

Primary umbilical endometriosis: case report and literature review

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Summary

We present a case of primary endometriosis of the umbilicus in a young nulliparous patient without any previous history of abdominal or pelvic surgery. Primary endometriosis of extra pelvic sites is unusual while umbilical endometriosis is quite rare. Diagnosis of endometriosis is difficult to obtain and sometimes diagnoses can be false-positive or false-negative. Some imaging procedures can be done to rule out other disorders but it is difficult to differentiate them from endometriosis. A definite diagnosis can only be established by histopathological examination. Hematoxylin and eosin (H&E) is the staining of choice. Conservative surgical excision of the lesion and drugs such as oral contraceptives and gonadotropin releasing analogues are the first-line treatment.

Key words: Endometriosis; Umbilical endometriosis; Umbilical inflammation; Hematoxylin and eosin (H&E) staining; CD10 staining; Vimentin staining.

Introduction

Endometriosis is defined as the presence of endometrial tissue, stroma and glands outside the uterine cavity. Common sites of endometriosis are the ovaries, fallopian tube, uterine ligament, pelvic wall, intestine and bladder.

Primary endometriosis of extra pelvic sites is unusual. Endometriosis of the umbilicus [1] and umbilical hernia sac is rare [2], especially if there is no history of abdominal, pelvic surgery or known preexisting endometriosis in other sites. The estimated incidence of umbilical endometriosis is 0.5-1% [3, 4].

Secondary endometriosis, (scar endometriosis) can develop after an umbilical hernia repair [5], an inguinal hernia repair [6] and laparoscopy performed through the umbilicus [7] or abdomen [8].

Cutaneous endometriosis could be an indicator of pre-existing abdominal endometriosis without any previous manifestation [9].

Case report

We present a case of a 26-year-old nulliparous patient without any history of previous abdominal or pelvic surgery. Over the past few years she had dysmenorrhea and was seen by a gynecologist. After an abdominal ultra-sound a cyst of 4.6 x 3.9 cm was found in one of the ovaries and a hemorrhagic corpus luteum cyst was suspected as a diagnosis. The gynecologists prescribed an anti-inflammatory drug, lornoxicam, and followed her up over time. The cyst was progressively decreasing in size and finally regressed but the dysmenorrhea remained.

During the past year the patient complained of umbilical pain and noticed a dark purplish nodule on the umbilicus with size changes – growth alternating with decreased size continuously over time.

No bloody secretions were observed.

The patient visited a dermatologist whose diagnosis was inflammation of the umbilicus. A local antibiotic cream was given with no result. After that she visited a surgeon who diagnosed an umbilical hernia and an operation was planned. Upon medical history a correlation was made between the painful nodule and menstrual cycle.

The patient realized that the enlargement together with the pain on the umbilicus was cyclic and was happening only a few days per month.

The operation for the umbilical hernia was performed and the specimen was sent for histopathological evaluation with the indication “umbilical nodule”.

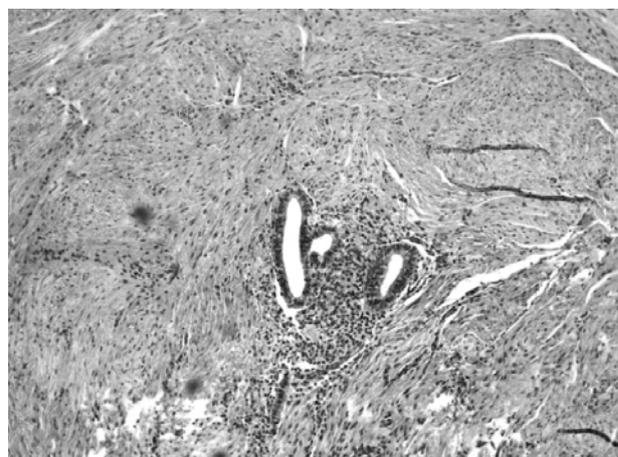


Figure 1. — Hematoxylin and eosin (H&E) staining of endometriotic tissue.

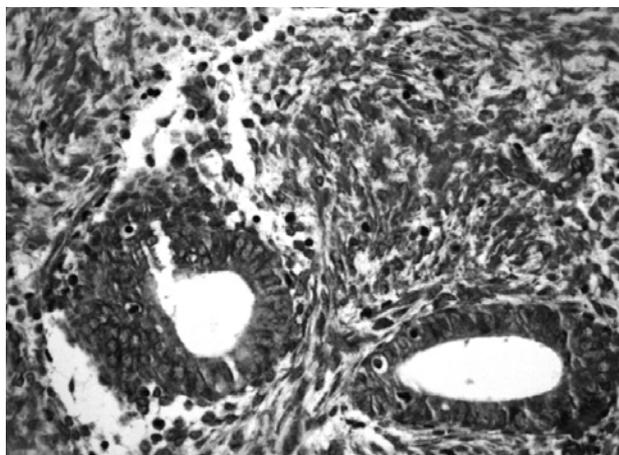


Figure 2. — Vimentin staining of endometriotic tissue.

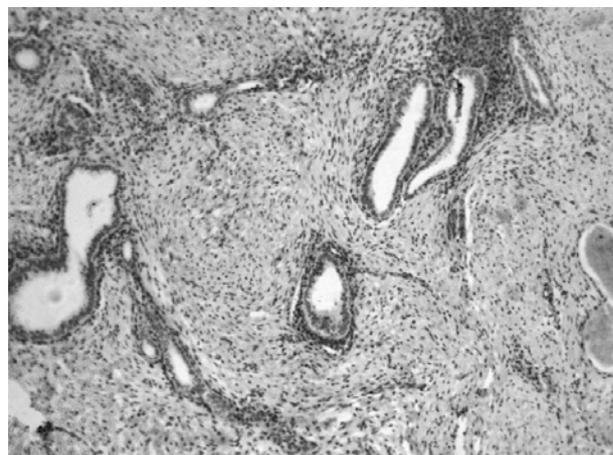


Figure 3. — CD10 staining of endometriotic tissue.

The specimen consisted of an ellipse of skin measuring 1.3 x 1 cm with underlying soft tissues that contained a circumscribed nodule 1 cm in diameter. It was routinely processed in paraffin and was stained by hematoxylin and eosin (H&E) staining (Figure 1).

Microscopically, there was a typical area of endometriosis consisting of endometrial-type glands and stroma.

Additional immunohistological stains for vimentin (Figure 2) and CD10 (Figure 3) were performed to support the histological finding of endometriosis.

Discussion

Primary endometriosis of the umbilical hernia sac is rare, especially if no previous abdominal or pelvic surgery has been performed, and only a few cases have been reported over the last decade [10-12].

The pathogenesis of extra pelvic primary endometriosis can be explained by the “retrograde theory”, according to which endometrial tissue can spread through the fallopian tube during menstruation. Retrograde menstruation occurs normally in healthy women but only a very small percent develop primary endometriosis [13].

In contrast to normal endometrial tissue, endometriotic tissue differentiates in the fact that it can produce estrogens by itself through the aromatase cycle [14] in vitro.

Genetic [15, 16], hormonal [17] and autoimmune factors [18] also play an important role in the pathogenesis of endometriosis.

According to the American Society for Reproductive Medicine, endometriosis may be classified based on the number, location and depth of endometriotic tissue and the presence of adhesion in four stages [Stage I (minimal), Stage II (mild), Stage III (moderate), Stage IV (severe)].

Imaging procedures such as ultrasonography, computed tomography and magnetic resonance imaging are done to rule out other disorders. Sometimes they are useful for determining the location and the size of endometriosis but cannot differentiate endometriosis from soft tissue tumors

[19]. On the other hand they can be useful in monitoring the disorder after it is diagnosed.

Histological examination is the only exam for a certain diagnosis. The diagnosis is usually straight forward with H&E stains, but false-negative results can be produced.

In a few cases immunocytochemistry stains, such as vimentin and CD10, are used to confirm the presence of endometrial type stroma around endometrial type glands [20].

The treatment of choice is conservative surgical excision of the lesion, with sufficient healthy margins to prevent recurrence, and drugs.

Drugs such as oral contraceptives and gonadotropin releasing hormone analogues inhibit the growth and activity of endometriotic tissue by suppressing ovarian function. However endometriosis recurs if the patient stops the drugs.

In conclusion, primary endometriosis is a rare entity and diagnosis can be difficult.

References

- [1] Iovino F, Ruggiero R., Irlandese E., Gili S., Lo Schiavo F: “Umbilical endometriosis associated with umbilical hernia. Management of a rare occurrence”. *Chir. Ital.*, 2007, 59, 895 (ISSN: 0009-4773).
- [2] Yuen J.S., Chow P.K., Koong H.N., Ho J.M., Girija R.: “Unusual sites (thorax and umbilical hernial sac) of endometriosis”. *J. R. Coll. Surg. Edinb.*, 2001, 46, 313 (ISSN: 0035-8835)
- [3] Rosina P., Pugliarello S., Colato C., Girolomoni G.: “Endometriosis of umbilical cicatrix: case report and review of the literature”. *Acta Dermat. Croat.*, 2008, 16, 218 (ISSN: 1330-027X)
- [4] Krumbholz A., Frank U., Norgauer J., Ziemer M.: “Umbilical endometriosis”. *J. Dtsch. Dermatol. Ges.*, 2006, 4, 239 (ISSN: 1610-0379).
- [5] Majeski J., Craggie J.: “Scar endometriosis developing after an umbilical hernia repair with mesh”. *South Med. J.*, 2004, 97, 532 (ISSN: 0038-4348).
- [6] Ducarme G., Uzan M., Poncelet C.: “Endometriosis mimicking hernia recurrence”. *Hernia*, 2007, 11, 175 (ISSN: 1265-4906).
- [7] Yu C.Y., Perez-Reyes M., Brown J.J., Borrello J.A.: “MR appearance of umbilical endometriosis”. *J. Comput. Assist. Tomogr.*, 1994, 18, 269 (ISSN: 0363-8715).
- [8] Sirito R., Puppo A., Centurioni M.G., Gustavino C.: “Incisional hernia on the 5-mm trocar port site and subsequent wall endometriosis on the same site: a case report”. *Am. J. Obstet. Gynecol.*, 2005, 193, 878 (ISSN: 0002-9378).

- [9] Elm M.K., Twede J.V., Turiansky G.W.: "Primary cutaneous endometriosis of the umbilicus: a case report". *Cutis*, 2008, 81, 124 (ISSN: 0011-4162).
- [10] Spaziani E., Di Filippo A., Picchio M. *et al.*: "Umbilical primary endometrioma. Case report". *G. Chir.*, 2009, 30, 230 (ISSN: 0391-9005).
- [11] Khaled A., Hammami H., Fazaa B., Zermani R., Ben Jilani, Kamoun M.R.: "Primary umbilical endometriosis: a rare variant of extragenital endometriosis". *Pathologica*, 2008, 100, 473 (ISSN: 0031-2983).
- [12] Wiegatz I., Kissler S., Engels K., Strey C., Kaufmann M.: "Umbilical endometriosis in pregnancy without previous surgery". *Fertil Steril.*, 2008, 90, 199 e17.
- [13] D. T. Y. Liu, A. Hitchcock: "Endometriosis: its association with retrograde menstruation, dysmenorrhoea and tubal pathology". *Int. J. Obstet. Gynaecol.*, 1986, 93, 859.
- [14] Jafar Ai, N. Esfandiari, R. Casper: "Detection of aromatase in human endometrial tissue cultured in three-dimensional fibrin matrix in vitro". *Iranian J. Reprod. Med.*, 105.
- [15] Simpson J.L., Bischoff F.Z., Kamat A., Buster J.E., Carson S.: "Genetics of endometriosis". *Obstet. Gynecol. Clin. North Am.*, 2003, 30, 21.
- [16] Vigano P., Somigliana E., Vignali M., Busacca M., Blasio A.M.: "Genetics of endometriosis: current status and prospects". *Front. Biosci.*, 2007, 12, 3247.
- [17] N. Sinaii, S.D. Cleary, M.L. Ballweg, L.K. Nieman, P. Stratton: "High rates of autoimmune and endocrine disorders, fibromyalgia, chronic fatigue syndrome and atopic diseases among women with endometriosis: a survey analysis". *Hum. Reprod.*, 2002, 17, 2715.
- [18] S. Mathur, M.R. Peress, H.O. Williamson, C.D. Youmans, S.A. Maney, A.J. Garvin *et al.*: "Autoimmunity to endometrium and ovary in endometriosis". *Clin. Exp. Immunol.*, 1982, 50, 259.
- [19] A. Marinis, J. Vassiliou, D. Kannas, T.K. Theodosopoulos, A. Kondi-Pafiti, E. Kairi, V. Smymiotis: "Endometriosis mimicking soft tissue tumors: diagnosis and treatment". *Eur. J. Gynaec. Oncol.*, ISSN 0392-2936, XXVII, n. 2, 2006.
- [20] Potlog-Nahari C., Feldman A.L., Stratton P., Koziol D.E., Segars J., Merino M.J., Nieman L.K.: "CD10 immunohistochemical staining enhances the histological detection of endometriosis". *Fertility and sterility*, 2004, 82.

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