CEOG Clinical and Experimental Obstetrics & Gynecology

Umbilical endometriosis: a rare case of spontaneous cutaneous umbilical endometriosis

M. Kalinderis, U. Singh

Department of Obstetrics & Gynaecology, Darent Valley Hospital, Dartford, Kent (United Kingdom)

Summary

Umbilical endometriosis is an extremely rare type of endometriosis, with an incidence of 0.5% to 1% of all endometriosis cases. This report describes a case of umbilical endometriosis that developed in the absence of previous abdominal or uterine surgery. Clinical diagnosis of umbilical endometriosis is difficult and the differential diagnosis of umbilical lesions can be confusing, thus a high level of clinical suspicion is required.

Key words: Villar's nodule; Endometriosis; Umbilical lesion.

Introduction

Endometriosis is defined by the presence of endometrial glands and stroma outside the endometrial cavity. It is a common condition that affects about 10% of women of reproductive age and 35–50% of women with pelvic pain and infertility [1]; however its cause remains currently poorly understood. The classic triad of symptoms dysmenorrhea, dyspareunia, and dyschezia is highly suggestive of endometriosis. Endometriosis is an estrogen-dependent disease that can be pharmacologically or/and surgically treated [2].

Endometriosis is dominantly found in the pelvis, but can also develop outside the uterus or ovary and is called ectopic or extra-gonadal/extragenital endometriosis. Extragonadal endometriosis, although rarely described, its seed can be found almost throughout the female body including bowel, bladder, lungs, brain, umbilicus, and surgical scars [3]. This is a case report study describing a cutaneous umbilical endometriosis in a surgically naïve young woman.

Case Report

A 41-year-old Asian woman initially presented to the surgical department complaining of umbilical non-reducible painful swelling, associated with purulent and bloody discharge over an 18-month period. She was previously treated with meticulous hygiene and courses of antibiotics without any improvement.

After clinical examination, the surgeon suspected an umbilical hernia. Under general anesthesia, exploration of umbilicus was offered and dissection down to the sheath and around the umbilicus was performed. A small hernia was repaired and the affected area was excised and was sent for histological examination. The abdominal defect was repaired with Vicryl one on J-shape needle in two layers and the umbilicus was reconstructed. The postoperative period was uneventful and the patient was discharged home the following day. After the histology results that reported fibroadipose tissue, with endometriosis associated with haemorrhage and chronic inflammation, the patient was referred to gynaecology clinic complaining of recurrent discharge from the umbilicus.

The patient had two uncomplicated vaginal deliveries with no history of subfertility. Her menstrual cycles were regular, however associated with excruciating painful and heavy periods. She also reported deep dyspareunia. She did not report any intermenstrual or postcoital bleeding and she was up to date with the smears. She did not provide history of urinary or bowel symptoms. The past medical history was unremarkable and she was surgically naïve before the excision of the granulomatous lesion.

From detailed history umbilical bleeding was reported to increase during menstruation. On physical examination abdomen was soft, not tender. Gynaecological examination revealed an anteverted, non-tender, mobile, and normal-sized uterus. Adnexal masses were not palpable on either side and no nodules were felt in the uterosacral ligaments or in the rectovaginal space. Gynaecological ultrasound scan was performed in which the anteverted uterus appeared normal in size and shape. The myometrium appeared heterogeneous and contained a small myometrium cyst; the appearance was suggestive of adenomyosis. The endometrium was regular in outline and within normal limits measuring 8.3 mm. Both ovaries appear sonographically normal. The right ovary measured 3×2.8×2.6 cm and contained a 2-cm normal dominant follicle. The left ovary measured 2.4×3×2.8 cm. No adnexal cyst or mass was seen, as well as no free fluid.

Discussion

Umbilical endometriosis is a rare manifestation of endometriosis, with an estimated incidence of 0.5–1% of all endometriosis cases and up to 30–40 % of cases with cutaneous endometriosis, only 15% of which is associated with

Revised manuscript accepted for publication July 18, 2016

Clin. Exp. Obstet. Gynecol. - ISSN: 0390-6663
XLIV, n. 5, 2017
doi: 10.12891/ceog3839.2017

7847050 Canada Inc.
www.irog.net

pelvic endometriosis [4]. Umbilical endometriosis is also known as Villar's nodule as is attributed to a physician who first described the disease in 1886 [5]. From 1996 to 2007, Victory *et al.* found only 122 reported cases of umbilical endometriosis worldwide [6]. It can be divided into primary and secondary. Primary umbilical endometriosis arises de novo. Spontaneous or primary umbilical endometriosis, occurring without any previous abdominal or uterine surgery, is extremely rare. The development of secondary umbilical endometriosis more often appears after a surgery, especially laparoscopic surgical procedures involving the umbilicus [4]. The present patient presented with a negative surgical history, which rendered the pathophysiology of the disease even more enigmatic.

Multiple theories have been proposed as causes of endometriosis. Sampson in 1927 was the first to propose the retrograde menstruation or implantation theory suggesting that during menstruation blood refluxes through the fallopian tubes and endometrial tissue implants into the nearby organs. However, this theory has been disputed in the past since retrograde menstruation occurs in 76–90% of women with patent fallopian tubes and only a minority will develop the disease; this theory cannot also explain the occurrence of endometriosis in pre-pubertal girls, newborns or males. A direct transportation of endometrial cells *via* blood or lymph vessels or even through surgical manipulations has also been proposed [1]. According to the theory of metaplasia, endometriosis originates from extrauterine cells that abnormally transform into endometrial cells. The Coelomic metaplasia theory proposes metaplasia of peritoneal mesothelial tissue cells into endometrial cells. Hormones, especially estrogen, are thought to stimulate this transformation. The embryonic rest theory proposes that the presence of cells of Müllerian origin within the peritoneal cavity could be induced to form endometrial tissue when subjected to the appropriate stimuli [1]. Inflammation and oxidative stress may contribute to the pathogenesis of endometriosis, as well as apoptosis suppression and survival of endometrial cells, immunological dysfunction, and genetic predisposition. Moreover specific attention has been paid to the role of stem cells through endometrial self-generation in specific niches of the endometrium [6].

In the development of spontaneous umbilical endometriosis, as in the case presented, some etiologies have been proposed, but none of them can entirely explain the appearance of the tumor. It is possible that the umbilicus, considered a physiologic scar, could have tropism for endometrial cells [7]. It has been also suggested that isolated umbilical endometriosis may arise through metaplasia of urachal remnants [8]. The theory of lymphatic or vascular transportation and reimplantation at the umbilicus could also be possible.

Umbilical endometriosis is commonly found in the reproductive age group. The mean age of diagnosis has been reported to be 37.7 years with the youngest being 23 years [5]. Typical symptoms involve cyclical swelling, pain, and bleeding from umbilicus, however there are case reports were the main symptom is not bleeding [9, 10]. Clinically, it presents as a reddish-brown painful nodule with cyclic variations in size with or without bleeding that many coincide or not with patient's menstrual cycle. Its size ranges from 0.5-3 cm, but larger masses have been described, as well [11]. The diagnosis is primarily clinical, but often puzzling and misguiding. Umbilical endometriosis can be misdiagnosed in 20-50% of the cases. The differential diagnoses of umbilicus endometriosis should include mainly melanoma, umbilical metastases (Mary Sister Joseph Nodule), and pyogenic granuloma. Lipoma, abscess, omphalitis, cyst, hernia, various granulomas, urachal lesions, nodular melanoma, primary or metastatic carcinoma, and keloid and residual embryonic tissue should be considered in the differential diagnosis, as well [8]. A biopsy is mandatory especially when melanoma cannot be ruled out. In a case of umbilical endometriosis malignization can occur, however the risk of malignancy is reassuringly low. Dermoscopy may be used as an auxiliary tool, however the diagnosis is often made incidentally by histological examination after surgical exploration and excision of the lesion.

Umbilical endometriosis treatment involves medical management such as progestational drugs, danazol, and GnRH analogues or surgical excision. Medical treatment could potentially be a pre-treatment option especially in cases of large tumours. However, surgical excision with complete wide resection of the lesion with free margins and minimal spillage is considered as a first line treatment. If the umbilicus cannot be preserved then reconstruction is recommended in conjunction with plastic surgeons. Mesh is required in cases of abdominal wall defects, however the possibility of mesh contamination by endometriosis needs further evaluation. Follow up following excision is paramount and is recommended every six months for a period of two years. The patient should be informed about the risk of malignancy and local recurrence.

Conclusion

Cutaneous endometriosis is a rare skin pathology that may present to the gynaecologist, general surgeon, dermatologist or plastic surgeon. This report describes a case of umbilical endometriosis that developed in the absence of previous abdominal or uterine surgery. Clinical diagnosis is difficult and umbilical endometriosis may go unrecognized because of its rarity, leading to multiple medical visits and a delayed diagnosis. A definite diagnosis can only be established by histopathological examination. Its pathogenesis remains currently uncertain. Since pelvic endometriosis may be present, referral to a gynaecologist is recommended in every case. Increased clinical surveillance and suspicion is required for the diagnosis of endometriosis, a multifaceted

and enigmatic disease with variable appearance.

Acknowledgements

The authors thank Dr. Kallirhoe Kalinderi for the critical reviewing of the manuscript

References

[1] Vercellini P., Viganò P., Somigliana E., Fedele L.: "Endometriosis: pathogenesis and treatment". *Nat. Rev. Endocrinol.,* 2014, *10*, 261.
[2] Kodaman P.H.: "Current strategies or endometriosis management". *Obstet. Gynecol. Clin. North Am.,* 2015, *42*, 87.
[3] Agarwal N., Subramanian A.: "Endometriosis morphology, clinical presentations and molecular pathology". *J. Lab. Physicians*, 2010, 2, 1.
[4] Pramanik S.R., Mondal S., Paul S., Joycerani D.: "Primary umbilical endometriosis: a rarity". *J. Hum. Reprod. Sci.,* 2014, *7*, 269.
[5] Victory R., Diamond M.P., Johns D.A.: "Villar's nodule: a case report and systematic literature review of endometriosis externa of the umbilicus". *J. Minim. Invasive Gynecol.,* 2007, *14*, 23.
[6] Sourial S., Tempest N., Hapangama D.K.: "Theories on the pathogenesis of endometriosis". *Int. J. Reprod. Med.,* 2014, *2014*, 179515.
[7] Attia L., Ben Temime R., Sidhom J., Sahli A., Makhlouf T., Chachia A., *et al.*: "A case of cutaneous endometriosis development on an abdominal scar". *Tunis Med.,* 2010, *88*, 841.
[8] Frischknecht F., Raio L., Fleischmann A., Dreher E., Luscher K.P., Mueller M.D.: "Umbilical endometriosis". *Surg. Endosc.,* 2004, *18*, 345.
[9] Chatzikokkinou P., Thorfinn J., Angelidis I.K., Papa G., Trevisan G.: "Spontaneous endometriosis in an umbilical skin lesion". *Acta Dermatovenerol. Alp. Panonica Adriat.,* 2009, *18*, 126.
[10] Dessy L.A., Buccheri E.M., Chiummariello S., Gagliardi D.N., Onesti M.G.: "Umbilical endometriosis, our experience". *In Vivo,* 2008, *22*, 811.
[11] Latcher J.W.: "Endometriosis of the umbilicus". *Am. J. Obstet. Gynecol.,* 1953, *66*, 161.

Corresponding Author:
M. KALINDERIS M.D., PhD
Department of Obstetrics & Gynaecology
Darent Valley Hospital
Darent Wood Road
Dartford, Kent DA2 8DA (United Kingdom)
e-mail: m.kalinderis@hotmail.com