate entity, they all may share similar pathogenic pathways (8). The diagnosis of DIE is based on clinical examination supported by ultrasound and MRI. The typical appearance of these lesions on ultrasound shows low-level homogenous echoes, otherwise described as a ground-glass appearance. This is consistent with old hemorrhagic debris. These lesions are also typically devoid of any vascularity when examined with doppler flow. MRI has shown a higher sensitivity for detecting a pelvic mass than ultrasonography(7). Like ultrasound, MRI also has limitations in detecting diffuse pelvic endometriosis. Therapeutic options include medical and surgical treatment, and the decision should be dictated by the patient's medical history, disease stage, symptom severity, and personal choice. Medical therapy can control the symptoms and stop the development of pathology, keeping in mind the side effects derived from a long-term treatment and the risk of recurrence once suspended. Surgical treatment should be proposed only when it is strictly necessary (failed hormone therapy, contraindications to hormone treatment, severity of symptoms, infertility, size of lesion)(7), preferring whenever possible, a conservative approach performed by a multidisciplinary team. All therapeutic possibilities have to be explained by the physicians in order to help the patients to make the right choice and minimize the impact of the disease on their lives.

Conclusion:

Isolated uterine endometrioma is extremely rare to find with no attachment to the ovary or to the tubes, there is no literature till date which has been found with such presentation. This was diagnosed only intraoperatively, again emphasizing the importance of laparoscopy in diagnosing and assessing the extent of lesions. As a clinician having known the difficulty of enucleating a 17 cm endometriotic cyst without compromising the ovarian reserve, this particular case was a pleasant intraoperative surprise as we could provide complete removal of the large endometriotic cyst without compromising the ovarian reserve . With continued medical management the patient is currently symptom free. If medical management fails, thorough surgical removal of the endometriotic cyst whatever may be the location of the endometrioma by an experienced laparoscopic surgeon followed by medical management is warranted. Counselling about the prolonged management with regular follow up in an endometriosis clinic is mandatory. Laparoscopy is no wonder , still the gold standard for diagnosing, evaluating and managing endometriosis .

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