Case report

Primary umbilical endometriosis coexisting with multiple uterine fibroids: A case report

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ABSTRACT

Introduction: Uterine fibroids and endometriosis are independent causes of infertility/subfertility in women of reproductive age. Primary umbilical endometriosis is rare.

Presentation of case: Here, we report a case of primary umbilical endometriosis coexisting with multiple uterine fibroids in a 35 year old nulliparous woman who presented with abdominal swelling as well as cyclical pain and swelling of the umbilicus without any previous surgery. She had abdominal myomectomy and excision of the umbilical lesion with histological confirmation of uterine fibroids and umbilical and peri-umbilical endometriosis.

Discussion: Primary umbilical endometriosis should be considered as a possible differential diagnosis in cases of umbilical disorders even if the patient has no typical symptoms of pelvic endometriosis. The clinical features include an umbilical swelling (90%), often associated with cyclical pain (81.5%) and bleeding or discharge (49.2%); while some patients may be asymptomatic. The diagnosis of umbilical endometriosis could be made based on clinical findings but histological confirmation is the gold standard for diagnosis. The definitive treatment for umbilical endometriosis is surgical excision.

Conclusion: Although rare, primary umbilical endometriosis may coexist with uterine fibroids and should be suspected in women of reproductive age who complain of cyclical umbilical disorders in addition to abdominal swelling or other symptoms of uterine fibroids.

1. Introduction

Endometriosis is one of the common gynaecological disorders among women of reproductive age. It is defined as the presence of functional endometrial glands and stroma outside the normal uterine cavity. It is a benign gynaecological disorder reported to affect about 10–15% of all women of reproductive age and 6% of the perimenopausal women [1,2]. The pathogenesis of endometriosis is not very clear. However, there are several theories that try to explain the possible causes of the development of endometriosis, including retrograde menstruation, direct spread, embryonal rest, coelomic metaplasia, and lymphatic or hematogenous spread. It is associated with chronic pelvic pain, dysmenorrhea, dyspareunia and infertility.

Endometriosis is an estrogen-dependent disease that can appear anywhere in the body. Commonly affected areas include the peritoneum, the ovaries, anterior and posterior pouch of Douglas, posterior broad ligaments, uterosacral ligaments, fallopian tubes, sigmoid colon, appendix and round ligaments; and less common in the umbilicus, lungs, thorax, brain and pericardium [3].

The umbilicus is the commonest cutaneous site of endometriosis outside of the pelvis; however, it still remains a rare occurrence with an incidence of approximately 0.5–1.2% in all cases of endometriosis [4]. Primary (spontaneous) umbilical endometriosis was first described by Villar in 1886, and it represents 75% of all cases of umbilical endometriosis [4]. Secondary umbilical endometriosis develops following previous surgery (caesarean section, abdominal hysterectomy, appendicectomy, laparoscopy etc.) and is due to iatrogenic seeding of endometrial tissue [5].

Here we discuss a case of primary umbilical endometriosis in a nulliparous woman who was being managed for multiple uterine fibroids. The SCARE Guidelines were strictly followed in making this report [6].
2. Case report

A 35 year old single nulliparous patient presented to the gynaecology clinic on referral following an ultrasound finding of uterine Fibroid. She had noticed an abdominal swelling which gradually increased in size over four years and was subsequently associated with dysmenorrhea and heavy menstrual bleeding. About a year prior to presentation she developed cyclical increase in size of her umbilicus during her menstruation with associated pain. There was no bleeding or discharge from the umbilicus which usually reduced in size after the menses. She had no past history of any surgery. There was no history of dyspareunia, change in bowel habit, urinary symptoms or weight loss (Figs. 1, 2).

On examination, the abdomen was distended and moved with respiration. There was a hyperpigmented umbilical swelling (mass) that measured 4 × 4 cm, non-tender, firm with no expansile cough impulse. It was not attached to the underlying structures or overlying skin and not reducible. There was also a non-tender pelvic mass of 32 weeks size, with nodular surface. Abdominopelvic ultrasound scan revealed a large uterus that harboured multiple roundish heteroechoic masses, some with rim calcification. The largest measured about 8.9 cm in diameter. The endometrial stripe was distorted. The ovaries were visualized and appeared normal. There was no adnexal mass and no fluid in the pouch of Douglas. Preoperative hemogram, biochemical and coagulation profile were normal.

Following a clinical diagnosis of huge uterine fibroid and umbilical endometriosis, she had abdominal myomectomy and excision of the umbilical mass. Intra-operative findings were an umbilical nodule that was firm and measured 4x4cm; a periumbilical mass that was soft, measuring 3 × 2 cm and located in the subcutaneous layer, multiple uterine fibroids of various sizes, about 14 in number located within the subserosa, intramural and submucous areas, with the largest one measuring about 20 × 15 cm. Both uterine tubes were adherent to the uterus with the left tube kinked. The ovaries looked apparently normal. No evidence of intrapelvic endometriosis was identified (Figs. 3 and 4).

Histological findings of the fibroid nodules showed fourteen encapsulated nodular masses measuring between 0.7 cm and 13 cm in their widest diameter. Cut surfaces were whitish, whorled and similar. On microscopic examination, sections of the nodular masses were similar.
and showed benign mesenchymal proliferations consisting of long interlacing fascicles of smooth muscle cells. These cells had abundant eosinophilic fusiform cytoplasm and did not present nuclear atypia or mitotic activity. There were hyalinized areas within the stroma. The histological diagnosis was uterine leiomyomatosis (Fig. 5).

The umbilical end tissue was a spherical nodular mass measuring 4 cm × 3 cm × 2 cm. There was an ellipse of skin fragment on the surface measuring 4 cm × 2 cm. Cut surface was greyish white with some dark brown spots. Periumbilical tissue was a cystic spherical mass measuring 3 cm × 2 cm × 1 cm. Cut section revealed a smooth monocystic cavity containing dark brown fluid.

Sections of the umbilical and peri-umbilical tissues, on microscopic examination, showed connective tissue within which there were frequent foci of endometrial glands and stroma. The glands were lined by a single layer of columnar cells which did not show any atypia. The stroma did not show any features of invasion by the glands or tissue reaction. The histological diagnosis was umbilical and peri-umbilical endometriosis.

She remained stable throughout the post-operative days and was discharged on the 10th day post operatively. She was seen in the gynaecology clinic two weeks later and the histology report discussed with her. She had been seen again in the Gynaecology clinic on two occasions and still remained stable.

3. Discussion

The theory of lymphatic and hematogenic transplantation has been suggested for cases of umbilical endometriosis with pelvic endometriosis. But in a case of isolated umbilical endometriosis as was with our patient, the disease may occur through metaplasia of urachal residues [7]. Primary umbilical endometriosis is a rare entity but should be considered as a possible differential diagnosis in cases of umbilical disorders even if the patient has no typical symptoms of pelvic endometriosis. Umbilical endometriosis presents as a rubbery or firm nodule, and the size varies from several millimeters to 6 cm. The clinical features include an umbilical swelling (90%), often associated with cyclical pain (81.5%) and bleeding or discharge (49.2%); while some patients may be asymptomatic [8,9]. The index patient presented with cyclical pain and swelling of the umbilicus with an umbilical lesion that measured 4 cm ×
in addition, she had multiple uterine fibroids that gave rise to a pelvic mass of 32 weeks’ size which was her major concern. In a case series of five African patients with umbilical endometriosis, the highest lesion measured 4 cm in diameter with an average size of 3.02 cm [10]. Yunusa et al. reported a case of umbilical and ovarian endometriosis coexisting with multiple uterine myomas [11]. Other studies have also shown the coexistence of pelvic endometriosis with uterine leiomyoma [9,12–14]. No evidence of pelvic endometriosis was seen in the index patient during surgery. Very few of similar cases had been reported [15].

The differential diagnoses of umbilical endometriosis include granuloma, umbilical polyps, haemangioma, melanocytic nevus, seborrhoeic keratosis, granular cell tumour, umbilical hernia, lipoma, keloid, hypertrophic scars, and cutaneous metastasis of cancers. However, the history of cyclical pain and swelling as in the index patient may not be present in the other lesions [16].

The diagnosis of umbilical endometriosis could be made based on clinical findings but histological confirmation is the gold standard for diagnosis [16]. Other investigations that could be helpful in the diagnosis include ultrasonography, computed tomography, magnetic resonance imaging, and Doppler ultrasonography [17].

The definitive treatment for umbilical endometriosis is surgical excision. Medical treatment with analgesics and hormonal therapy may be used to achieve temporary relief of the symptoms and is not curative [10]. Some of the drugs that could be used include non-steroidal anti-inflammatory drugs, combined oral contraceptive pills, danazol, gestrinone, gonadotrophin releasing hormone agonists and antagonists. The index patient had abdominal myometrectomy and excision of the umbilical lesion with good outcome.

4. Conclusion

Although rare, primary umbilical endometriosis may coexist with uterine fibroids and should be suspected in women of reproductive age who complain of cyclical umbilical disorders in addition to abdominal swelling or other symptoms of uterine fibroids. The treatment of choice is surgical excision which could be done at the time of myomectomy as was done for our patient.

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Consent

Consent was obtained from the patient for the publication of her case.

Ethical approval

Ethical approval was obtained from Ethic and Research Committee of the Enugu State University of Science and Technology Teaching Hospital, Parklane Enugu with reference number: ESUTH/CMAC/RA/034/Vol.3/10 and dated 31st March 2022.

Author contribution

• Mba SG: Conceptualization of study, Study design, Data interpretation, Revising the manuscript critically for important intellectual content, Final approval of the version to be submitted.

• Omeke CA: Conceptualization of study, Study design, Data interpretation, Drafting of the manuscript, Final approval of the version to be submitted.

• Enebe JT: Study design, data interpretation, Revising the manuscript critically for important intellectual content, Final approval of the version to be submitted.

• Anyanwu OC: Data Collection, Data interpretation

Registration of research studies

Not applicable.

Guarantor

There was no external guarantor.

Declaration of competing interest

There was no conflict of interest to declare.

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