



CASE REPORT

Primary Cutaneous Endometriosis of Umbilicus

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Cutaneous endometriosis is defined by the presence of endometrial glands and/or stroma in skin and represents less than 1% of all ectopic endometrium. Cutaneous endometriosis is classified as primary and secondary. Primary cutaneous endometriosis appears without a prior surgical history and secondary cutaneous endometriosis mostly occurs at surgical scar tissue after abdominal operations. The most widely accepted pathogenesis of secondary endometriosis is the iatrogenic implantation of endometrial cells after surgery, such as laparoscopic procedures. However, the pathogenesis of primary endometriosis is still unknown. Umbilical endometriosis is composed only 0.4% to 4.0% of all endometriosis, however, umbilicus is the most common site of primary cutaneous endometriosis. A 38-year-old women presented with solitary 2.5 × 2.0-cm-sized purple to brown colored painful nodule on the umbilicus since 2 years ago. The patient had no history of surgical procedures. The skin lesion became swollen with spontaneous bleeding during menstruation. The skin lesion was diagnosed as a keloid at private hospital and has been treated with lesional injection of steroid for several times but there was no improvement. Imaging studies showed an enhancing umbilical mass without connection to internal organs. Biopsy specimen showed the several dilated glandular structures in dermis. They were surrounded by endometrial-type stroma and perivascular infiltration of lymphocytes. The patient was diagnosed as primary cutaneous endometriosis and skin lesion was removed by complete wide excision without recurrence. We report an interesting and rare case of primary umbilical endometriosis mistaken for a keloid and review the literatures. (*Ann Dermatol* 29(5) 621 ~ 625, 2017)

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-Keywords-

Cutaneous endometriosis, Endometriosis of umbilicus, Primary cutaneous endometriosis, Umbilical endometriosis

INTRODUCTION

Endometriosis is histopathologically defined by the presence of endometrial glands and/or stroma outside of the endometrium¹. Ectopic endometriosis could be developed in many other tissues, most commonly affects pelvic organs such as ovaries, fallopian tubes, uterine ligaments, pelvic wall². Primary cutaneous umbilical endometriosis, which is also known as Villar's nodule, is a rare manifestation of endometriosis³. Secondary endometriosis mostly occurs at surgical scar tissue after abdominal operations⁴. The most widely accepted pathogenesis of secondary endometriosis is the iatrogenic implantation of endometrial cells after surgery, commonly after laparoscopic procedures⁵. However, the pathogenesis of primary endometriosis is still unknown.

To date, umbilical endometriosis has been reported to represent about 0.4% to 4.0% of all endometriosis and accounts for 30% to 40% cases of cutaneous endometriosis. Among cutaneous endometriosis, primary umbilical endometriosis was considered even less common.

CASE REPORT

A 38-year-old multigravida female visited our department because of a painful nodule on her umbilicus. The patient recalled that the lesion was observed 2 years ago and the

lesion became swollen with spontaneous frank bleeding during menstruation. The patient had no history of surgical procedure, nor any family history of malignancy. The nodule was first diagnosed as a keloid at a private clinic and had been treated with intralesional injection of steroid for several times without any signs of improvement. Physical examination revealed a 2.5×2.0-cm-sized brownish to purple colored nodule on the umbilicus (Fig. 1). Imaging studies were carried out for differential diagnosis with Sister Mary Joseph nodule and keloid. Umbilical ultrasonography showed a mass with heterogenous echogenicity, increased vascularity and abdominal computed tomography (CT) revealed enhancing mass at umbilicus without connection to abdominal organs. Histopathological examination showed dilated glandular structures surrounded by cellular endometrial-type stroma and deep perivascular infiltration of lymphocytes (Fig. 2). According to these findings, the umbilical lesion was diagnosed as primary cutaneous endometriosis and it was removed by



Fig. 1. About 2.5×2.0-cm-sized brownish to purple colored nodule on the umbilicus.

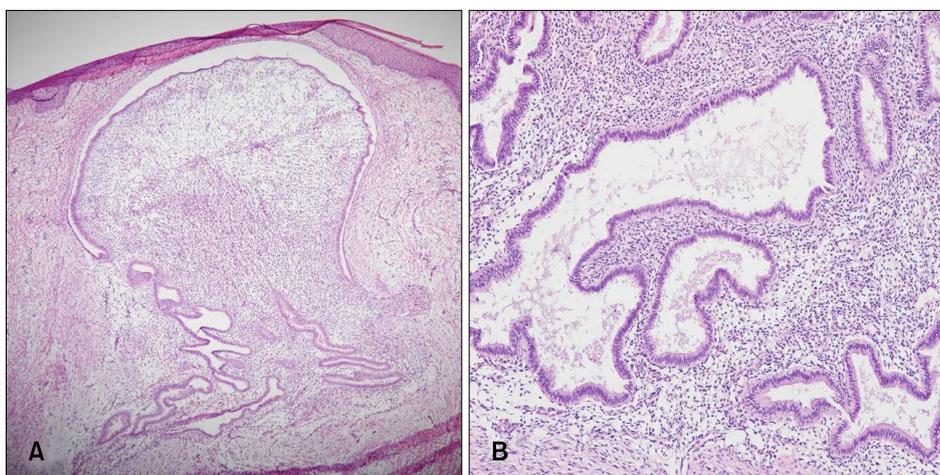


Fig. 2. (A, B) Specimen showed lesions in the superficial dermis and deep dermis comprising dilated glandular structures, surrounded by cellular endometrial-type stroma (H&E; A: ×40, B: ×100, respectively).

local surgical excision. Postoperative period was unremarkable and the patient was followed up for 2 years without recurrence.

DISCUSSION

Cutaneous endometriosis represents 0.5% to 1.0% of all patients with ectopic endometriosis. Less than 30% of cutaneous endometriosis presents without prior surgical operative history, which is termed as primary spontaneous cutaneous endometriosis³. Umbilical endometriosis is composed 0.4% to 4.0% of all endometriosis, high as two-fifths of extragenital endometriotic lesions. Moreover, umbilicus is the most common site of primary cutaneous endometriosis⁶. Umbilical endometriosis occurs in female of reproductive age and associated symptoms are cyclic pain, bleeding and swelling of the lesion according to the menstrual cycle⁴.

Several possible pathogenesis of umbilical endometriosis were suggested by multiple investigators. The most commonly accepted mechanisms are lymphatic or vascular migration, cellular metaplasia, and iatrogenic metastasis⁵. Suggested theory includes migration of endometrial tissue from retrogression of menstruation. Survival of endometrial implants after implantation may depend on local and systemic factors. Inflammatory process is then stimulated by microvascular endothelial injury. Accordingly, it might enhance adhesion of tissue implants in outside of endometrial tissues via production of adhesion molecules such as integrin and e-cadherins⁷. Major etiologic pathogenesis of secondary umbilical endometriosis could be explained by iatrogenic metastasis, endometrial cells implant in scars after surgery. In comparison, primary umbilical endometriosis may be explained by the theory of vascular or lymphatic migration.

Table 1. Literature review of primary umbilical endometriosis

Reference	Age at diagnosis (yr)	Age at initial (yr)	Initial diagnosis	Presenting symptom	Treatment
Theunissen and Ijpma ⁹	47	47	Umbilical hernia	Asymptomatic umbilical nodule	Surgical excision
Calagna et al. ¹⁰	33	33	Umbilical granuloma	Spontaneous catamenial bleeding	Surgical excision
Chikazawa et al. ¹¹	46	44		Swelling during menstrual period	Surgical excision
Chikazawa et al. ¹¹	27	23		Pain during menstrual period	Surgical excision
Pariza and Mavrodin ¹²	26	25		Pain and discharge	Surgical excision
Paramythiotis et al. ¹³	46	-	Uterine leiomyoma	Abdominal and pelvic pain	Total hysterectomy with excision of nodule
Ghosh and Das ¹⁴	33	33	Umbilical endometriosis	Cyclic pain and swelling	Excisional biopsy
Gin et al. ¹⁵	31	31		Swelling during menstrual period	Surgical excision
Kahlenberg and Laskey ¹⁶	24	20	Abcess	Bloody discharge during menstrual period	Surgical excision
Fancellu et al. ¹⁷	24	24	Umbilical endometriosis	Concomittant bleeding on menstruation	Surgical excision
Jaime et al. ³	33	33		Spontaneous bleeding	Not described
Efremidou et al. ¹⁸	44	38~39	Granuloma	Pain during menstrual period	Surgical excision
Kesici et al. ¹⁹	38	38	Omphalitis	Umbilical secretion and mass	Surgical excision
Fernández-Aceñero and Córdoba ⁴	38	35	Umbilical endometriosis with uterine fibroids	Cyclic pain	Abdominal hysterectomy with excision of nodule
Dadhwal et al. ⁸	42	42		Cyclic pain and blackish discoloration	Surgical excision
Bagade and Guirguis ²⁰	35	35	Umbilical endometriosis	Spontaneous and cyclic bleeding	Goserelin acetate, and then surgical excision
Victory et al. ⁶	47	46		Umbilical bleeding	Surgical excision
Boesgaard-Kjer et al. ²¹	28.5 (mean age of 10 patients)	-		Periodic color change and tenderness	Surgical excision
Wiegatz et al. ²²	27	25	Umbilical endometriosis	Increasing cyclic pain	Oral contraceptive, and then surgical excision
Taniguchi et al. ²³	45	42	Umbilical endometriosis	Painful umbilical mass	Surgical excision
Chew et al. ²⁴	44	44	Umbilical endometriosis	Progressively enlarging umbilical nodule	GnRH analogue leuprorelin acetate
Claas-Quax et al. ²⁵	27	27		Catamenial bleeding	Surgical excision
Sidani et al. ²⁶	37	37		Cyclic swelling and discharge	Surgical excision
Sengupta et al. ²⁷	29	28		Painful nodule	Excisional biopsy
Minaidou et al. ¹	26	-	Umbilical hernia	Umbilical pain and dark purplish nodule	Surgical excision
Weng and Yang ⁵	37	-		Cyclic bleeding	Gestrinone, and then surgical excision
Kim et al. ²⁸	42	42	Epidermal cyst	Size increase and pain	Excisional biopsy
Song et al. ²⁹	25	23	Dermatofibroma	Size increase, pain, discoloration	Surgical excision
Kyamidis et al. ³⁰	37	27	Umbilical endometriosis	Tenderness and occasional bleeding	Not described

Twenty-nine published studies with primary umbilical endometriosis were identified in the literature written in English and Korean language during the period 2000~2016 (Table 1)^{1,3-6,8-30}. Primary umbilical endometriosis is initially very rare condition, but it is now increasing in number. Based on all the reports, the mean age of patients was 35.1 years.

Differential diagnosis of umbilical endometriosis includes keloid, metastasis of visceral carcinoma, which is referred as Sister Mary Joseph nodule and melanoma³. Therefore, physicians should work on imaging studies such as ultrasonography or CT or magnetic resonance imaging. Furthermore, diagnosis must be confirmed histopathologically to exclude malignancy. More importantly, keloid is clinically very similar to umbilical endometriosis. Clinicians should pay particular attention to patients, especially history of surgery or trauma, and presenting symptoms that are related to menstrual cycle. If treatment with steroid intralesional injection does not improve the symptom, umbilical endometriosis should be considered for differential diagnosis.

Surgical excision is the definitive treatment. Hormonal therapy with gonadotropin-releasing hormone agonists, oral contraceptive and danazol can be used before surgical excision to decrease the size of the lesion and make symptom relief^{8,31}. Recurrence rate is very rare⁹. In our case, the lesion was confirmed by umbilical ultrasonography and abdominal CT and histopathological finding, and removed by local surgical excision.

In conclusion, cutaneous endometriosis of umbilicus should now be recognized as a primary or metastatic presentation or iatrogenic complication of endometriosis. Patients with primary umbilical endometriosis should undergo careful history and physical examination to rule out potential malignancies. Moreover, differential diagnosis with keloid is very important. If the lesion diagnosed with keloid has cyclic symptoms with menstrual period, and does not improve with treatment, umbilical endometriosis should be suspected. Surgical excision is the treatment of choice to prevent recurrence and to reduce the risk of malignant transformation.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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