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Case Report

Multidisciplinary management of primary umbilical endometriosis (Villar's nodule) with coexistent uterine leiomyomata: A case report

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Abstract

Introduction: Primary umbilical endometriosis, also known as Villar's nodule, is a rare manifestation of endometriosis within the umbilicus.

Case description: A 41-year-old female who presented with a 4-year history of umbilical swelling associated with cyclic umbilical bleeding, dysmenorrhoea and heavy menstrual bleeding is herewith reported. Imaging procedures revealed an umbilical mass and uterine leiomyomas. The patient underwent combined surgery by the gynaecology and plastic surgery teams where an open myomectomy followed by omphalectomy, mesh repair of the rectus sheath and panniculectomy was done. Histopathological examination confirmed the presence of estrogen and progesterone receptor-positive endometrial glands and stroma in the umbilicus consistent with primary umbilical endometriosis and coexistent uterine leiomyomas. Clinical improvement was demonstrated 3 months following the surgery.

Conclusion: Umbilical swelling associated with catamenial symptoms should raise suspicion for primary umbilical endometriosis. Inter-specialty approach is key in the management of primary umbilical endometriosis.

Keywords

Primary umbilical endometriosis, uterine fibroids

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Introduction

Primary umbilical endometriosis defined as spontaneous ectopic tissue possessive of endometrial function within the umbilicus is rare and adopts its name, Villar's nodule, from Dr. Villar who first described it in 1886.[1] There is limited literature on the coexistence of primary umbilical endometriosis and uterine leiomyomas. We document a case of primary umbilical endometriosis to familiarize a global readership with the occurrence of this rare phenomenon that required resection and reconstruction of the anterior abdominal wall in an indigenous African woman with symptomatic uterine leiomyomas.

Case presentation

A 41-year-old para 1 on treatment for hypertension presented to the Kenyatta National Hospital Gynaecology clinic with a 4-year history of umbilical swelling associated with umbilical bleeding that was concurrent with her menstrual bleeding. The umbilical bleeding was

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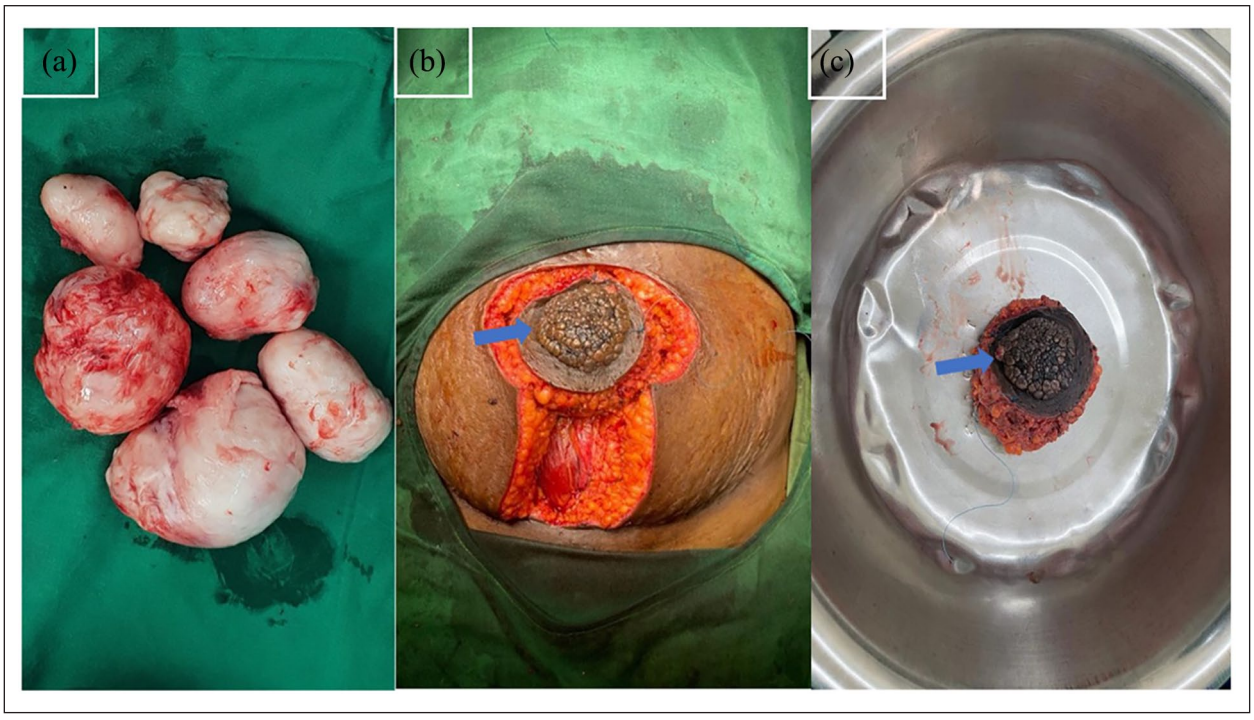

Figure 1. (a) Leiomyoma specimens, (b) wide excision of the endometriotic nodule and umbilicus (omphalectomy) and (c) macroscopic appearance of the omphalectomy specimen.

significant enough to require the placement of a sanitary towel and was accompanied by severe dysmenorrhoea and heavy menstrual bleeding. The symptoms were associated with emotional distress, anxiety and low self-esteem as she had parted ways with her sexual partner due to the same. She did not report dyspareunia, dyschezia, haematochezia or haematuria and had visited three general practitioners who put her on antifungal creams with no regression of symptoms.

The patient reported to have had a term hospital vaginal delivery to a male infant who weighed 3.8 kg and was alive and well. There was no history of postpartum haemorrhage or endometritis following delivery. The pregnancy was conceived naturally. There was no familial or personal history of endometriosis, abdominopelvic surgery or uterine instrumentation.

Abdominal examination revealed central adiposity and a brown multinodular and irregular umbilical mass measuring 6 cm × 5 cm. It was non-tender, firm and fixed to the anterior abdominal wall. There was no obvious bleeding or discharge from the mass. Multiple, regular, firm, non-tender masses that corresponded to a fundal height of 20 weeks were palpated on bimanual examination. Ultrasonography showed a heterogenous hypoechoic umbilical mass measuring 7.5 cm × 7.4 cm × 6.5 cm and a bulky uterus illustrating multiple heterogenous intramural masses. Her cervical cytology was normal.

Under a presumptive diagnosis of primary umbilical endometriosis and symptomatic uterine leiomyomata, she underwent a combined exploratory laparotomy by the gynaecological and plastic and reconstructive surgeons. The umbilical mass was adherent to the rectus sheath. The uterine wall was intact with no communication to the umbilical mass. There was no evidence of pelvic endometriotic implants. The uterus was bulky with six hybrid leiomyomas, the largest measuring 7 cm × 5 cm, which were meticulously enucleated with successful repair of the uterus. Care was taken not to breach the endometrium and haemostasis was achieved. Wide excision of the endometriotic nodule and the umbilicus with a 2 cm margin of normal skin (omphalectomy) was then performed (Figure 1).

Repair of the rectus sheath using a sub-lay prolene mesh followed by panniculectomy which involved undermining and apposition of the lateral flaps below the Scarpa's fascia and trimming off of the overlapping ends was done. Closure was then done in layers without recreating an umbilicus (Figure 2). Umbilicoplasty was scheduled as a secondary procedure.

The histopathology examination demonstrated a normal epidermis beneath which irregular endometrial glandular spaces surrounded by endometrial stroma were appreciated consistent with primary umbilical endometriosis. Immunohistochemistry showed estrogen receptor (ER) and progesterone receptor (PR) expression with negative cytokeratin-20 staining. Sections from the uterine masses revealed fascicles of spindled cells with eosinophilic cytoplasm in keeping with leiomyomas. There was no evidence of malignant tissue. (Figure 3)

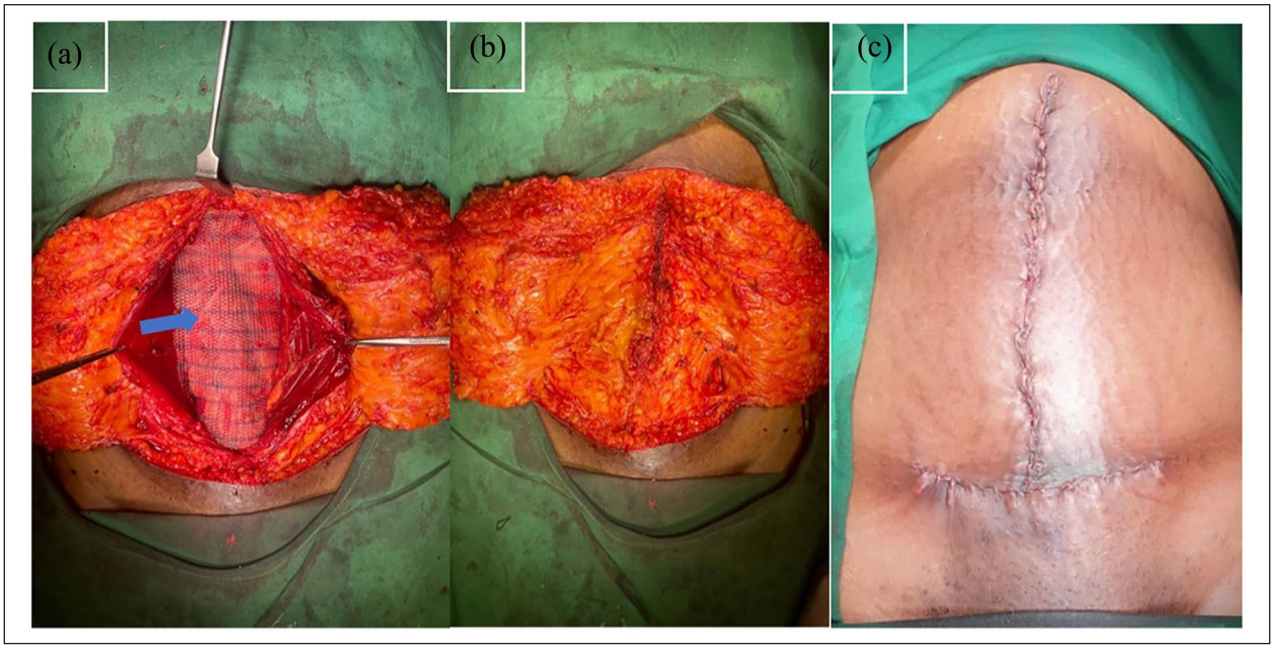

Figure 2. (a) Repair of the rectus sheath using a sub-lay prolene mesh (blue arrow), (b) panniculectomy and (c) skin closure.

The immediate postoperative period was uneventful. She was discharged home on the seventh postoperative day having undergone contraception counselling. She was appraised of the obstetric complications associated with myomectomy scars as well as the need to have a planned cesarean delivery in her subsequent pregnancy. The risk of subfertility following myomectomy was also explained and she was discharged through the mental health, plastic surgery, hypertension and gynaecology clinics. She reported recurrence of her menses with complete resolution of the umbilical bleeding, heavy menstrual bleeding, pelvic pain and emotional distress on her 3-month follow-up visit. She was not keen on having the umbilicoplasty.

Discussion

Umbilical endometriosis is rare with an incidence of 0.5-1%.[2] While secondary umbilical endometriosis occurs as an aftermath of endometrial tissue dissemination during laparotomy or laparoscopy with umbilical port entry, primary umbilical endometriosis occurs spontaneously. The pathophysiology of primary umbilical endometriosis is not well understood. It is thought to arise from metaplastic changes of urachal remnants or contamination of the umbilical cord with endometrial tissue during childbirth.[3]

The mean age at diagnosis of primary umbilical is 37.7 years.[4] The case described developed an umbilical mass at the age of 37 years which demonstrates the estrogen-dependent susceptibility of women in the childbearing age to endometriosis. About 90% of patients with primary umbilical endometriosis present with an umbilical swelling associated with cyclic pain and umbilical bleeding.[4] This explains the importance of this finding in aiding the clinical suspicion of umbilical endometriosis. The umbilical swelling is often brown but may be blue-purple, red or black in decreasing order.[4] The average size of the nodule at diagnosis is 1–3 cm.[5] The case in discussion had a larger lesion measuring 6 cm × 5 cm, reflecting a delay in the diagnosis and treatment of her endometriosis. In Kenya, the delay is attributed to lack of societal awareness about endometriosis, the normalization of pain and stigma.[6]

The definitive diagnostic method for primary umbilical endometriosis is the histopathological demonstration of endometrial glands or stroma within the umbilicus following excisional biopsy which presents both a diagnostic and therapeutic advantage compared to fine needle aspiration which is solely diagnostic.[5] Preoperative imaging using ultrasonography or magnetic resonance imaging(MRI) is useful in further examining the nodule, its relation to surrounding tissue, co-existing pelvic pathologies and optimally preparing for surgery.[5] It also aids in ruling out differential diagnoses like polyps, hernias and Sister Mary Joseph's nodule among others. The sonographic features of primary umbilical endometriosis include an irregular heterogenous and hypoechoic lesion. The presence of homogeneous hyperintense lesions on T1-weighted or hypointensity on T2-weighted MRI highly suggests endometriosis.[4] The diagnosis of Villar's nodule and uterine leiomyomas in the case presented was aided by ultrasonography.

Only 20% of umbilical endometriosis coexists with pelvic endometriosis.[3] There was no evidence of pelvic endometriotic implants in our case, but multiple uterine leiomyomas were noted intraoperatively. The simultaneous

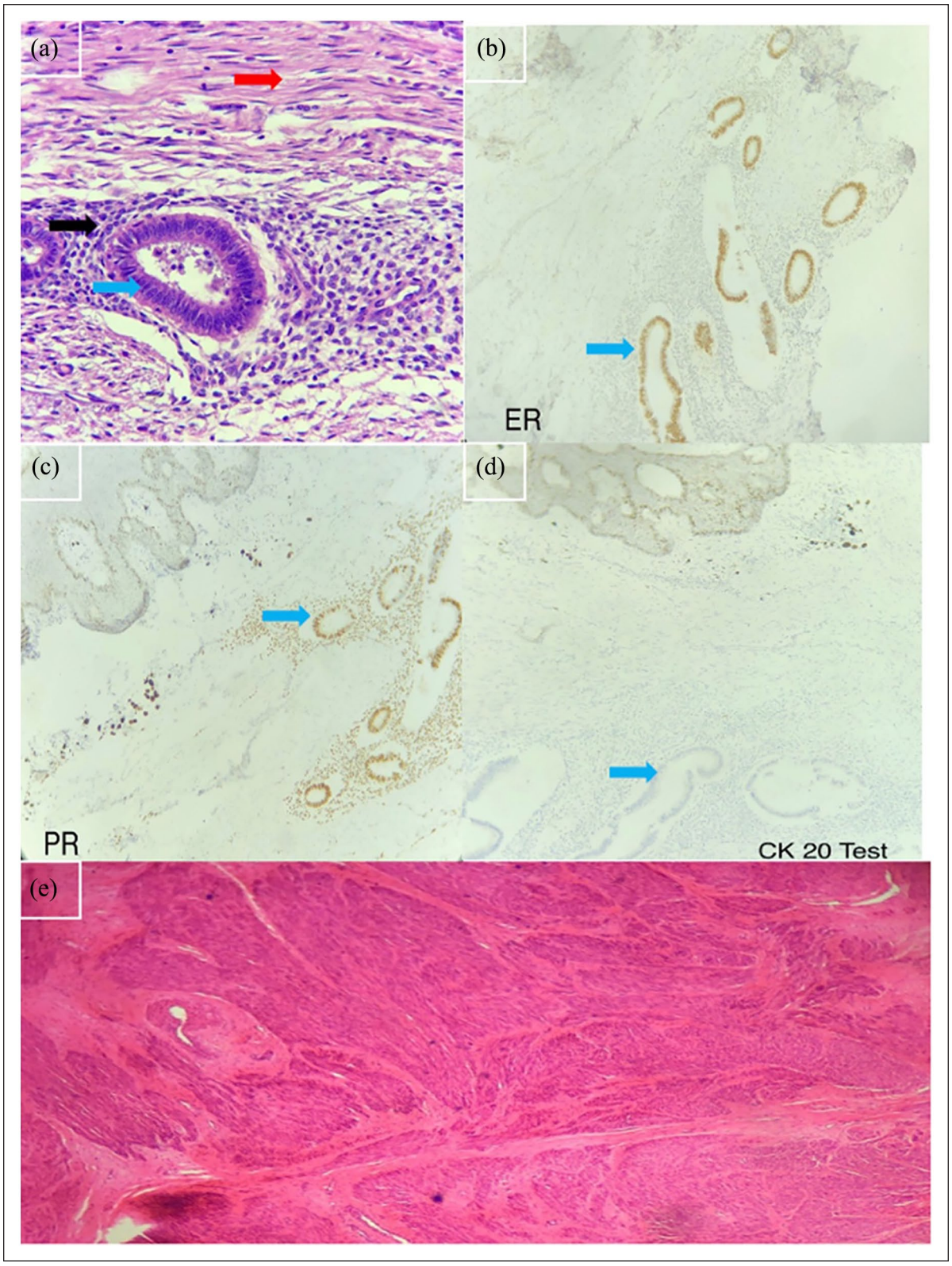

Figure 3. (a) Photomicrograph of the excised nodule showing endometrial glands (blue arrow) surrounded by endometrial stroma (black arrow) beneath the epidermis (red arrow). Immunohistochemistry images showing (b) estrogen receptor (ER) positivity (c) progesterone receptor (PR) positivity and (d) Cytokeratin-20 negative staining. (e) microscopic image of the uterine masses showing fascicles of spindled cells with eosinophilic cytoplasm in keeping with uterine leiomyoma.

occurrence of primary umbilical endometriosis and uterine fibroids is similarly rare.[7] It has been documented that T homozygote and T allele of epidermal growth factor receptor gene 2073 polymorphism increases the risk of both endometriosis and leiomyoma development.[8]

Surgical excision is the gold standard treatment for Villar's nodule.[5] An inter-specialty approach with plastic and reconstructive surgeons in treating primary umbilical endometriosis improves patient outcomes.[9] In this case, the patient presented a large umbilical nodule that required resection of part of the anterior abdominal wall. She ended up having a tummy tuck from the panniculectomy, which improved the patient's aesthetic appearance in addition to the anterior abdominal wall reconstruction.

The risk of recurrence following excision of umbilical endometriosis is as low as 1%.[10] A recent study found no significant difference in the recurrence of umbilical endometriosis between those who underwent wide local excision including the peritoneum and those who underwent excision without peritoneal excision. There was also no difference in the recurrence rate between those who received and those who did not receive postoperative hormonal therapy.[10] This suggests that surgery is an effective treatment for this type of endometriotic lesion.

Being a chronic disease, endometriosis significantly reduces the quality of life of the affected women. According to an analysis of women living with endometriosis in Kenya, chronic pain disrupts women's social and professional lives and in turn, negatively impacts their mental health.[6] Psychotherapy therefore forms an integral part of the management of endometriosis patients. Our case was linked to the mental health clinic for appropriate care.

Conclusion

Women of reproductive age presenting with umbilical swelling associated with catamenial symptoms should be investigated for primary umbilical endometriosis. Though rare, the coexistence of this condition and uterine leiomyomas is not unusual. The treatment of primary umbilical endometriosis calls for a multidisciplinary approach including gynaecologists, plastic and reconstructive surgeons, pathologists and psychologists. Surgical excision is the mainstay of treatment.

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None

Author contributions

RSG wrote the case report under the supervision of SM and EM. RSG is a resident in Obstetrics and Gynaecology. SM was the lead gynaecologist in this case. EM is a senior lecturer in biochemistry and a board member of the World Endometriosis Society. He critically reviewed the case report. ED is a resident in human pathology. He examined the specimens and wrote the histopathology aspect of the case report under EW's supervision. MFK is a resident in plastic surgery. He was the assistant plastic surgeon and wrote the plastic surgery aspect of the case report under the supervision of BW who was the chief plastic surgeon. All authors contributed to the final manuscript.

Data availability statement

Data sharing not applicable to this article as no datasets were generated or analysed during the current study.

Declaration of conflicting interests

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Ethical approval

The Kenyatta National Hospital, University of Nairobi Ethics and Review Committee granted authority for publication of this case report (Ref: KNH-ERC/ 01/PUB/1).

Informed consent to participate

Written informed consent was obtained from the patient.

Informed consent to publish

Written informed consent was obtained from the patient for their anonymized information to be published in this article

Consent for publication

Informed consent for participation and publication was obtained from the patient.

Trial registration

Not applicable.

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