



# Umbilical Hernia as Forerunner of Primary Umbilical Endometriosis: A Case Report

## *Primer Umbilikal Endometriozisin Öncüsü Olan Bir Umbilikal Herni: Olgu Sunumu*

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### ABSTRACT

Umbilical endometriosis is a type of cutaneous endometriosis that usually follows laparoscopic or surgical procedures that involve the umbilicus. Primary umbilical endometriosis (PUE) is an extremely rare condition and its association with an umbilical hernia is an equally rare condition. To date, only very few cases of PUE with umbilical hernia association have been reported in the medical literature. Report herein is a case of PUE associated with an umbilical hernia who presented with classical umbilical nodule symptoms with cyclical pain and bleeding due to menstruation. The patient underwent omphalectomy with abdominal wall defect repair using prosthetic mesh. The diagnosis was confirmed by histopathological examination of the excised umbilical nodule. This case report highlights a rare entity that should be considered as a differential diagnosis in females of the reproductive age group that presents with the umbilical nodule.

**Keywords:** Primary umbilical endometriosis, umbilical hernia, omphalectomy

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### Öz

Umbilikal endometriozis, genellikle umbilikus içeren laparoskopik veya cerrahi prosedürleri takip eden bir kutanöz endometriozis türüdür. Primer umbilikal endometriozis (PUE) son derece nadir bir durumdur ve PUE ile umbilikal herni birlikteliği de aynı derecede nadir bir durumdur. Tıp literatüründe bugüne kadar umbilikal herni ile ilişkili çok az sayıda primer umbilikal endometriozis vakası bildirilmiştir. Biz burada, umbilikal nodülün klasik semptomları olan ve menstrüasyona bağlı döngüsel ağrı ve kanama ile başvuran umbilikal herni ile ilişkili bir PUE olgusunu sunuyoruz. Hastamıza, protez ağ kullanılarak karın duvarı defekt onarımı ile omfalektomi yapıldı. Eksize edilen umbilikal nodülün histopatolojik incelemesi ile tanı doğrulandı. Bu olgu sunumu umbilikal nodül ile başvuran üreme çağındaki kadınlarda ayırıcı tanı olarak düşünülmesi gereken nadir bir durumu vurgulamaktadır.

**Anahtar kelimeler:** Primer umbilikal endometriozis, umbilikal herni, omfalektomi

### INTRODUCTION

Endometriosis is defined as the presence of endometrium or endometrium-like tissue outside the endometrial cavity. Globally, up to 10% of women in the reproductive age group suffer from endometriosis<sup>1</sup>.

Umbilical endometriosis is a rare condition, with an estimated incidence of 0.5-1% of all endometriosis<sup>2</sup>. It generally develops following surgical procedures involving the umbilicus, whereas spontaneous umbilical endometriosis without preexisting pelvic endometriosis or

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abdominal surgeries is an extremely uncommon variant<sup>3</sup>. To date, only a few cases of umbilical endometriosis with umbilical hernia have been reported. Herein, reported is a case of spontaneous umbilical endometriosis associated with umbilical hernia, for which surgical excision and abdominal wall reconstruction using mesh was performed after obtaining the patient consent for the publication process.

### CASE REPORT

A 24-year-old female patient presented to the gynecology outpatient department with complaints of swelling over the umbilicus, which had gradually increased in size over the past 2 years and was associated with pain. She also noticed a slight increase in size and bleeding from the swelling during menses for the last 3 months. She had a history of abnormal uterine bleeding, for which she had received hormonal treatment. No history of significant dysmenorrhea, menorrhagia, dyspareunia, or subfertility was found. She had undergone suction and evacuation twice for first-trimester abortion. No uterine surgery or any other pelvic surgery, including laparoscopy, as well as endometriosis, and family history of autoimmune disease, malignancy, or any other gynecological disorders were found.

General examination revealed no abnormality, except for mild hirsutism. Local examination revealed a brownish-black nodule of approximately 2×1.5 cm on the umbilicus,

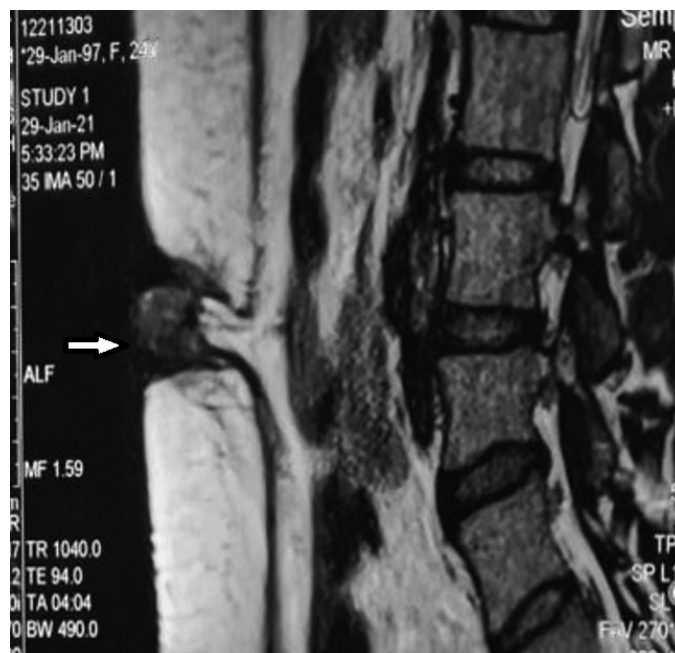


**Figure 1.** Macroscopic presentation of the 2×1.5 cm size and hyperpigmented firm lesion of umbilical endometriosis, marked by a white arrow.

which was tender and nonreducible, without any active bleeding (Figure 1). Gynecological examination revealed an anteverted uterus, which was normal in size, mobile, and nontender.

Ultrasound (US) showed a well-defined hypoechoic lesion of 2.3×1.2 cm with internal vascularity in the umbilicus, most likely an endometriotic implant, and the uterus was normal size with bilateral ovaries showing polycystic ovarian morphology (Figure 2). Magnetic resonance imaging (MRI) showed a hyperintense lesion at 1.9×2.0×1.6 cm with a bright signal in the umbilicus that suggests endometriosis and a tiny defect of 7×6.6 mm in the umbilicus with herniation of the omentum that suggest umbilical hernia. Fine needle aspiration cytology (FNAC) of the umbilical nodule revealed spindle-shaped cells with moderate cytoplasm with dense chromatin in the hemorrhagic background and possible benign spindle cell tumor or endometriosis.

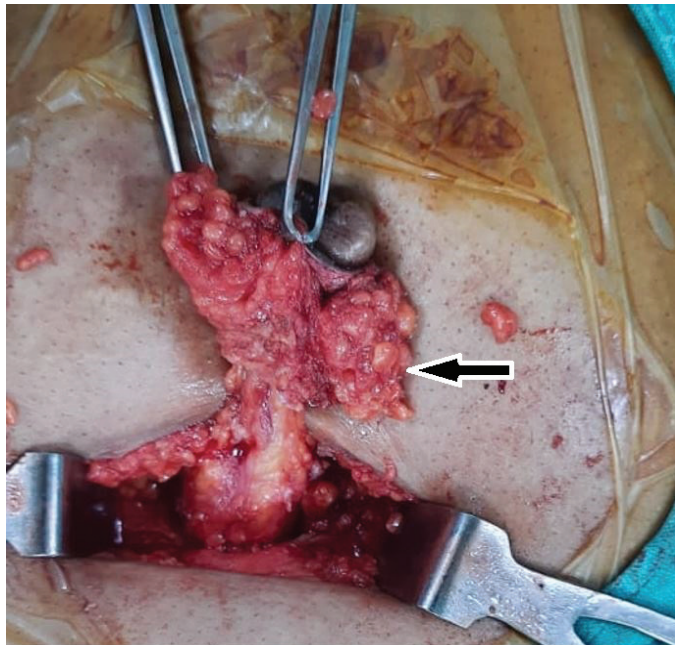
Based on clinical evaluation and investigation, a preoperative diagnosis of umbilical endometriosis with the umbilical hernia was made. Surgery was performed by a multidisciplinary team, involving gynecologists and a general surgeon without any delay after diagnosis. The elliptical skin incision was made with a wide margin of 0.5-1.0 cm and omphalectomy was performed and tissue



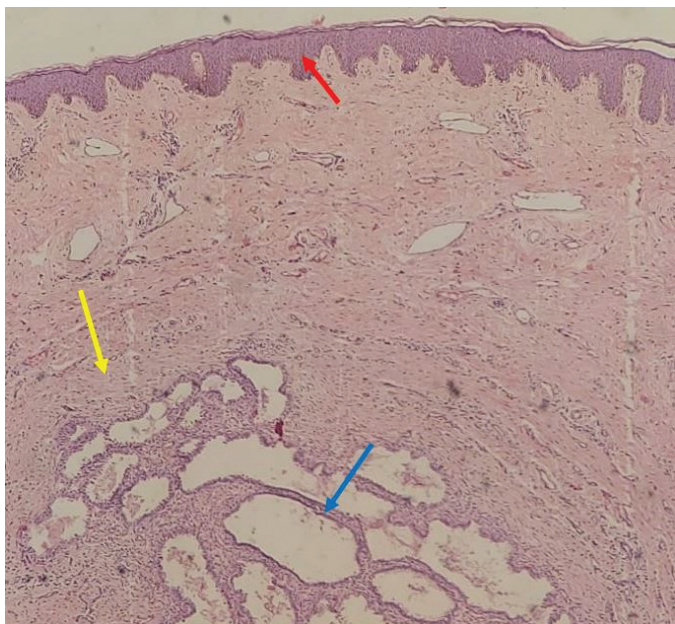
**Figure 2.** MRI showing primary umbilical hernia with umbilical nodule, marked by a white arrow.

MRI: Magnetic resonance imaging

was sent for histopathological examination (Figure 3). The hernia was repaired using polyester mesh. No finding of intra-abdominal endometriosis was detected in imaging or during surgery. Postoperatively, the patient had an uneventful course and was discharged on the fourth postoperative day. Histopathology confirmed the diagnosis of cutaneous endometriosis (Figure 4).



**Figure 3.** Intraoperative picture of location and excision of an umbilical nodule, marked by a black arrow.



**Figure 4.** H&E, 40X, section showing epidermis (red arrow), endometrial stroma (yellow arrow), and endometrial glands (blue arrow).

## DISCUSSION

Umbilical endometriosis is the most common site of primary cutaneous endometriosis. In 1886, Villar first described umbilical endometriosis, and hence, the condition is sometimes referred to as Villar's nodule<sup>4</sup>.

The pathogenesis of spontaneous endometriosis is not clearly understood. Among the various hypotheses, the most accepted one is the "hypothesis of migration or retrograde menstruation," in which the menstrual blood reflux and endometrial cells implant in the target organs. The second theory is "induction theory," or the theory of "coelomic metaplasia," where the mesothelium transforms into endometrium-like tissue under the influence of regurgitated endometrium<sup>5</sup>. This theory is based on the observation that pluripotent cells of the coelom differentiate into both endometrial and peritoneal cells<sup>6</sup>. In cases of cutaneous endometriosis, another possible mechanism could be a retrograde lymphatic flow with implantation of intra-abdominal endometrial cells into the subcutaneous tissue<sup>7</sup>.

In spontaneous umbilical endometriosis development, as in our case, the umbilicus possibly behaves as a physiological scar with a predilection for endometrial tissue or developed by metaplasia of urachal remnants stimulated by inflammation<sup>8</sup>. This may also represent intra-abdominal endometriosis within the hernia sac, which had herniated through the umbilical defect<sup>3</sup>.

However, our case did not have any features of intraperitoneal or pelvic endometriosis. Other possible mechanisms could be the migration of endometrial cells to the umbilicus through the abdominal cavity, the lymphatic system, and the umbilical vessels, which support the hematogenous or lymphatic spread theory.

Primary umbilical endometriosis (PUE) can present as single or multiple brownish or dark bluish and painful umbilical swelling of different sizes<sup>8-10</sup>, which can be asymptomatic or present with symptoms like pain, swelling, discharge, or bleeding during menstrual flow<sup>10</sup>. Our case had brownish swelling over the umbilicus with associated pain and bleeding during menstrual flow.

The umbilical nodule can be confused with various other conditions, such as subcutaneous abscess, cyst, desmoid tumor, lipoma, subcutaneous hematoma, lymphadenopathy, lymphoma, melanoma, soft tissue sarcoma, or metastatic tumors<sup>11</sup>. Umbilical endometrioma associated with an umbilical hernia is uncommon and can

remain unrecognized during the surgical intervention, which recurs after the surgery<sup>12</sup>.

A preoperative US scan can determine the cystic or solid components of the mass; however, this is not diagnostic of cutaneous endometriosis. Computed tomography scan or MRI shows the extent of the disease<sup>13</sup>. Fernandes et al.<sup>10</sup> advocated the use of FNAC for preoperative diagnosis based on cytological features of cutaneous and subcutaneous endometriosis related to cyclic hormonal changes. The cytological smears contain epithelial and stromal fragments admixed with hemorrhage and hemosiderin-laden macrophages. The gold standard diagnostic method is a histopathological examination of the excised mass<sup>10</sup>.

The most appropriate modality of treatment in symptomatic umbilical endometriosis associated with an umbilical hernia is total removal of the umbilicus with an adequate margin of normal tissue and abdominal wall repair<sup>12,14</sup>. In umbilical endometriosis, medical management is based on hormonal therapy (norethisterone, progesterone, danazol, and gonadotropin-releasing hormone analog), which can be used to reduce symptoms and downsize the endometrial nodule before surgery<sup>1</sup>. In our case we performed omphalectomy along with mesh repair of abdominal wall defect.

Among the reported cases of umbilical endometriosis associated with umbilical hernia, neither recurrence of endometriosis was seen post excision nor any malignant transformation<sup>12</sup>. A longer follow-up is required to establish recurrence and malignant transformation.

Umbilical endometriosis should be suspected in a female patient presenting with umbilical swelling associated with localized cyclical pain and/or bleeding in menstruation. PUE associated with an umbilical hernia is a very rare condition and only a few cases have been reported in the medical literature. Gold standard treatment is omphalectomy with abdominal wall reconstruction using mesh.

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### Ethics

**Informed Consent:** Herein, reported is a case of spontaneous umbilical endometriosis associated with

umbilical hernia, for which surgical excision and abdominal wall reconstruction using mesh was performed after obtaining the patient consent for the publication process.

**Peer-review:** Externally peer-reviewed.

### Author Contributions

Concept: J.B., P.K., Design: J.B., P.K., Data Collection and/or Processing: S.H., A.K., Analysis and/or Interpretation: J.B., P.K., A.K., Critical revision: J.B., P.K., S.S., D.B., Writing: S.H., J.B.

**Conflict of Interest:** The authors declare that they have no conflict of interest.

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