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Case Report

Endometriosis externa: A rare presentation

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ABSTRACT

Endometriosis externa is presence of endometrial glands and stroma outside uterus. Umbilical endometriosis (also called Villar's nodules) and subcutaneous endometriosis are rare conditions that occur in elderly women without any menstrual complaints and pelvic endometriosis. The overall incidence of umbilical endometriosis is very low (0.1-0.5%). Here we report a rare case of endometriosis externa with umbilical menstruation in a woman with a previous history of cesarean section and myomectomy. After complete evaluation, the patient underwent excision of the endometriotic lesion with abdominoplasty with mesh repair. The possible cause of this type of endometriosis can be secondary type due to direct implantation of endometrial tissue in the subcutaneous tissue during the previous surgeries.

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1. Introduction

Endometriosis is a condition that can be endometriosis interna or externa. If the endometrial glands and stroma present in the myometrium are called endometriosis interna or adenomyosis. The presence of the same outside uterus is called endometriosis externa. Endometriosis externa is commonly present in ovaries, Pouch of Douglas but it can be seen anywhere outside the pelvis like gastrointestinal tract, ureters, lungs, and previous surgical scar.¹ Often, the women have complaints related to menstruation or fertility like dysmenorrhea, dyspareunia, menorrhagia, and infertility.¹

Umbilical endometriosis is also called Villar's nodules described in 1886.² Endometriosis could be suspected if a patient presented with umbilical swelling or any other swelling with cyclical pain or bleeding from the swelling in correspondence with the menstrual cycles. The suspicion should increase if there is a history of any surgery related to the uterus like cesarean section, myomectomy,

or laparoscopic surgery for pelvic endometriosis for the possibility of secondary implantation.^{1,3,4} So, the umbilical endometriosis can be primary or secondary.¹ Primary umbilical endometriosis, could be due to migration of endometrial tissue via the abdominal cavity or lymphatic system or the embryonic remnant in the umbilical fold such as umbilical vessel and urachus to the umbilicus. Secondary umbilical endometriosis can be due to implantation of endometrial tissue iatrogenically during the surgery.³

2. Case Report

A 41 years old woman came with complaints of swelling in the left side of the abdominal wall for 2 years associated with cyclical pain during the menstrual cycle, the pain is intermittent associated with a mild increase in the size of the swelling observed only during menstruation.

The patient had a similar swelling in the umbilicus for the past 2years and observed a minimal amount of bleeding through the umbilicus in the previous cycle which was stopped spontaneously after the menstruation. There was no associated pain or increase in the size of the umbilical

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lesion.

Her menstrual cycles were regular, 3-5/30 days, with moderate flow and it was not associated with clots or any dysmenorrhea. She had a history of one cesarean delivery 12 years back and a history of open myomectomy surgery 3 years back. There was no significant medical history. There was no significant family history of a similar condition.

On examination, the patient was moderately obese. Abdominal examination revealed a healthy midline vertical scar just below the umbilicus to just above the pubic symphysis noted. The umbilicus was replaced by a bluish, irregular, non-tender nodular lesion of size 3cm, which was firm to hard in consistency. A swelling in the abdominal wall of size 5x5cm, 2-3cm below and left lateral to the umbilicus noted, which was tender, firm with ill-defined borders and the overlying skin shows reticular vascularity (Figure 1). External genitalia healthy, on bimanual examination uterus normal sized and anteverted, mobile with no forniceal tenderness or mass.

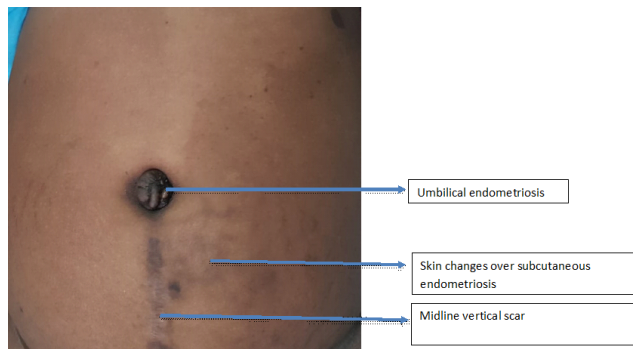


Fig. 1:

On evaluation, the patient was diagnosed as hypertensive. Hematological and biochemical examinations are within normal limits. Pap smear was done, which was negative for CIN or malignancy. Magnetic resonance imaging of abdomen and pelvis suggestive of the anteverted bulky uterus with few intramural and subserosal fibroids of uterine walls, largest measuring 2.9x2.7cm with mild focal adhesion of anterior abdominal wall and uterine wall. A well-defined, 5.6x4.3cm T2 weighed hyperintense lesion with the hypointense area, T1 hyperintense area, and surrounding fat strand extending over the muscular and deep subcutaneous plane of infraumbilical region of anterior abdominal to the left side. A similar appearing lesion of size 3.1x2.6cm in the umbilical region was present. A small abdominal wall defect of 5-6mm with herniation of omentum present.

Based on the imaging report and the chances of recurrence of the same condition, the patient was given the option of total abdominal hysterectomy with bilateral salpingo-oophorectomy along with endometrial lesion excision, excision of umbilical endometriosis,

and abdominoplasty. Intraoperatively, 4x3cm umbilical endometriotic mass was noted. There was no herniation of omental contents but omentum was adherent with the umbilicus. 5x5cm dense fibrotic endometriotic mass noted in the abdominal wall above the rectus sheath and adjacent to the left side of the umbilicus. Minimal adhesion noted between the anterior abdominal and the uterus. The uterus was irregularly enlarged to 8 weeks size due to multiple fibroids, the right ovary was healthy and the left ovary showed a cyst of size 4x4cm. There was no evidence of pelvic endometriosis. The umbilical mass was removed along with the umbilicus (Figure 2). Abdominal wall lesion above the rectus sheath was removed from the subcutaneous plane which was situated away from the suture line (Figures 3 and 4). Total abdominal hysterectomy with bilateral salpingo-oophorectomy done (Figure 4). To prevent the incidence of incisional hernia, abdominoplasty was done with mesh repair (Figure 5). The excised lesion and the specimens were sent for histopathological examination.

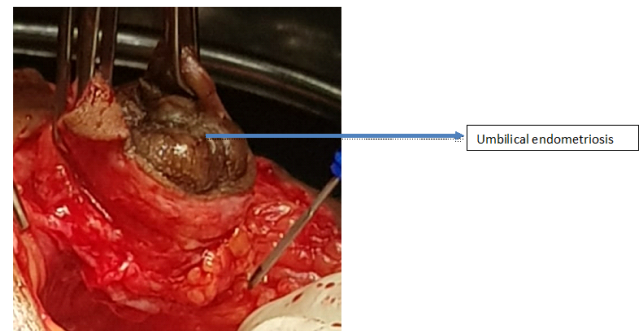


Fig. 2:

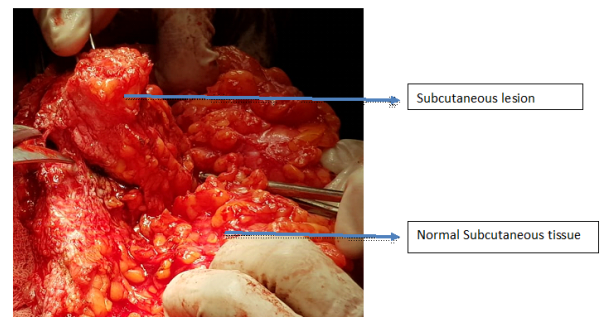


Fig. 3:

Cut section of the uterus – multiple intramural fibroids, largest size 3x3cm. endometrium and cervix appeared normal. No growth or polyp. Umbilical lesion- shows yellowish deposits. The abdominal wall lesion showed reddish-brown deposits. Histopathological examination of the specimens showed the presence of endometrial glands and stroma.

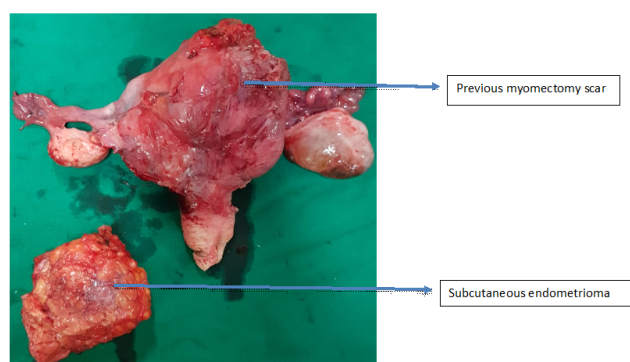


Fig. 4:

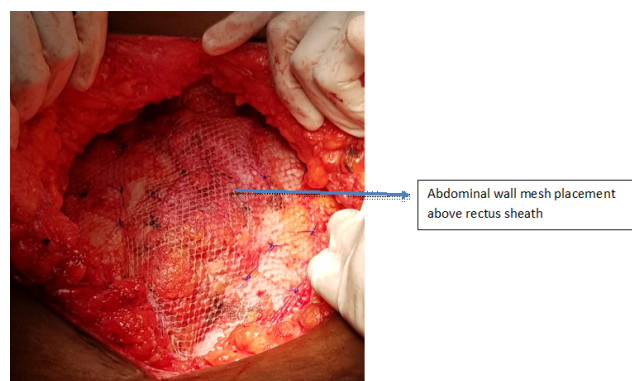


Fig. 5:

3. Discussion

The incidence of Endometriosis in a reproductive woman is about 6-10%.⁵ It can be primary or secondary type. Primary Endometriosis has been suggested by various theories. The embryonal rest theory, the coelomic metaplasia theory, and the migratory pathogenesis theory. It states that under the influence of inflammation or trauma, the coelomic pluripotent mesothelium cells in the skin dedifferentiate into endometrium or by direct extension, lymphatic and vascular dispersion (main) or surgical manipulation of endometrial tissue. Also, the endometrial cell proliferation along the urachus suggested theory. But the exact mechanism was unknown.^{1,3-5} Secondary endometriosis, the widely accepted theory, iatrogenic implantation of endometrial cells as a result of surgery, commonly laparoscopic procedures or implantation of an endometriotic cell by direct contact.^{4,6} Uterine endometrial fragment deposition in the skin is an uncommon but well-recognized phenomenon. Differential diagnosis of umbilical nodules should include pyogenic granuloma, hernia, residual embryonic tissue, primary or metastatic adenocarcinoma (Sister Joseph's nodule), nodular melanoma, and cutaneous Endosalpingosis.^{1,4,5} Malignant transformation of umbilical endometriosis into endometrial carcinoma has also been reported.^{2,3}

The diagnosis was based on the symptoms correlating with the menstrual cycle along with USG and MRI imaging shows hyperintense on T1 weighted imaging is difficult sometimes.³ But the definitive diagnosis is based on the histopathological examination considered the gold standard and also to exclude malignancy.³ If malignancy is detected, the more common is clear cell carcinoma followed by endometrioid carcinoma, but it has a poor prognosis.⁶

The association of subfertility or infertility with endometriosis is controversial. About 44% of patients with moderate to severe endometriosis, conceived normally.² The treatment of choice for endometriosis is wide local excision.¹⁻³ Hormonal therapy can also be used to reduce the size of the lesion and to reduce the symptoms of associated pelvic endometriosis. The drugs used are danazol, combined oral contraception pills, and gonadotropins releasing hormones agonist or antagonist.^{3,4}

In our case, since the patient was 41 years, not planning for pregnancy as well as the uterus was enlarged with multiple fibroids and history of myomectomy pointed towards the future need for hysterectomy. So, to avoid recurrence of disease and the future need of surgery, the patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy with excision of umbilicus and the subcutaneous lesion. Because of the midline incision and the obesity, the chance of incisional hernia is high. So, the abdominal wall is reinforced with nonabsorbable prolene mesh (on-lay mesh over the rectus sheath) repair has been done.

4. Conclusion

To conclude, the most common cause of endometriosis outside pelvic cavity is due to secondary implantation. Due to the increased rate of caesarean section in obstetrics and advancement of surgical techniques in gynaecology like diagnostic and operative minimal invasive surgery may contribute to rising incidence of endometriosis externa. As a preventive measure, surgeons should have an utmost care to avoid secondary implantation during surgeries like caesarean section, myomectomy by minimally handling endometrial tissues.

5. Source of Funding

None.

6. Conflicts of Interest

The authors declare no conflicts of interest.

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