Primary umbilical endometriosis: a case report

Muluka L, Omanwa K

Department of Obstetrics and Gynaecology, University of Nairobi, Kenya

Correspondence to: Dr. L. Muluka. Email: mulukatila@gmail.com

Abstract

Umbilical endometriosis is a rare presentation especially in the absence of prior pelvic surgery. This report presents a rare case of symptomatic primary umbilical endometriosis in a 28 year old female who presented with a 2 year history of umbilical mass associated with cyclical bleeding at the time of her menses. There was no previous history of abdominal or pelvic surgery and no history of endometriosis associated symptoms. An excisional biopsy was performed with histology confirming the diagnosis of endometriosis.

Introduction

Endometriosis is defined by the presence of endometrial tissue outside the uterus (1). The precise prevalence of umbilical endometriosis is not known. Primary umbilical endometriosis however is a rare disorder. The incidence of this disease is estimated to be about 0.5% to 1% of all cases of extragenital endometriosis (2).

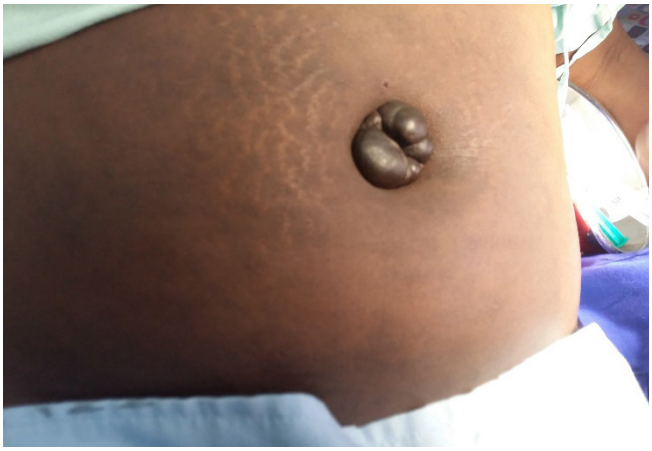
The most common symptom of primary umbilical endometriosis is cyclical pain and bleeding that is associated with an umbilical tumour. Although the pathogenesis of this disease is not fully understood, possibilities include the migration of endometrial cells to the umbilicus through the abdominal cavity, the lymphatic system or through embryonic remnants in the umbilical fold such as the urachus and the umbilical vessels. In contrast, secondary umbilical endometriosis is caused by iatrogenic dissemination of the eutopic endometrial cells following surgery. The iatrogenic implantation occurs in the surgical scars including caesarean section, laparoscopic surgery, and episiotomy(3). The patient considered here has a rare case of primary umbilical endometriosis.

Case presentation

A 28 year old, Para 2+0 female presented with history of an umbilical nodule of 2 years duration. This was associated with cyclical bleeding and pain coinciding with her menses. The nodule progressively increased in size over time. The patient reported no other symptoms such as dysmenorrhea, menorrhagia, dyspareunia, dyschezia or pelvic pain. She also had normal bowel movements and did not report nausea or vomiting. She had a regular 28 day cycle with average flow and had two children both delivered vaginally with no past history of surgery.

Physical examination revealed a 3x2cm dark brown coloured, lobulated, tender, firm and irreducible nodule of about 3 cm in diameter in the center of a widened umbilicus (Figure 1). Pelvic examination was otherwise normal.

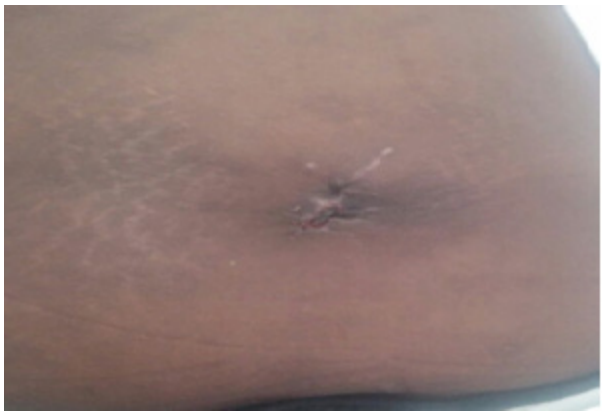
Figure 1: Lobulated umbilical endometrioma pre operatively



An abdominopelvic ultrasound done was normal. A provisional diagnosis of umbilical endometriosis was made and the patient worked up and scheduled for excisional biopsy under general anaesthesia. The diagnosis of endometriosis was confirmed histopathologically with sections showing skin tissue with the deep dermis bearing endometrial glands and stroma, smooth muscles and focal haemorrhagic areas. There was no atypia or malignant changes noted.

A 2 week and 3 month follow up was uneventful with the patient remaining symptom free. Figure 2 shows the wound 2 weeks after excision.

Figure 2: Surgical wound 2 weeks post operatively



Discussion

Extrapelvic endometriosis accounts for up to 15% of all cases of endometriosis. Umbilical endometriosis in particular, has been reported to be around 0.4–4% of all patients with endometriosis and accounts for upto 30–40% of all cases of cutaneous endometriosis (3).

The particularity of the case described herein, is that the patient had primary or spontaneous umbilical endometriosis, i.e. the presence of ectopic endometrial tissue located in the umbilicus in absence of previous surgery for either gynaecological disorders or caesarean section. The pathogenesis of primary endometriosis still remains unclear. Theories proposed are coelomic metaplasia, congenital presence of developmentally displaced endometrial tissue, direct extension through the round ligament or the patent omphalo-mesenteric duct, mechanical seeding of endometrial tissues via the lymphatic or venous system. It is possible that the umbilicus acts as a physiological scar with a predilection for endometrial tissue (4). As for clinical presentation, the typical symptoms of umbilical endometriosis are the presence of a discrete bluish-purple mass in the umbilicus, becoming swollen, painful and bleeding concomitantly with the menstrual cycle (3).

In the review of Boesgaard-Kjer *et al* (4) the mean age of the women was 28.5 years which corresponds to our patient's age. He observed that in young women with periodic complaints with bleeding from a tender and discolored umbilicus, endometriosis should always be considered. Differential diagnosis would include umbilical hernia, nodular melanoma, primary and metastatic neoplasms, granulomas, keloid and embryogenic remnants (4).

Surgical excision remains the treatment of choice. However other methods of treatment identified in literature include the use of combined oral contraceptive pill and gonadotropin releasing hormone agonist (4,5).

Conclusion

Although a rare condition, primary umbilical endometriosis should be suspected in a woman in her reproductive age with cyclical pain and bleeding from an umbilical tumour without prior history of surgery. Surgical excision remains the mainstay of treatment with good outcomes reported.

Funding

We would like to declare that the authors did not have any conflict of interest.

References

1. Dunselman GAJ, Vermeulen N, Becker C, Calhaz-Jorge C, D'Hooghe T, De Bie B, *et al.* ESHRE guideline: management of women with endometriosis. *Hum Reprod*. 2014; **29**(3): 400–412.
2. Krumbholz A, Frank U, Norgauer J and Ziemer M. Umbilical endometriosis. *JDDG*. 2006; **4**(3): 239–241.
3. Fancellu A, Pinna A, Manca A, Capobianco G and Porcu A. Primary umbilical endometriosis. Case report and discussion on management options. *Int J Surg Case Rep.* 2013; **4**(12):1145–8.
4. Boesgaard-Kjer D, Boesgaard-Kjer D and Kjer JJ. Primary umbilical endometriosis (PUE). *Eur J Obstet Gynecol Reprod Biol* [Internet]. 2016 Jun [cited 2016 Aug 22]; Available from: http://linkinghub.elsevier.com/retrieve/pii/S0301211516302445
5. Calagna G, Perino A, Chianetta D, Vinti D, Triolo MM, Rimi C, *et al*. Primary umbilical endometrioma: Analyzing the pathogenesis of endometriosis from an unusual localization. *Taiwan J Obstet Gynecol*. 2015; **54**(3):306–312.