

# Umbilical Endometriosis in a Surgically Naïve Multiparous Young Female

Renu Singh, Sushma Bharti, Harpreet Kaur<sup>1</sup>, Saroj Kumar Yadav<sup>2</sup>

Departments of Pathology, <sup>1</sup>Obstetrics and Gynaecology and <sup>2</sup>General Surgery, All India Institute of Sciences, Bilaspur, Himachal Pradesh, India

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### ABSTRACT

The existence of functional endometrial tissue outside the uterus is known as endometriosis. It is a benign estrogen-dependent gynecological condition that affects 5%–10% of women who are of reproductive age. Endometriosis often affects the ovary and fallopian tubes, although it can also occur in nonpelvic areas. The most typical location for extra-pelvic endometriosis is the abdomen. Umbilical endometriosis is a rare condition accounting for 0.5%–1.0% of all cases of endometriosis. In 3% of cases, there is a chance of malignant change. This disorder's precise etiology is uncertain. Recurrent discomfort and swelling around the umbilicus are the classic manifestations. In this instance, we describe a patient with primary umbilical endometriosis (PUE) who had cyclical bleeding and swelling over the umbilicus and was surgically naïve. This case will demonstrate how, particularly in surgically naïve instances, diagnosis of PUE is frequently delayed due to ignorance of the entity.

**KEYWORDS:** *Cyclical pain, naïve surgically, primary umbilical endometriosis, umbilical swelling, Villar nodule*

## INTRODUCTION

Endometriosis is defined as the presence of endometrial tissue outside the uterine cavity. It is a benign gynecological disorder that affects about 5%–10% of reproductive-aged women. Commonly seen in the pelvic cavity including the ovary and fallopian tubes but extra-pelvic implantation is not common and is seen in <12% of cases.<sup>[1]</sup> These include the abdominal wall, diaphragm, pulmonary, urinary bladder, kidney, brain, intestine, lymph nodes, extremities, umbilicus, hernial sac, heart, and brain.<sup>[2]</sup> Among the extra-pelvic locations, the abdominal wall is the most common site. The umbilical endometriosis is described as two types: primary and secondary. Primary umbilical endometriosis (PUE) can occur spontaneously, hence also called spontaneous endometriosis, while secondary umbilical endometriosis (SUE) occurs following open or laparoscopic surgeries. Both can be seen with or without a prior history of endometriosis.<sup>[2]</sup> PUE is an uncommon occurrence that accounts for 30%–40% of abdominal wall endometriosis. It occurs in around 0.4%–4% of extragenital lesions and 0.5%–0.1% of all endometriosis

cases.<sup>[3]</sup> This case study features a PUE who had two previous vaginal births and complained of cyclical abdominal pain and swelling over the umbilicus for the past year only. The patient visited the gynecology outpatient department. She had no prior history of endometriosis, and she lacked surgical experience.

## CASE REPORT

A 34-year-old multiparous female presented to the gynecology outpatient department with complaints of umbilical pain and swelling during the menstrual cycle for the previous year. The pain was gradually increasing. She had no prior experience with abdominal surgery. On clinical examination, there was a 2.5 cm × 2.5 cm superficial hyperpigmented enlarged swelling over the umbilicus [Figure 1]. The abdominal ultrasound was normal. Local sonography, on the other hand, revealed the presence of a relatively well-defined lesion measuring

**Address for correspondence:** Dr. Sushma Bharti, Department of Pathology, All India Institute of Sciences, Bilaspur - 110 017, Himachal Pradesh, India. E-mail: sushmabharti2222@gmail.com

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2.8 cm × 2.5 cm and closely abutting the underlying bilateral rectus muscle, with no gross infiltration and no increase in angiogenesis. Umbilical endometriosis was considered and planned for wide local excision based on the clinical history and radiological findings. A 2 cm × 2 cm mass was discovered intraoperatively at the inferolateral angle of the umbilicus which was received for histopathological examination [Figure 2a and b]. Upon microscopic analysis, fibrocollagenous tissue contained endometrial glands and stroma. Some of these endometrial glands were filled with secretions and ranged in shape from round to oval to cystically dilated. The endometrial glands are surrounded closely by a compact stroma. There was no evidence of malignancy, granuloma, or hyperplasia. The characteristics aligned with those of umbilical endometriosis [Figure 3a and b]. She had a trouble-free postoperative period and was doing well after a week of observation.

## DISCUSSION

Endometriosis is a chronic, benign condition that depends on estrogen. Scientist Rokitansky coined the term endometriosis in 1860. Villar discovered PUE in 1886, which is why the condition is also referred to as Villar's nodule.<sup>[4]</sup> Because of the overproduction of prostaglandins, cytokines, and chemokines brought on by this ectopic tissue, endometriosis is also a chronic inflammatory disease that causes vague pain.<sup>[5,6]</sup>

Several theories explain the unclear genesis of extra-pelvic endometriosis, including immunologic defects, coelomic metaplasia, retrograde menstrual flow theory, hematogenous or lymphogenic spread, and embryonic remnants in the umbilical fold (such as the urachus and umbilical vessels). However, in the case of isolated umbilical endometriosis, it is thought that the disease may develop as a result of urachus remnant metaplasia.<sup>[5]</sup>

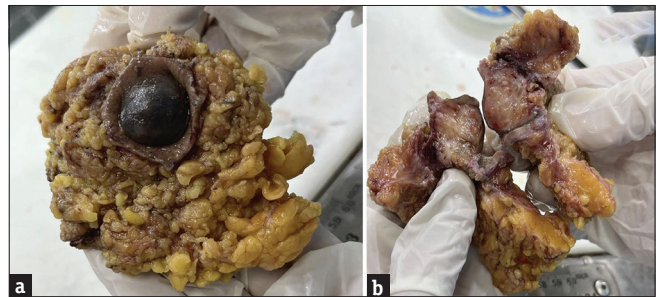
Although it can occur in premenopausal females, it is typically observed in the reproductive age group.<sup>[7]</sup> Recurrence is observed even though the condition is benign. According to Hirata *et al.*, there is a 3% chance that umbilical endometriosis (UE) will transform malignantly.<sup>[3]</sup>

SUE in patients with abdominal endometriosis is primarily seen in those who have had surgery in the past. The most typical location for SUE is the umbilicus. It hypothetically acts as a physiological scar and has a predilection for ectopic endometrial implants.<sup>[8]</sup> The classification of primary and SUE is done to exclude the disease's underlying cause.

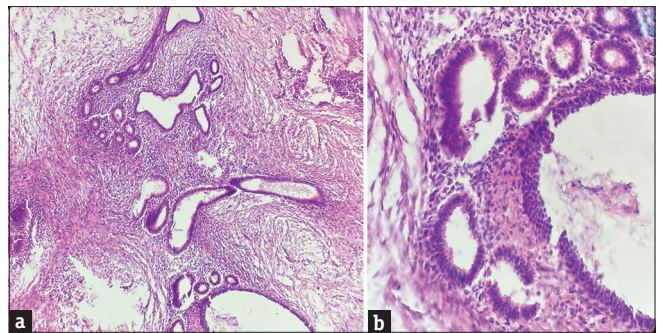
Red-to-brownish or flesh-colored nodules are the typical presentation of UE. Dysmenorrhea, bleeding, or swelling



**Figure 1:** Clinical picture of umbilical nodule



**Figure 2:** (a and b) Gross appearance of the umbilical nodule



**Figure 3:** (a and b) Microscopic examination shows fibrocollagenous tissue with endometrial glands and stroma surrounded closely by a compact stroma (H and E, ×20 and ×40)

of the lesion, however, are the most common symptom that indicates the diagnosis.<sup>[4]</sup>

The imaging modalities are not diagnostically specific, and the symptoms lack always-cyclic characteristics.<sup>[6]</sup> Magnetic resonance imaging and ultrasounds are typically performed, though. The most prominent symptom in this case was unbearable pain that was followed by umbilical swelling during her menstrual cycle.

The most frequent clinical differential diagnoses for swelling around the navel are granular cell tumor, hemangioma, urachal residual, pyogenic granuloma, keloid, and pemphigus vegetans. It is important to rule out malignant lesions such as melanoma, adenocarcinoma, squamous, and basal cell carcinoma.<sup>[9,10]</sup>

Wide local excision is the cornerstone of treatment, and only a histopathological examination can confirm the diagnosis. After surgery, the patient must be monitored because recurrence is possible.

## CONCLUSION

PUE is an uncommon thing. Usually, it remains undetected until menarche. Despite being a benign condition, there is a reported risk of malignant transformation. Even when there is growth and periodic bleeding, the diagnosis is frequently postponed. Even in the absence of a prior history of abdominal surgery, a woman presenting with pain and swelling at the umbilicus should be treated with the utmost suspicion. Histopathological examination is the gold standard for diagnosis confirmation, but follow-up is necessary because of the chance of recurrence.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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