

Recurrence of primary umbilical endometriosis: case report and review of the literature

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Summary

The authors report the case of a 33-year-old nulliparous patient with recurrence of primary spontaneous cutaneous umbilical endometriosis and no history of abdominal or pelvic surgery. The literature describing similar cases is reviewed.

Primary endometriosis at extrapelvic sites, and umbilical endometriosis in particular, is very uncommon. The diagnosis is confirmed only by histopathological examination, specifically hematoxylin and eosin staining. Conservative surgical excision with wide margins is the recommended treatment. Adjuvant and neoadjuvant treatment for endometriosis is based on medications which induce endometrial atrophy, such as gonadotropin releasing hormone superactive analogs, or oral estrogen-progestogen formulations given continuously.

KEY WORDS: *umbilical endometriosis, macroscopic appearance, pathogenesis, surgical excision, Villar's nodule.*

Introduction

Endometriosis is defined as the presence of endometrial tissue, with its typical stromal and glandular components, at anatomical sites other than the uterine cavity. The ovaries are most commonly involved, followed by the

uterine ligaments, pelvic wall, intestine, bladder and skin. Cutaneous endometriosis of the umbilical scar, also known as Villar's nodule, is rare, especially if there is no history of abdominal or pelvic surgery or concomitant endometriosis at other sites.

Villar's nodule, which accounts for up to 5% of cases of cutaneous endometriosis, can occur as a primary lesion resulting from spontaneous metastatic implantation, or as secondary endometriosis, i.e. following a iatrogenic displacement of endometrial tissue during surgical procedures such as umbilical or inguinal hernia repair or any laparoscopic surgery performed using an umbilical access (2,3). On physical examination, the lesion appears as a solid nodule varying in color from red to blue or brown-black, depending on the amount of hemorrhage and the depth of penetration of the ectopic endometrial tissue (1). A case of recurrent primary cutaneous endometriosis of the umbilical scar is here reported.

Case report

In February 2013, a 33-year-old nulligravida black African woman referred herself to the Gynecological Surgery Unit at the "Ss Annunziata" Hospital in Chieti (Italy) with a tender and discolored nodule of the umbilical skin. Characteristically, the lesion showed cyclic hematic discharge associated with evident changes in size and color concomitantly with the menstrual period.

Subjective pain was described by the patient as constant and not related to any particular activity, but markedly increased by the onset of the menstrual period.

Her past history included surgical excision of a lesion with identical clinical, anatomical and histopathological features, performed in 2006. The nodule was excised with free margins and pathology examination confirmed the lesion nature as cutaneous endometriosis. The patient's history was negative for any other surgery on the umbilical skin, including cosmetic procedures such as piercings, and she reported no previous pelvic or abdominal surgery. Her history was also negative for significant medical problems, with the exception of dysmenorrhea, and her family history was negative for significant related pathology.

The patient's body mass index, blood cell count, liver function test and renal function test were all in the normal range. Her serum CA125 was 77.2 U/ml the day before the procedure (cut-off 35 U/ml).

A head-to-toe physical examination showed two reddish-blue nodules with a solid consistency involving the umbilical skin, moderately tender on palpation (Fig. 1). No other lesions were identified on thorough inspection of other cutaneous regions. No pathologically enlarged lymph nodes were evident on palpation.



Figure 1. Umbilical endometriosis that presented as a two reddish-blue nodules and occupied the deep part of the umbilical circumference.

The nodule was excised with wide margins under local anesthesia and the specimen was sent for histopathological examination. The macroscopic report described an elliptical specimen of skin including two nodules, a lateral one with a maximum diameter of 12 mm and a more central one, deeply embedded in the umbilicus, with a maximum diameter of 9 mm. Neither nodule showed superficial hematic scabs. Light microscopy with hematoxylin and eosin (H&E) staining showed a typical area of endometriosis consisting of endometrial-type glands and stroma. Subsequent histological examination showed evidence of stratified endometrial glands and stroma.

The post-operative course was uneventful and the patient was given continuous oral estrogen-progestins for six months to induce atrophy of other possible microscopic foci of ectopic endometrium.

Discussion

Overall, the estimated incidence of umbilical endometriosis is 0.5-1% (4,5), and it has a highly variable clinical presentation.

Cutaneous endometriosis affects patients with a mean age of 35-38 years (6,7) and most commonly involves scars.

Less than 30% of cutaneous endometriosis occurs in the absence of previous surgery (3-6); in such cases, the site most commonly affected is the umbilicus, followed by the inguinal area and the abdominal wall (8).

The clinical features of the lesion described in different studies, e.g. color, consistency and tenderness on palpation, vary widely, depending on the depth of penetration of the ectopic endometrial tissue and the amount of bleeding (1). Its color reportedly varies from pink to dark brown and black, while palpation commonly discloses a nodule with a solid consistency and moderate to severe tenderness. The nodule can be single or multilobed (9).

Menstrual bleeding into the dermis leads to hemosiderin deposition, scarring and chronic inflammation.

The pathogenesis of spontaneous umbilical en-

ometriosis is still not well understood. Several hypotheses have been proposed, each one explaining some specific characteristic of different clinical presentations of endometriosis. Most of the theories lay the development of ectopic endometrial foci either on a primitive lesion of the involved site, arising from tissue metaplasia or from embryonic remnants (the *in situ* theory), or on the endometrial transplantation from i) retrograde menstruation; ii) iatrogenic displacement by surgical procedures; iii) lymphatic or vascular benign metastatic transport (the implantation theory) (10).

In contrast to normal, eutopic endometrial tissue, endometriotic tissue shows, *in vitro*, the capability, by itself, to produce estrogens through the aromatase cycle.

Genetic (11, 12), hormonal (13) and autoimmune factors (14) also play an important role in the pathogenesis of endometriosis.

The diagnosis can be suspected on the basis of clinical signs and symptoms collected through a detailed history and physical examination, but it must always be confirmed pathologically through H&E staining. The histological image will typically show irregular glands lined by columnar epithelium among a myxoid stroma with extravasation of blood into the gland lumina.

In the event of doubt, immunocytochemistry stains such as vimentin and CD10 are used to confirm the presence of endometrial-type stroma around endometrial-type glands (15).

The treatment of choice is conservative surgical excision of the lesion, with sufficient healthy margins to prevent recurrence, with a subsequent course of medication effective in inducing endometrial atrophy (16).

It is recommended that surgical excision be performed at the end of the menstrual period, when the nodule is smaller and a smaller excision can therefore be performed (18).

Postoperative medications to induce endometrial atrophy include continuous oral contraceptives, i.e. given without the one-week cyclical suspension, or gonadotropin releasing hormone superactive analogs. Both medications effectively inhibit ovarian function and therefore the cyclical stimulation of the endometrium (16-18), while the prevalently progestogenic environment induced by oral estrogen-progestins has a direct atrophying effect on the endometrium.

Although endometriosis may recur after the course of medication is stopped, the prognosis is usually from good to excellent, with little or no symptoms at all remaining after proper treatment. Relapses are uncommon if excision is performed with clean and wide margins (1).

In conclusion, primary endometriosis is a rare condition whose diagnosis may be sometimes challenging. It can be suspected clinically from the patient's history, but should be confirmed with histological examination.

References

1. Kyamidis K, Lora V, Kanitakis J. Spontaneous cutaneous umbilical endometriosis: report of a new case with immunohistochemical study and literature review. *Dermatol Online J* 2011; 17(7):5.

2. Douglas C, Rotimi O. Extragenital endometriosis: a clinico-pathological review of a Glasgow hospital experience with case illustrations. *J Obstet Gynaecol* 2004; 24(7):804-808.
3. Agarwal A, Fong YF. Cutaneous endometriosis. *Singapore Med J* 2008; 49(9):704-709.
4. Rosina P, Pugliarello S, Colato C, Girolomoni G. Endometriosis of umbilical cicatrix: case report and review of the literature. *Acta Dermatovenerol Croat* 2008; 16(4):218-221.
5. Krumbholz A, Frank U, Norgauer J, Ziemer M. Umbilical endometriosis. *J Dtsch Dermatol Ges* 2006; 4(3):239-241.
6. Fernández-Aceñero MJ, Córdova S. Cutaneous endometriosis: review of 15 cases diagnosed at a single institution. *Arch Gynecol Obstet* 2011; 283(5):1041-1044.
7. Victory R, Diamond D, Johns DA. Villar's nodule: a case report and systematic literature review of endometriosis externa of the umbilicus. *J Minim Invasive Gyneol* 2007; 14(1):23-32.
8. Steck WD, Helwing EB. Cutaneous endometriosis. *Clin Obstet Gynecol* 1996; 9(2):373-383.
9. Weller CV. Endometriosis of the umbilicus. *Am J Pathol* 1935; 11(2):281-286.
10. van der Linden P. Theories on the pathogenesis of endometriosis. *Hum Reprod* 1996; 11 Suppl 3:53-65.
11. Simpson JL, Bischoff FZ, Kamat A, Buster JE, Carson SA. Genetics of endometriosis. *Obstet Gynecol Clin North Am* 2003; 30(1):21-40.
12. Vignano P, Somigliana E, Vignali M, Busacca M, Blasio AM. Genetics of endometriosis: current status and prospects. *Front Biosci* 2007; 12:3247-3255.
13. Sinaii N, Cleary SD, Ballweg ML, Nieman LK, Stratton P. High rates of autoimmune and endocrine disorders, fibromyalgia, chronic fatigue syndrome and atopic diseases among woman with endometriosis: a survey analysis. *Hum Reprod* 2002; 17(10):2715-2724.
14. Mathur A, Peress MR, Williamson HO, Youmans CD, Maney SA, Garvin AJ et al. Autoimmunity to endometrium and ovary in endometriosis. *Clin Exp Immunol* 1982; 50(2):259-266.
15. Potlog-Nahari C, Feldman AL, Stratton P, Koziol DE, Segars J, Merino MJ, Nieman LK. CD10 immunohistochemical staining enhances the histological detection of endometriosis. *Fertil Steril* 2004; 82(1):86-92.
16. Minaidou E, Polymeris A, Vassiliou J, Kondi-Paphiti A, Karoutsou E, Katafygiotis P, Papaspyrou E. Primary umbilical endometriosis: case report and literature review. *Clin Exp Obst Gynecol* 2012; 39(4):562-564.
17. Chatzikokkinou P, Thorfinn J, Angelidis IK, Papa G, Trevisan G. Spontaneous endometriosis in an umbilical skin lesion. *Acta Dermatovenerol Alp Panonica Adriat* 2009; 18(3):126-130.
18. Bagade PV, Guirguis MM. Menstruating from the umbilicus as a rare case of primary umbilical endometriosis: a case report. *J Med Case Rep* 2009; 3:9326.