

# Abdominal Wall Endometriosis – Case Series

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## Abstract:

*Endometriosis is primarily a disease of pelvis. Extra pelvic endometriosis is defined, when eutrophic endometrial tissue is detected in any site other than pelvic cavity with the specification that this invasion is benign (non-neoplastic) in nature. Endometriotic lesions are usually found on the peritoneal surfaces of the reproductive organs and adjacent structures of the pelvis, but it can occur anywhere in the body. Extra pelvic endometriosis is a rare condition and it's most common extra-pelvic location is the abdominal wall, occurring most frequently after cesarean delivery or any other abdominal surgery. Other sites are gastrointestinal tract, respiratory tract, pleura, pericardium, diaphragm and primary umbilical endometriosis. Patient's usually present with painful mass in the abdominal wall during menstrual period, sometimes with a typical presentation, but recently incidences of scar endometriosis is increased. Two cases are presented here. To obtain a definitive diagnosis and optimal treatment for abdominal wall endometriosis, complete excision & histopathological examination are necessary. Here we presents 3 cases with umbilical endometriosis who presented between 13 to 28 years of age. Our all cases presented with a cyclical painful, abdominal mass and sometimes bleeding from umbilical mass. Also we highlight the importance of early recognition and appropriate surgical intervention to minimize morbidity and mortality from endometriosis.*

## Introduction:

Endometriosis is defined as the presence of eutrophic endometrial tissue outside the normal uterine cavity and it is a chronic, benign, estrogen dependent inflammatory disease. True prevalence is unknown due to lack of formal epidemiological study but approximately 10% women of reproductive age suffer from this disease. Regarding the prevalence of disease, Practice Bulletin No. 114 published in 2010<sup>1</sup> and Brugg mann D et al in 2016<sup>2</sup> mentioned that 2% to 50% women are thought to have silent endometriosis. They also mentioned that 40% to 60% women suffer from severe dysmenorrhea due to endometriosis and 20% to 30% women are sub-fertile due to endometriosis.

So, endometriosis is considered to be a common condition. It has different clinical manifestations, and there is debate on its diagnostic and therapeutic aspects. Extra pelvic endometriosis has been reported in any region of the body including bowel, bladder, lung, kidney, extremities, perineum and umbilicus. Less than 12% of reported cases of endometriosis are refers to implants found else anywhere in the body. Extra-pelvic endometriosis may be found in the surgical scar area and subcutaneous tissue after obstetric and gynecologic surgery. Endometriosis may be seen after the interventions that contain endometrial tissue such as caesarean section, episiotomy for a vaginal birth, hysterectomy and operation for ectopic pregnancy, or in laparoscopic port<sup>3,4,5</sup>. Characteristic presentation is

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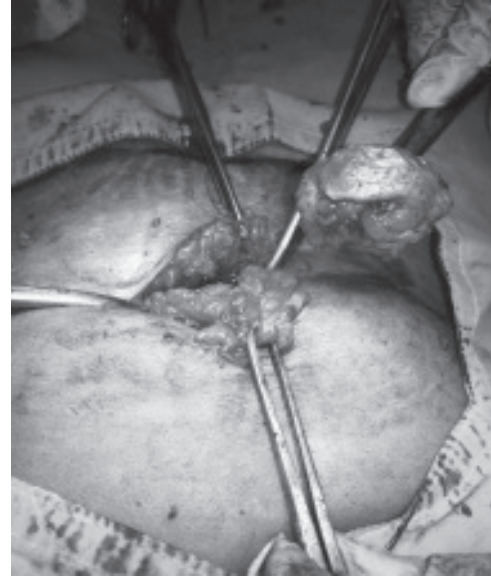
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of a painful, palpable mass with cyclic bleeding or discharge with worsening of pain and bleeding during menstruation. The diagnosis of this entity is neither easy nor routine. Many diagnostic methods both clinical and laboratory have been used, but none of them is of gold standard. Regarding diagnosis MRI and ultrasound guided fine needle aspiration cytology provide more definitive diagnosis.<sup>6</sup> Treatment option includes medical and surgical management. We reported here our experience of surgical and medical management of 5 cases of abdominal wall endometriosis. Also some literature review has been presented here.

### Case presentation:

#### Case: 1

A 28 years old woman presented with a painful swelling adjacent to caesarean scar and she gives history of gradual increase in size of the mass at the time of menstruation. She had a caesarean delivery 2yrs back followed by lactational amenorrhoea for 11 months, there after her regular menstrual cycle was established. The couple did not practiced any contraceptives. Then she noticed gradual development of a swelling beneath the caesarean scar. On palpation a firm tender mass measuring about 4 X 3 cm subcutaneously was found. Review of symptoms did not reveal any history of pelvic endometriosis and pelvic examination reveals no abnormality. A subcutaneous soft tissue hypo echoic lesion 4 X 3cm was detected by ultrasonography



**Case 1:**

imaging. The lesion was excised with the preliminary diagnosis of endometriosis. The histopathological examination confirmed the diagnosis of endometriosis. No recurrence was seen in 3 years of follow-up period after surgery.

#### Case: 2

A 23 year-old-lady, P-2, both caesarean delivery, house wife of middle class family complains of cyclical pain with nodular swelling within the umbilicus which gradually increased in size for last 4yrs. The umbilical mass used to increase in size and become painful during her menstrual period. Thereafter when she



Before surgery



After surgery

**Case 2:**

became pregnant the mass became pain free. After 8 months of her delivery when her regular menstrual cycle was established, she felt similar type of discomfort like before. Size of nodule and intensity of pain gradually increased. Cyclical bleeding occurred from umbilicus for the first 2 days of menstruation. Her general condition reveals normal. On P/A examination a soft, cystic, dark colored tender mass measuring about 0.5 x 0.5 cm within umbilicus was found. Pelvic examination reveals no abnormality. So from history and clinical examination it was diagnosed as a case of umbilical endometriosis. She took OCP without dummy pill for 6 months but not cured the excision of endometriotic nodule from the umbilicus under G/A was done. No recurrence was found after 5 years follow up.

### Case: 3

A 28 year-old-lady, Para 2 (NVD)+1(MR) house wife of middle class family complains of cyclical pain and nodular swelling within umbilicus for last one year. She was regularly menstruating woman without pain with average flow and duration and was reasonably well 1 year back. Then she noticed cyclical pain with nodular swelling within the umbilicus. She also

noticed that the pain was gradually increasing and at the same time the size of mass was increasing. Initially her pain would persist for first 2 to 3 days of menstruation. Her general condition reveals normal. A soft, cystic, dark-colored tender mass measuring about 1 X 0.5 cm was found inside the umbilicus. Pelvic examination reveals no abnormality. Patient took OCP without dummy pill for 3 months and then came for follow up, when the size of the nodule was found to be decreased. Then she was advised to continue OCP without dummy pill for next 6 months. After completion of conservative treatment, patient was symptom free.

### Case: 4

A 13 year-old girl complain of cyclical per umbilical bleeding with pain and swelling for 1 year. With this complain she consulted with a surgeon and excision of nodule done but after 3 months her bleeding and pain from the umbilical scar recurred. So she consulted with a gynecologist and was advised for continuous uninterrupted estrogen-progesterone oral pill taking without dummy pill. Bleeding was stopped after 3 months of OCP taking and it continued up to 9 months. She is now symptom free and under observation.



Before OCP



After OCP

Case 3:

**Case 4:****Case – 5:**

A 25 year-old lady had a history of laparoscopic right sided salpingo-oophorectomy due to large chocolate cyst done in 2014. During last 1 year she felt pain in left lower port with swelling which was related to menstruation. Patient was under Dinogest treatment and now she is symptom free.

**Case 5:****Discussion:**

Endometriosis is a benign disorder, which affects 6% of all women in the reproductive age<sup>7</sup> Endometriosis outside the pelvic cavity is rare, but its most common extra pelvic location is in the abdominal wall, occurring most frequently after caesarean delivery. Pelvic endometriosis in many cases is crippling for affected women. Symptoms of endometriosis like dysmenorrhoea, dyspareunia, pelvic pain, pain during defecation and infertility are common<sup>8,9</sup> ; but these aren't useful for localization of endometriotic lesions.

So it is very important that endometriosis can be diagnosed as soon as possible and with certainly, because this is the necessity of highest therapeutic efficacy. In this regard fundamental detection of all sites involved by this pathology, superficial, deep, pelvic and extra pelvic sites are particular interest. The endometriosis localization at abdominal wall is challenging. They appear like a mass of respectable size near a surgical scar, like caesarean, hysterectomy, appendectomy, a scar in the laparoscopic trocar passage way, even in the puncture sites of amniocentesis and in the episiotomy scars.<sup>10,11</sup> There are various theories about the origin of endometriosis. One explanation is that there is a problem with interactions between hormone and immune system. Our immune system usually makes defense that tissue from a particular organ does not grow elsewhere in the body. It is still not clear why endometrial tissue sometimes grows outside the womb.

Abdominal wall is the most frequent location of extra pelvic endometriosis. Endometriosis of the abdominal wall is usually associated with a surgical procedure in the uterus especially in women who have delivered by cesarean section. The most common clinical symptom in women with abdominal wall endometriosis is a constant focal abdominal pain which is mostly not associated with the menstrual cycle<sup>12</sup>. This pain is frequently atypical so it can be misdiagnosed. Patients can also feel a palpable mass in the area of the surgical section.

Diagnostic methods for an abdominal wall endometrioma are: ultrasonography, computer tomography (CT) and magnetic resonance tomography (MRI). Ultrasonography is not the gold standard diagnostic method for an abdominal wall endometrioma as it shows a mass in the abdominal wall. This mass is imaged as a solid, hypo-echoic lesion containing internal vascularity and it may also contain cystic acne. Abdominal wall mass must be submitted to differential diagnosis. Differential diagnosis includes neoplasms like sarcoma or lymphoma, suture granuloma, ventral hernia, abscess or haematoma, melanocytic nevus, seborrheic keratosis, epithelial inclusion cyst, desmoids, haemangioma and granulosa cell tumor.<sup>10</sup> The latter three can be excluded from the final diagnosis of endometrioma by ultrasonography, CT and MRI, as these tumors show a solid mass in the

abdominal wall. So these cannot be gold standard diagnostic tools of an endometrioma, but can depict the extent of the disease preoperatively<sup>13,14</sup>.

Primary umbilical endometriosis: More common form of umbilical endometriosis, develops spontaneously in the absence of any previous abdominal surgery. The umbilicus could be an extraordinary site of endometriosis and is affected in 0.5-1% of the women with endometriosis and up to 40% of patients with extra pelvic endometriosis. Symptoms occurring an average of  $17.8 \pm 3.9$  months before presentation<sup>15, 16</sup>. Umbilical endometriosis could have a secondary nature, but this disorder can also be spontaneous or primary. The exact pathogenesis is unknown, however possible theories include the spread of endometrial cells to the umbilicus through the abdominal cavity, via the lymphatic system or through the embryonic remnants in the umbilical folds<sup>17,18</sup>. Secondary umbilical endometriosis develop following previous surgery (caesarean section, abdominal hysterectomy, appendicectomy, laparoscopy etc.) and is due to iatrogenic seeding of endometrial tissue. Primary umbilical endometriosis is typically manifested by a firm, pigmented, or bluish nodule with pain and tenderness associated with cyclical bleeding or discharge during menstruation. Here the subcutaneous nodule presented by the patients did not show any skin symptoms, pigmentation, or bleeding in the umbilicus. It was difficult to establish a definitive diagnosis of umbilical endometriosis by only the USG or MRI findings. Zohai also stated that the FNB (fine needle biopsy) is a useful additional tool for diagnosing cutaneous endometriosis<sup>17, 18</sup>. Although there are no reports on the diagnostic accuracy of FNB in umbilical endometriosis, the clinical features include an umbilical swelling (90%), often associated with cyclical pain and bleeding (49.2%)<sup>19</sup>. Investigations may include an ultrasound to assess the echogenicity and vascular involvement. MRI can also be helpful, although no imaging modality is diagnostic. Medical treatment such as the Estrogen progesterone containing Oral Contraceptive Pill (OCP) or GnRH analogues is an option and can be effective in relieving symptoms temporarily.<sup>20</sup> However after cessation of hormonal treatment symptom recurrence is common. The definitive management involves surgery, which can vary from superficial

diathermy to the more radical omphalectomy with a concomitant laparoscopy approach. This allows for total excision of the umbilicus followed by repair of the underlying fascia and reconstruction of the umbilicus. Another option is local resection of the endometriotic tissue and sparing of the umbilicus. Whilst there is no real data on follow up of these surgically treated cases, Fedele et al.<sup>21</sup> described in their case series recurrence of umbilical endometriosis in those managed with superficial surgical approach.

Theoretically, drug treatment would be ideal in the treatment of endometriosis. In practice, however, drug therapy alone is accompanied with a temporary improvement of pain and tampering of the symptoms but in a time period they usually return. Also, drug therapy can certainly reduce the size of endometriomas and facilitate their surgical removal. Contraceptive pills are one of the main drug treatments, causing reduction of the quantity of menstrual blood loss and in that way they result in reducing pain during menstrual period. Additionally, Dinoprogesteron, Gonadotropin-releasing hormone (GnRH) agonists block the production of ovarian-stimulating hormones (FSH and LH), lowering estrogen levels and preventing menstruation. This causes endometrial tissue to shrink. GnRH agonists can force endometriosis into remission during the time of treatment and sometimes for months or years afterwards the effectiveness of drug therapy on the reproductive capacity is questionable. That is the reason why drug therapy is not recommended as the sole treatment of women with endometriosis, except for rare cases where surgery is not possible or presents a significant risk for the life of the patient<sup>22,23</sup>.

Here we had presented 3 cases of umbilical Endometriosis. First patient needed surgery, 2<sup>nd</sup> one had only 9 months OCP treatment helped to complete regression and 3<sup>rd</sup> case was recurrent umbilical Endometriosis after surgery and she was also treated by OCP. But simple OCP treatment would have a satisfactory result; none of them has any history of pelvic Endometriosis.

#### **Conclusion:**

There are 2 steps in the identification of extra pelvic abdominal wall endometriosis; analysis of symptoms that may be attributed to the presence of endometriosis and the macroscopic with

microscopic evaluation of lesions. Abdominal scar endometriosis is reported to be one of the rare types of endometriosis in the literature. However, it may be observed more common than it has been reported in the literature. A painful mass is found clinically, diagnosis depend on the clinical consideration a good medical history and physical examination. Radiological examinations are partially helpful for the diagnosis, but a definitive diagnosis is made by histopathological examination. Hormonal suppression therapy can be effective, once the diagnosis is made the management options depend on the severity of symptoms, location and extent of disease and the patients desire for future fertility. In surgical scar endometriosis, treatment modality to prevent recurrence of the disease is wide excision.

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