Bangladesh J Obstet Gynaecol, 2024; Vol. 39(1): 49-52

Umbilical Endometriosis: A Case Report

RABEYA AKTHER

Background:

Endometriosis is defined as the presence of the endometrium other than the uterine cavity. It is also considered a chronic inflammatory disease associated with immune processes.[1] This disease was first described by Daniel Shroen in 1690 in the work "*Disputatio Inauguralis Medica de Ulceribus Ulceri*". The symptoms of this disease were presented by Arthur Duff in 1769.[2] The ectopic endometrium is functionally similar to the eutopic endometrium. It is a benign, estrogen-dependent, gynecological disease; however, due to the accompanying ailments and chronic nature, it is a very important medical, social and economic problem. This disease affects from 10–15% of women of reproductive age and 35–50% of women with pelvic pain and/or infertility.[3] The vast majority of cases of endometriosis occur in women between menarche and menopause. The peak of the disease falls in the period between 25 and 45 years of age.[2] Endometrial foci outside the uterine cavity may appear, for example, in the peritoneal cavity, ovaries, bladder or ureters.[4] Extrapelvic endometriosis occurs less commonly. The extrapelvic sites include the diaphragm, pulmonary, urinary tract, gastrointestinal tract, brain, surgical scar and cutaneous endometriosis. Umbilicus is a physiological scar that is a preferred site for umbilical endometriosis, as described by Yu et al.[5] It represents 0.5% to 1% of all cases of extragenital endometriosis.

Primary umbilical endometriosis is first described by Villar in 1886; therefore, it is also known as Villar's nodule and since then more than 100cases have been described.[6,7] The pathogenesis of endometriosis is not well understood. Postulated theories include Sampson's theory of retrograde menstruation, which is the commonest, coelomic metaplasia, induction theory, embryonic Mullerian rests, bone marrow stem cell theory, and hematogenous/lymphatic spread. The theory favored in the case of umbilical endometriosis is hematogenous/lymphatic spread where there is co-existing pelvic endometriosis. Isolated umbilical endometriosis could develop from metaplasia of urachal remnants [2,7,8,9,10]

The aim of this study is to report one such case of umbilical endometriosis because of its rarity as well as the management option we experienced during the management of this patient.

Case report:

A 38-year-old woman presented with complaint of a swelling at her umbilical region which bleeds spontaneously. She was menstruating for last 3 days without any pain (picture 1). On examination, there was a dark bluish cystic swelling at umbilicus and dried clotted blood around the swelling. It was about 2cm x2cm size like a big black berry (Picture 2). She is married for 16 years, menstruating regularly with average flow and duration. Her husband work abroad and she never practices any contraceptive method. She had two children, one delivered vaginally and another by cesarean section 7 years back. For the last two cycles her menstrual flow was more than average. Her medical history was unremarkable. Her general and abdominal examination findings was unremarkable. Her hemoglobin level is 11.6gm/ dl; serum TSH level was 3.24mic IU/ml. Her day 12 ultrasonogram (USG) of pelvic organ was normal and usg of umbilical lesion slowed a solid area and measured 0.4 cm x0.4 cm. Our clinical diagnosis was umbilical endometriosis. Patient was offered both medical and surgical management and she opted to have oral contraceptive pill and Naproxen Sodium 500mg immediately. She again menstruates after 28 days, no umbilical bleeding and umbilical swelling was shrunken. (Picture 3).She was followed regularly and counseled to continue OCP and Naproxen 500mg during menstrual period to reduce menstrual flow and recurrence of umbilical swelling. (Picture 4, 5, 6)

Discussion and conclusion:

Umbilical endometriosis is the commonest type of cutaneous endometriosis. Some case reports have also described the presence of umbilical endometriosis during pregnancy. [7,11] The risk of developing endometriosis is the lowest in black women, the highest in Asian women. Caucasian women have a higher risk of getting sick than black women.[12]

Address of Correspondence: Dr. Rabeya Akther, Chief Consultant, Bangladesh Bank Medical Center, Motijheel, Dhaka. Mail address: 422 Malibag, Dhaka 1217, Mobile: 01817517100, e-mail:rabeya.akter@bb.org.bd

Endometriosis is a benign, estrogen-dependent, gynecological disease, although it is generally recognized that the condition almost always becomes quiescent with the cessation of ovarian function. We analyzed age, parity, contraceptive history, delivery history, duration of symptoms, chief presentation, associated symptoms, size of the lesion, and management of the patient. Patient's age, detail history, life style favors the pathogenesis of endometriosis. Umbilical endometriosis may occur spontaneously.[13] Majority of these cases occurred secondary to surgical, commonly laparoscopy scars. An umbilical endometriotic lesion without surgical history is a rare condition.[7, 14]

Here she had no prior laparoscopic surgery; however, she had one prior cesarean delivery. Secondary umbilical endometriosis can occur following cesarean sections in 1% of cases.[15] Most patients present with umbilical swelling with cyclical pain and bleeding. According to Victory et al.[14], umbilical swelling was present in almost 90% of cases with less than 50% having bleeding and about 80% having pain. Pain is caused by tissue inflammation, distention, and cyclical changes. The mean size of the lesion is about 2.29cm, with color changes ranging from brown to blue, purple, black, and normal in decreasing order. In our study, however, the color changes from red to dark blue color.[1,7,14] Discoloration occurs as a result of bleeding into the lesion with hemosiderin deposition.[16] Differential diagnosis of umbilical nodules should include pyogenic granuloma, hernia, residual embryonic tissue, primary or metastatic adenocarcinoma (Sister Joseph's nodule), nodular melanoma, and cutaneous endosalpingosis.[7]

Although preliminary diagnosis is made on the basis of history and physical examination, Ultrasound examination is the basic tool in the diagnosis of this disease. Ultrasound can be used to assess the nodule size and involvement of surrounding tissues and to evaluate other pelvic pathology; hence aiding the planning of medical/surgical management.[17] Pelvic ultrasound report is unremarkable in our case. However, the gold standard in the diagnosis of endometriosis is surgery, with simultaneous confirmation in histo-pathological examination.[18] Lymphatic and hematogenous spread to the umbilicus and direct extension of endometrial cells through round ligaments or omphalo-mesenteric remnants are possible theories to explain the etiology of umbilical endometriosis[7, 19]

Pharmacological, surgical and combination treatments are used to treat endometriosis. In pharmacological treatment, we distinguish between hormonal and symptomatic treatment. Currently, hormonal treatment is allowed, for three months without histo-pathological confirmation of the disease.[3] The choice of treatment is made depending on the age of the patient, her desires, the ailments present and the form of endometriosis.

The goal of pharmacological treatment is to reduce or eliminate pain, inhibit further development and regression of endometrial foci. Initiation of pharmacological treatment of endometriosis is possible only on the basis of a clinical exam without the need to confirm the existence of the disease in histological examination (empirical therapy).[3]

Pharmacotherapy may be part of the preparation for surgery, as well as a complementary procedure in the postoperative period. There are certain types of drugs that are used in the treatment of endometriosis (Table 1).[3] Surgical excision of the lesion with sparing of the umbilicus is the preferred treatment of umblical endometriosis [7,20,21] These lesions have a low risk of malignancy,[6,22] we continue hormone therapy because of relief of symptoms; reduce the size of large lesions, to avoid hospital admission and finally to reduce cost.[23]

Table-I

Pharmacotheraputics used in the treatment of endometriosis.[3]

	Types of Drugs
1	non-steroidal anti-inflammatory drugs
2	hormonal drugs:
	progestogens
	hormonal contraceptives,
	danazol,
	analogues and gonadoliberin (GnRH)–gonadoliberin agonists and antagonistss.
3	selective progesterone receptor modulators
4	aromatase inhibitors

Non-steroidal anti-inflammatory drugs inhibit the synthesis of prostaglandins; contribute to reducing the inflammatory process and resolve pain.[24] Complex estrogen-progestogen therapy can be used cyclically or continuously.[3]

Figure 1: *3rd day of menstruation (LMP 13/1/2021)*

Figure 2: *4th day of menstruation (LMP 13/1/2021)*

Figure3: *after addition of OCP (LMP 9/2/2021). Showing hemosiderin*

Figure 4: *2nd cycle after addition of OCP(LMP 9/3/2021)*

Figure 5: *3RD cycle after addition of OCP (LMP 10/4/2021)*

Figure 6: *14th day of the cycle after addition of OCP (LMP 10/10/2022)*

Reference:

1. Matarese G., DePlacido G., Nikas Y., Alviggi C. Pathogenesis of endometriosis: Natural immunity dysfunction or autoimmune disease? Trends Mol. Med. 2003;9:223–228. doi: 10.1016/S1471-4914(03)00051-0.

2. .Benagiano G., Brosens I., Lippi D. The history of endometriosis. Gynecol. Obstet. Investig. 2014;78:1–9. doi: 10.1159/000358919.

3. Smolarz B, Szy³³o K, Romanowicz H. Endometriosis: Epidemiology, Classification, Pathogenesis, Treatment and Genetics (Review of Literature). Int J Mol Sci. 2021 Sep 29; 22(19):10554. doi: 10.3390/ijms221910554. PMID: 34638893; PMCID: PMC8508982.

4. Nezhat C., Falik R., McKinney S., King L.P. Pathophysiology and management of urinary tract endometriosis. Nat. Rev. Urol. 2017; 14:359–372. doi: 10.1038/nrurol.2017.58.

5. .Yu CY, Perez-Reyes M, Brown JJ, Borrello JA. MR appearance of umbilical endometriosis. J Comput Assist Tomogr. 1994; 18(2):269–71.

6. Boesgaard-Kjer D, Boesgaard-Kjer D, Kjer JJ. Primary umbilical endometriosis (PUE). Eur J Obstet Gynecol Reprod Biol. 2017;209:44–5.

7. .Pallavi V Bagade and Mamdouh M Guirguis :Menstruating from the umbilicus as a rare case of primary umbilical endometriosis: a case report ; Journal of Medical Case Reports 2009, 3:9326 doi:10.1186/1752-1947-3-9326

8. Teh WT, Vollenhoven B, Harris PI. Umbilical endometriosis, a pathology that a gynecologist may encounter when inserting the Veres needle. Fertil Steril. 2006;86(6):1764. e1–2.

9. Rubegni P, Sbano P, Santopietro R Fimiani M: Case four: umbilical endometriosis. Clin Exp Dermatol 2003, 28:571-572.

10. Ploteau S, Malvaux V, Draguet AP: Primary umbilical adenomyotic lesion presenting as cyclical periumbilical swelling. Fertil Steril 2007, 88(Suppl 6):1674-1675..

11. Razzi S, Rubegni P, Sartini A, De Simone S, Fava A, Cobellis L, Fimiani M, Petraglia F: Umbilical endometriosis in pregnancy: a casere1port. Gynecol Endocrinol 2004, 18(Suppl 2):114-116.

12. Goldstein D.P., de Cholnoky C., Emans S.J., Leventhal J.M. Laparoscopy on the diagnosis and management of pelvic pain in adolescents. J. Reprod. Med. 1980;24:251–256.

13. Jeffcoate's Principles of Gynecology, 8th edition ;Endometriosis and allied states,Chapter 22; page 343

14. Victory R, Diamond MP, Johns DA. Villar's nodule: a case report and systematic literature review of endometriosis externa of the umbilicus. J Minim Invasive Gynecol. 2007;14(1):23–32.

15. Santos Filho PVD, Santos MPD, Castro S, Melo VA. Primary umbilical endometriosis. Rev Col Bras Cir. 2018;45(3):e1746.

16. Dorothy Makena , Timona Obura, Steve Mutiso and Felix Oindi: Umbilical endometriosis: a case series; Makena et al. Journal of Medical Case Reports (2020) ; 14:142. https://doi.org/10.1186/s13256-020-02492-9.

17. Genovese G, Passoni E, Veraldi S, Nazzaro G. Ultrasonographic findings in primary umbilical endometriosis. An Bras Dermatol. 2018;93(2):297–8.

18. Horne A.W., Daniels J., Hummelshoj L., Cox E., Cooper K.G. Surgical removal of superficial peritoneal endometriosis for managing women with chronic pelvic pain: Time for a rethink? BJOG. 2019;126:1414–1416. doi: 10.1111/1471-0528.15894.

19. Ouedraogo NLM, Ilboudo S, Ouattara AK, Ouedraogo AS, Zida M, Zongo N, et al. A case report of Villar's nodule in a woman without surgical history. Int J Surg Case Rep. 2018;53:186–8.

20. Schachter LR, Tash J, Olgac S, Bochner BH: Umbilical endometri2osis. J Urol 2003, 170:2388-2389.

21. Purvis RS, Tyring SK. Cutaneous and subcutaneous endometriosis: surgical and hormonal therapy. J Dermatol Surg Oncol. 1994;20(10):693–5.

22. Obata K, Ikoma N, Oomura G, Inoue Y. Clear cell adenocarcinoma arising from umbilical endometriosis. J Obstet Gynaecol Res. 2013;39(1):455–61.

23. Rashida Begum.Patient management in Obstetrics & Gynecology: "Endometriosis", p245,1st edition;.Jaypee Brothers Medical Publishers,2019.

24. Horne A.W., Daniels J., Hummelshoj L., Cox E., Cooper K.G. Surgical removal of superficial peritoneal endometriosis for managing women with chronic pelvic pain: Time for a rethink? BJOG. 2019; 126:1414–1416. doi: 10.1111/1471-0528.15894.