

SHORT REPORT

Primary umbilical endometriosis: Radical excision (omphalectomy) and laparoscopic management of associated pelvic endometriosis in a low resource tertiary hospital

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Abstract

Primary umbilical endometriosis (PUE) is a rare condition affecting 0.5 – 1% of all extragenital endometriosis cases. We reviewed the data of five women with umbilical endometriosis retrospectively. The age range was 29 – 46 years, and they were all nulligravid at presentation. Common clinical presentation was umbilical pain and masses, dysmenorrhea, and primary infertility. Radical umbilical excision was performed to remove the nodule as a definitive treatment. Diagnostic laparoscopy was performed, followed by varying degrees of operative laparoscopic procedures. They all had endometriosis in the pelvis. Three out of five women operated became pregnant and had live births. Complete resolution of clinical symptoms with a reduction in umbilical and menstrual pain scores occurred. In resource-constrained settings, diagnosis, and treatment of PUE may be challenging. Clinical suspicion and appropriate case management are critical for good reproductive outcomes and quality of life. (*Afr J Reprod Health* 2021; 25[4]: 167-173).

Keywords: Umbilical endometriosis, umbilical nodule, umbilical bleeding, infertility, reproductive outcome, quality of life

Résumé

L'endométriose ombilicale primaire (PUE) est une maladie rare affectant 0,5 à 1 % de tous les cas d'endométriose extragénitale. Nous avons examiné les données de cinq femmes atteintes d'endométriose ombilicale rétrospectivement. La tranche d'âge était de 29 à 46 ans, et ils étaient tous nulligravides à la présentation. Le tableau clinique commun était la douleur et les masses ombilicales, la dysménorrhée et l'infertilité primaire. Une excision ombilicale radicale a été réalisée pour retirer le nodule comme traitement définitif. La laparoscopie diagnostique a été réalisée, suivie de divers degrés de procédures laparoscopiques opératoires. Ils avaient tous une endométriose du bassin. Trois femmes opérées sur cinq sont tombées enceintes et ont eu des naissances vivantes. Une résolution complète des symptômes cliniques avec une réduction des scores de douleur ombilicale et menstruelle s'est produite. Dans les milieux à ressources limitées, le diagnostic et le traitement du PUE peuvent être difficiles. La suspicion clinique et une prise en charge appropriée des cas sont essentielles pour de bons résultats en matière de reproduction et une bonne qualité de vie. (*Afr J Reprod Health* 2021; 25[4]: 167-173).

Mots-clés: Endométriose ombilicale, nodule ombilical, saignement ombilical, infertilité, résultat reproductif, qualité de vie

Introduction

Endometriosis is a benign gynaecologic condition defined by endometrial glands and stroma outside the uterus¹. It affects 6 to 10% of women of reproductive age, with higher rates among infertile

women undergoing laparoscopy². It occurs mostly in the pelvic cavity, with the commonest sites being ovaries, the pouch of Douglas, and uterosacral ligaments³. Extra-pelvic endometriosis is encountered in 12% of women diagnosed with the disease. These include the intestinal tract, urinary

tract, peritoneum, omentum, lung, thoracic cage, and cutaneous tissues such as the surgical scars and abdominal wall⁴. Locations like nasal mucosa, brain, and eyes are exceedingly rare⁵.

Umbilical endometriosis (UE) is rare, accounting for 30% of cutaneous endometriosis (CE), and could be primary or secondary⁶. Primary umbilical endometriosis (PUE) accounts for 0.5 – 1% of all extragenital endometriosis and is defined as the presence of ectopic endometrial tissue in the umbilicus without previous abdominal surgery while secondary UE occurs following caesarean section, laparotomy, or laparoscopy^{6,7}. The lesion undergoes enlargement slowly over time, and malignant transformation has been described⁸. Management options include medical treatment, often palliative, and definitive surgical treatment. This study reviewed five PUE cases managed over five years in our center.

Description of case series

We followed up five women diagnosed and managed with UE in our hospital between 2015 and 2020. Data were retrospectively analyzed. Ethical review committee approval was waived because of the study's retrospective nature. The study protocol complied with the Declaration of Helsinki. The age range was 29 to 46 years, and they were all nulligravid at presentation. The duration of symptoms prior to presentation ranged between 1 to 4 years. All the patients had umbilical masses and pain, dysmenorrhea, and primary infertility. Four out of five cases had cyclical bleeding from the umbilicus (P2 to P5). Primary umbilical endometriosis was not recognized in patient no. 1 and 4 before referral when symptoms did not resolve. P1 had exploration of the umbilical mass by a general surgeon on account of suspected incarcerated umbilical hernia while P4 had open myomectomy by a general practitioner on account of uterine fibroid.

Recruitment and preoperative evaluation

Five women who presented with a history of painful umbilical nodules with or without cyclical bleeding during or after menstruation were included in the series. They had no previous gynaecological surgery/caesarean section or symptoms predated surgery. They were counselled about their condition, the need for surgery, and the nature of

the surgery, they were informed of the possibilities of encountering pelvic endometriosis, and the possible therapeutic options and prognosis were explained. Informed consent was obtained from all the patients before surgery, and it included permission to collect non-surgical and surgical information and use the patients' specimens, images, and videos for any study.

Routine investigations, including full blood count (FBC), renal function test, urinalysis, blood glucose, cardiac function test, and viral screening, were carried out and normal. Evaluation of male infertility with semen analysis was within normal limits. Transvaginal scan (TVS) showed normal uterus, ovarian cysts, and persistent fluid at the pouch of Douglas at a variable period. Hysterosalpingography (HSG) and hormonal assays were normal. All patients had an umbilical biopsy to confirm the diagnosis before the radical umbilical excision was performed to remove the nodule as a definitive treatment. The first (P1) and second (P2) patients refused preoperative Goserelin 3.6mg (Zoladex®) a GnRH analogue because of the possible side effects while others P3, P4, and P5 received preoperative Goserelin.

Surgical technique

Surgery was performed in the proliferative phase of the menstrual cycle, under general anaesthesia with cuffed endotracheal intubation. Prophylactic antibiotics were administered with Intravenous Ceftriaxone and Metronidazole. Radical excision (Omphalectomy) was performed by excising the umbilicus en block with the nodule. The incision was carried around the umbilicus, ensuring normal tissue was visible at the margins. The dissection involved the skin, subcutaneous tissue, and the rectus fascia. Haemostasis was ensured using monopolar diathermy.

Diagnostic laparoscopy was performed using Hasson's technique, entering through the opening where the umbilicus was removed. A 10-mm cannula without trocar was inserted, an airtight seal was ensured around the cannula with the use of nylon sutures and the peritoneum was insufflated with CO₂ gas. A 10-mm 30-degree Karl Storz telescope and light source connected to a Karl Storz camera was inserted into the peritoneal cavity. Additional 5-mm trocar and cannula were inserted as appropriate under vision, and the abdominal cavity was inspected.



Figure 1: Preoperative umbilical images of all the patients

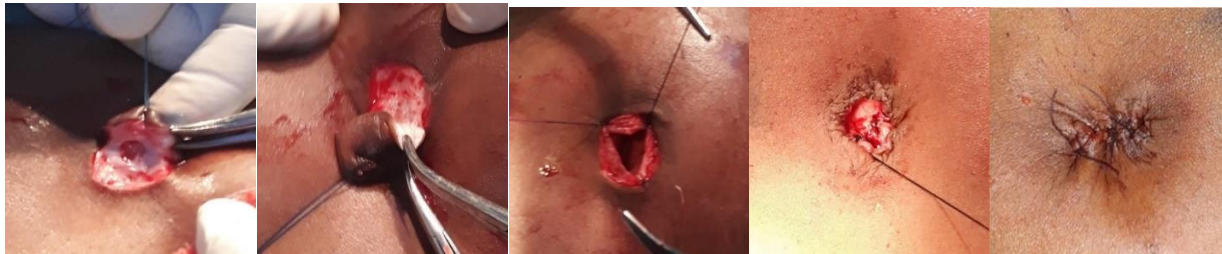


Figure 2: Intraoperative images for patient No. 3 (P3)



Figure 3: Postoperative Umbilical images of the patients

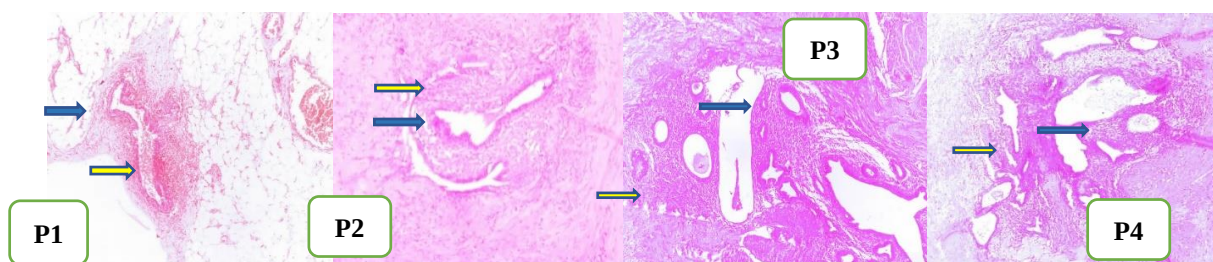


Figure 4: Photomicrographs showing endometrial glands and stroma H&E X 10. Blue arrows pointing to the glands while yellow arrows are pointing to the stroma

Varying degrees of operative laparoscopic procedures: excision/ablation of pelvic endometriotic deposits, cystectomy, and adhesiolysis were performed using a combination of graspers, scissors, and bipolar forceps. Specimens were retrieved using improvised retrieval bags made with sterile surgical hand gloves, and peritoneal lavage was done with normal saline. An Anti-adhesion agent (Guardix Sol® 5gm

to 10gm gel) was instilled into the pelvic cavity. Accessory ports were removed under vision, Carbon dioxide gas was let out, and primary ports and instruments were removed.

The closure of the umbilical wound was done by the plastic surgeon using Vicryl sutures (0 for fascia and 2/0 for skin in subcuticular fashion). Surgically, endometriosis was staged I – IV using the American Society of Reproductive Medicine

Table 1: Intraoperative findings, operative interventions, medical treatment, complications and outcome

Patients	Intraoperative findings	Operative intervention	Medical Treatment (GnRHa) ^a	Complications	Pregnancy Outcome
P1	Umbilical mass, pelvic endometriotic deposits and adhesions	Omphalectomy, excision/ablation of pelvic endometriosis, and adhesiolysis.	Refused to consent for fear of side effects	None	1 st pregnancy conceived by IVF (Live birth female baby delivered at 38weeks GA via elective CS). 2 nd pregnancy spontaneously conceived (Miscarriage at 18 weeks GA).
P2	Umbilical mass, pelvic endometriotic deposits and peri-tubal adhesions	Omphalectomy, excision/ablation of pelvic endometriosis, and adhesiolysis.	Refused to consent for fear of side effects	None	1 st pregnancy spontaneously conceived (Miscarriage at 12 weeks GA). 2 nd pregnancy spontaneously conceived (Live birth male baby at 38weeks via elective CS).
P3	Umbilical mass, omental and peritoneal endometriosis	Omphalectomy, excision/ablation of omental and peritoneal endometriosis.	Had pre & post-operative GnRHa	Port site infection	On follow-up in Gynaecology clinic. Referred for IVF
P4	Umbilical mass, pelvic endometriotic deposits and adhesions	Omphalectomy, excision/ablation of pelvic endometriosis, adhesiolysis and ovarian cystectomy.	Had pre & post-operative GnRHa	None	Pregnancy conceived by IVF (Live birth male baby at 38weeks GA via elective CS).
P5	Umbilical mass and pelvic endometriosis.	Omphalectomy, excision/ablation of pelvic endometriosis.	Had pre & postoperative GnRHa	None	Lost to follow-up in Gynaecology clinic.

Note: All patients had anti-adhesion agents (Guardix Sol®) instilled intraoperatively. ^a GnRHa= Gonadotropin Releasing Hormone Analogue, GA=Gestational age, CS=Caesarean section. IVF=In-Vitro fertilization.

(ASRM) revised classification. P1, P2, P4, and P5 had moderate (Stage III) endometriosis while P3 had mild (Stage II) endometriosis.

Intraoperative findings, operative intervention, medical treatment, complications, and pregnancy outcome

Findings at diagnostic hysteroscopy were normal in all cases. Laparoscopic intraoperative findings, operative intervention, medical treatment, complications, and pregnancy outcomes are documented in Table 1. They all had endometriotic deposits in the pelvis or abdominal cavity. Three out of five women operated became pregnant and had live births. One patient is on follow-up while another was lost to follow-up in the gynaecology clinic.

Postoperative period

All the patients did well and were discharged on the first day post-surgery. Pain relief was achieved with

parenteral analgesics and suppository Diclofenac 100mg 12 -hourly for 24 hours. Patient (P3) had a deep port-site wound infection post-surgery that was managed with antibiotics and wound dressing. Patients P3, P4 and P5 received postoperative Goserelin 3.6mg (Zoladex®). They were followed up postoperatively, and pain score was assessed using the Numeric Pain Rating Scale (NPRS). All had complete resolution of clinical symptoms with a significant reduction in umbilical and menstrual pain scores six months after surgery. None had a recurrence of CE. Pre, Intra, and Postoperative images of the umbilicus are shown in Figures 1, 2, and 3.

Histopathology confirmed the diagnosis of endometriosis in all the patients (Figure 4) (photomicrographs). Ovulation induction was used for a maximum of 6 months for the first patient (P1), and 3 months for the second (P2) and third (P3) patients. Due to advanced maternal age, P4 was referred for IVF after the procedure. Pregnancy outcome is documented in Table 1.

Discussion

Primary umbilical endometriosis is a rare condition described as endometrial glands and stroma in the umbilicus in the absence of abdominal surgeries⁶. The rare nature of this condition may pose a challenge to achieving a correct diagnosis. In our study, two out of five had had abdominal surgeries in other centers to treat their complaints before referral to us when symptoms did not resolve due to non-recognition of PUE. However, the most important thing to note was that the UE symptoms predated the abdominal surgeries in those cases. Abdominal wall endometriosis is often mistaken for other surgical conditions, as seen in patient no. 1, where exploration was carried out by a general surgeon on account of suspected incarcerated umbilical hernia without considering PUE⁹. The general practitioner who carried out the open myomectomy for patient no. 4 may not have been aware of or consider PUE before performing surgery. In 1993, Sevdel *et al.* reported that correct preoperative diagnosis was made in 20 – 50% extra-pelvic endometriosis¹⁰. Therefore, knowledge of UE, as well as early referral of umbilical lesion when in doubt, is essential for all cadre of medical doctors as this will prevent unnecessary intervention and ensure that patients receive appropriate care.

Clinical evaluation can be made from history and physical examination. Differential diagnosis of UE must be kept in mind, including benign and malignant disease conditions, while considering endometriosis¹. Ultrasonography with or without Doppler, computed tomography (CT), magnetic resonance imaging (MRI), fine needle aspiration cytology (FNAC), biopsy, are some of the ancillary investigations that can help in diagnosis with varying degrees of specificity and sensitivity. Magnetic resonance imaging may help assess the lesion's depth, primarily when related to the fascia or peritoneum¹⁰. Carrying out MRI is a challenge in our environment where the test is not easily accessible, and the average cost (\$200) is out of reach for most patients. However, histopathological examination is the hallmark of diagnosis using the standard hematoxylin and eosin-stained slide¹¹. This test is useful not only to establish a diagnosis when endometrial glands, stroma, and hemosiderin pigments are seen but also to rule out the presence of malignancy¹¹.

The age range at presentation in this study (29 to 46 years) was within the reproductive age. This finding is similar to the age range of 23 to 48 years recorded by Chrysostomou and colleagues in South Africa¹². The study by Victory *et al.*, in Canada, recorded a wider age range of 23 to 58 years, although all the patients were within the reproductive age except the singular case of a 58-year old woman who presented with UE coexisting with metastatic endometrial adenocarcinoma¹³. The range of duration of symptoms before presentation was 12 to 48 months. This implies that patients suffer UE for a long period of time before seeking intervention. This finding was similar to the range of 6 to 48 months reported in South African. Late presentation was expected to be worse among Africans but surprisingly, a much wider range of 2 weeks to more than 12 years was reported in the Canadian study¹³. The late presentation among their study population was attributed to misdiagnosis which also occurred in two patients in our case series.

The most common symptoms in this study were cyclical pain, umbilical mass, and infertility. All patients in this study had these symptoms. Four out of five cases also had cyclical bleeding from the umbilicus. These symptoms are also reported by other researchers^{1,6-14}. Pelvic endometriosis was found during diagnostic laparoscopy in all the patients. This finding was similar to that of Malebranche and co-workers who reported a case with associated pelvic endometriosis¹⁴. However, the finding is different from Chrysostomou and colleagues' work, where the most common symptoms are cyclical pain and umbilical mass without infertility, and none of their patients had pelvic endometriosis¹². They recommended that there is no need for laparoscopic assessment in cases of PUE. This view was also shared by other researchers¹². However, our study's findings of associated pelvic or peritoneal endometriosis in all the five patients studied showed that this recommendation might need critical evaluation.

Radical excision of the umbilicus, along with the nodular mass, was performed in all cases, ensuring that an adequate rim of normal tissue surrounds the edges, and the umbilicus was reconstructed in collaboration with a plastic surgeon. This treatment was adequate to prevent recurrence in all cases. This practice is comparable to that of other researchers^{1,6-14}. In our study, an

omphalectomy was done before diagnostic laparoscopy, and abdominal entry was via an open (Hasson) technique. This technique's advantage is the perceived reduction in failed-entry, vascular, and bowel injury during the primary trocar's insertion at laparoscopy. This approach contrasts with some other researchers who performed diagnostic laparoscopy before the umbilicus' radical excision¹⁵. In their technique, the ability to visualize the intraabdominal portion of the umbilicus, ensured there was no attachment of intra-abdominal structures before excision is an advantage. However, any entry technique can safely be done when it is established via USS or MRI that there is no attachment of intra-abdominal structures to the umbilicus. Both techniques have not been reported to be associated with increased complications and any could be used.

Conclusion

Although it is difficult to extrapolate findings from a case series to the general population, the outcome obtained in this study indicates the possible effectiveness of omphalectomy and the usefulness of laparoscopic management of associated pelvic endometriosis. Lastly, the resolution of clinical symptoms suggests a reduction in disease burden and though this should be interpreted with care considering the small sample size, the management approach may have contributed to the good fertility outcome that was recorded among our study population. Further studies with a larger sample size could inform the applicability of our combined approach to the management of PUE and associated fertility challenges whether through spontaneous or assisted conception.

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Conflicts of interest

All authors have no conflicts of interest to declare.

Authors contribution

AAA conceived the study. AAA, AOA, MAO, NCN, AKT, AAN designed the study. AOA, AAA, MAO operated on P1 as first, second, and third assistants respectively. AAA, MAO, NCN, MUF, AO, OSC, operated on P2, P3, P4, and P5. EOS performed umbilical reconstruction. AAA, AOA, MAO, NCN, MUF, AO, OSC, AKT, AAN were involved in pre-and post-operative management of the patients. FOO, ADI prepared the specimens, read, and reported the histopathology, they also provided the photomicrographs. Acquisition of data and analysis of data was done by AAA, AOA, MAO, NCN, MUF, AO, OSC. The manuscript was prepared by AAA, MUF. The manuscript review was done by AAA, AOA, MAO, CNC, MUF, FOO, AAN, AKT. All authors reviewed the draft manuscript and approved the final version of the manuscript.

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