

RECURRENT UMBILICAL ENDOMETRIOSIS: A CASE REPORT AND REVIEW OF THE LITERATURE

DAMIR DANOLIĆ¹, MARIO PULJIZ¹, DARKO TOMICA¹, ILIJA ALVIR¹ and IVICA MAMIĆ¹

¹ Department of Gynecologic Oncology, University Hospital for Tumors,
Zagreb, Croatia

Summary

Umbilical primary endometriosis is a rare localization of the functional endometrial tissue outside of uterine cavity and it represents 0.5-1% of ectopical endometriosis.

We are reporting a case in which recurrence of umbilical endometriosis has occurred 5 months after excision of umbilical endometriotic lesion and 19 years after the surgery of endometriotic cyst of the right ovary. It caused periodical bleeding from the umbilicus, associated with severe abdominal pain. Endometriosis was suspected and treated by wide surgical excision. Pathohistological analysis confirmed the diagnosis of endometriosis.

KEYWORDS: *umbilical endometriosis, differential diagnoses, surgical excision, patohistological analysis*

RECIDIVIRAJUĆA PUPČANA ENDOMETRIOZA: PRIKAZ SLUČAJA I PREGLED LITERATURE

Sažetak

Primarna endometrioza funkcionalnog endometrijskog tkiva izvan maternice s lokalizacijom u pupku je rijetkost sa zabilježenom pojavnošću 0,5-1% među ektopičnim endometrioza.

Opisujemo bolesnicu u koje je recidiv pupčane endometrioze nastupio 5 mjeseci nakon ekscizije pupčane endometriotičke lezije i 19 godina nakon operacije endometriotičke ciste u desnom jajniku. Endometrioza je uzrokovala povremeno krvarenje iz pupka, povezano s jakim boli u trbuhu. Zbog sumnje na endometrioza primijenjena je široka kirurška ekscizija. Patohistološka analiza potvrdila je dijagnozu endometrioze.

KLJUČNE RIJEČI: *pupčana endometrioza, diferencijalna dijagnoza, kirurška ekscizija, patohistološka analiza*

INTRODUCTION

Endometriosis is characterized by ectopic endometrial tissue with specific histological traits (glands, stroma, hemosiderin in macrophages).

It is estimated that there is a 10% prevalence of endometriosis in the general population (1). In asymptomatic women, the prevalence ranges from 2% to 22%, depending on the diagnostic criteria used and populations studied. Among women with chronic pelvic pain, dysmenorrhoea and subfertility the incidence of endometriosis ranges

60-70%, 40-60% and 20-30%, respectively (2). The reported prevalence of endometriosis in infertile patients is 10%-25% (2-4), and incidence peaks are at about age 40 (2).

In spite of its being a relatively common condition, endometriosis remains a professional and scientific challenge to modern medicine.

Symptoms depend on extension and localization of endometriosis. The pelvis is the most common site of the disease, giving rise to the common presenting symptoms of pelvic pain, dysmenor-

rhoea, dyspareunia and infertility (5). Endometriosis can be divided into intra- and extra-peritoneal sites. Intra-peritoneal locations are most common, and they include ovaries (30%), uterosacral and large ligaments (18%-24%), fallopian tubes (20%), pelvic peritoneum, pouch of Douglas, and gastrointestinal (GI) tract (10%) (6, 7). Extra-peritoneal locations are less common but even more difficult to diagnose due to variability of the symptoms. They include cervical portion (0.5%), vagina and rectovaginal septum, round ligament and inguinal hernia sac (0.3%-0.6%), navel (0.5%-1%), abdominal scars after a gynecological surgery (1.5%) and Cesarean section (0.5%). In extra-abdominal organs such as the lungs, urinary system, skin and the central nervous system endometriosis is extremely rare (6-9).

Umbilical endometriosis is a very rare disease, but it should be considered on the differential diagnosis of umbilical lesions (9). It has a documented neoplastic risk (10-12).

Umbilical endometriosis can occur after a surgery but it is generally spontaneous (13).

CASE REPORT

We are reporting a case of umbilical endometriosis in a 39-year-old woman. The patient had two abdominal surgeries. Nineteen years ago the patient underwent her first surgery for an endometriotic cyst on the right ovary. Laparotomy-Pfannenstiel incision, right ovariectomy and appendectomy were performed. Second abdominal surgery was 8 years ago, she gave birth by Cesarean section.

The patient presented at our Department with a dark brown nodule associated with severe abdominal pain and umbilical bleeding occurring only during her menstrual period.

The patient had similar symptoms a year ago and at that time medical examination revealed a umbilical nodule, measuring 15 mm in diameter. The patient then underwent surgical treatment. The nodule was excised and the subsequent histological examination set the diagnosis of umbilical endometriosis. Surgical excision was not effective: at follow-up 5 months later, there was a recurrence and the patient was hospitalized in our Department.

Physical examination revealed a 10 x 5 mm brown umbilical nodule. No other masses were observed in the abdomen or by the low transverse scar.

The ultrasonographic evaluation showed a hypoechogenic umbilical lesion suggestive of endometriosis and no other spots of endometriosis in the pelvis. CA-125 serum level was measured. It was within normal ranges.

The patient was submitted for a surgical excision and histological examination of the lesion. Wide excision was performed and the specimen was sent for patohistological analysis.

Pathohistological analysis confirmed the diagnosis of endometriosis. Histologically, there was the normal epidermis on the surface, and in the dermis there were fragments of endometrial stroma and glands coated with endometrial epithelium cell type. The described changes do not reach the edges of the resected tissue specimen.

The patient was discharged on the third postoperative day and sutures removed on the seventh postoperative day. At 8-month follow-up, the patient was asymptomatic and there were no signs of recurrence (Figure 1).

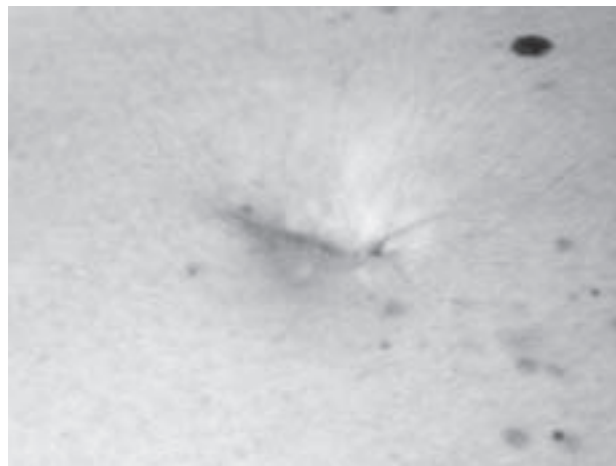


Figure 1. Umbilical scar after excision of endometriotic lesion, with no signs of the disease

DISCUSSION

Endometriosis of the skin is rare and its most frequent form develops over gynecologic or obstetric scars, but it may also appear spontaneously in the umbilical area (14).

The cause of umbilical endometriosis appears to be unknown, although there are two theories: metastases and metaplasia. The metastasis theory suggests that the implantation is either by lymphatic or hematogenous spread. The metaplasia theory suggests that multipotential cells can develop into endometrial tissue, often stimulated by inflammation (15, 16).

Clinical diagnosis is difficult, and umbilical endometriosis can be easily confused with other conditions such as benign and malignant tumors (11). Differential diagnoses for the umbilical nodule with no discharge are umbilical granuloma, simple inclusion cyst, irreducible umbilical hernia, umbilical polyp, melanocytic nevus, folliculitis, seborrheic keratosis, primary malignancy such as malignant endometriosis in the umbilicus and secondary metastatic tumor from an intra-abdominal malignancy (9, 15, 17). Differential diagnoses for umbilical nodule with discharge or weeping include pilonidal sinus, urachal cyst, urachal sinus, foreign body granuloma and rare cause as in the presented case is umbilical endometriosis (15, 17). In our case, it was easy to predict final diagnosis. Endometriosis was suspected because of the presence of typical cyclic bleeding, swelling, abdominal pain and history of pelvic and umbilical endometriosis.

Victory R, Diamond MP and Johns DA analyzed 122 patients with documented umbilical endometriomas from 1966 to 2007 and 109 cases before 1953.

In their review mean age of the study population was 37.7 +/- 0.98 years, lesions were predominantly brown (19.1%), blue (13.2%), or purple (10.3%) and patients frequently had with pain (77.93%), cyclical bleeding (47.1%), and swelling (88.2%). Most patients had no history of endometriosis (73.1%) and only one patient required repeat surgical therapy (18).

Umbilical endometriosis usually has typical presentation with little variation. The clinical presentation of umbilical endometriosis in our patient is similar as in most of the cases. Wide local excision with clear margins is the key point to prevent recurrence, as indicated in our case.

Surgical excision with removal and pathological analysis of the specimen is recommended for both therapeutic and diagnostic purposes. Other diagnostic methods have been proven to be nonspecific and unreliable in the literature (19).

Cutaneous endometriosis could be a sign of internal endometriosis (9) and ultrasonographic evaluation of the pelvis should always be performed.

CONCLUSION

The literature review summarizing knowledge and our experience leads us to conclusion that wide excision with free margins is the most important element of the treatment strategy (10, 11). Umbilical endometriosis is a rare entity, but it should be remembered as a possibility in cases of umbilical nodulations or bleeding, especially if there is a history of pelvic surgery with endometrial manipulation. Surgical excision is the treatment of choice.

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Author's address: Damir Danolić, M.D., Department of Gynecologic Oncology, University Hospital for Tumors, Illica 197, 10000 Zagreb, Croatia; e-mail: damir.danolic@gmail.com