Spontaneous cutaneous umbilical endometriosis: a rare variant of extragenital endometriosis

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Summary

Umbilical endometriosis is a very uncommon condition which presents as a pigmented umbilical nodule, papular or cystic, with symptoms punctuated rhythmed by menses. The authors report the case of a 32-year-old with spontaneous umbilical endometriosis. Surgical resection was performed with a good cosmetic result and no recurrence at six months. A review of the literature allowed the authors to discuss the diagnosis difficulties and treatment in a underdeveloped country.

Key words: Endometriosis; Surgery; Extragenital.

Introduction

Endometriosis was first described by Rokistansky in 1860 and was defined as the presence of proliferation of endometrium (endometrial glands and stroma) outside the uterine cavity, with the most common site being the pelvis [1]. It is a common gynecological condition that affects up to 22% of all women, eight to 15% of women of reproductive age, and six percent of premenopausal women [2, 3], and commonly occurs in pelvic organs of women presenting with dymenorrhea, menorrhagia, pelvic pain, and infertility [4, 5]. Furthermore, ectopic endometrioma occurs in the abdominal wall in 0.03% to 1.08% with anamnesis of obstetrics or gynecologic procedures [1]. However it could sometimes be found with no previous scar (iatrogenic or not) [6].

Primary (spontaneous) umbilical endometriosis (SUE) was first described by Villar in 1886 [6] and represents a rare condition, estimated in 0.5 % to 1% of all extragenital endometriosis [5].

The authors present a case of umbilical painful skin nodule that presented first to the general surgeons, which was clinically diagnosed as umbilical papilloma but finally resulted histologically as an abdominal wall endometrioma. This is a rare case of primary umbilical spontaneous abdominal wall endometriosis.

Case Report

A nulliparous woman, 32 years of age, presented with a pigmented umbilical mobile mass, which was tender to palpation. over a period of five years. Her lesion was associated with cyclical changes in size with worsening during menses and severe pain over the past eight months. There was no bleeding. The mass replaced the umbilicus entirely. She had never been pregnant nor had any abdominal surgery. She had no history of dysmenorrhea and never used hormonal contraception.

On physical examination, she had a hard, black umbilical mass that measured three by two cm, stiff, and painful and irreducible to palpation (Figure 1). The nodule was movable from all skin plans. Given the clinical history and the physical appearance of the lesion, the diagnosis of umbilical endometriosis was strongly suspected.

Ultrasound scan of the mass showed a well-defined, oval-shaped anechoic area. The preliminary diagnosis was incarcerated umbilical hernia. There were no other localizations. The determination of CA-125 came back normal at 13.2 IU/ml. Hysterorraphy had found no other site of endometriosis.

Surgical excision of the umbilicus nodule and reconstruction was performed using a purse-string suture technique. Histological examination of the surgical specimen confirmed the diagnosis of endometriosis (Figure 2). Surgical pathology revealed a 5.0 × 4.0 × 3.0 cm area of endometriosis with negative margins at the umbilicus. Characteristic of cutaneous endometriosis, endometrial glands in a fibrous eosinophilic stroma were noted within the dermis.

Discussion

Endometriosis is a very common gynecological disease which usually occurs in the pelvic cavity [1, 7]. Extrapelvic endometrioma is an uncommon gynecological problem [8] with an estimated incidence of 0.5% to 1% [5]. It is the presence of ectopic endometrial tissue in almost any organ and cavity of the female body, including the lung, bowel, ureter, and brain, but the most including location is the abdominal wall [4, 5, 6]. Endometriosis involving the abdominal wall is termed as cutaneous endometriosis, and it is mostly associated with surgical scars after abdominal or pelvic operations, or may rarely occur spontaneously [7]. The primary umbilical endometriotic lesion was firstly described by Villar in 1886. There is no systematic literature review on published cohorts of patients having umbilical endometrioma [6].

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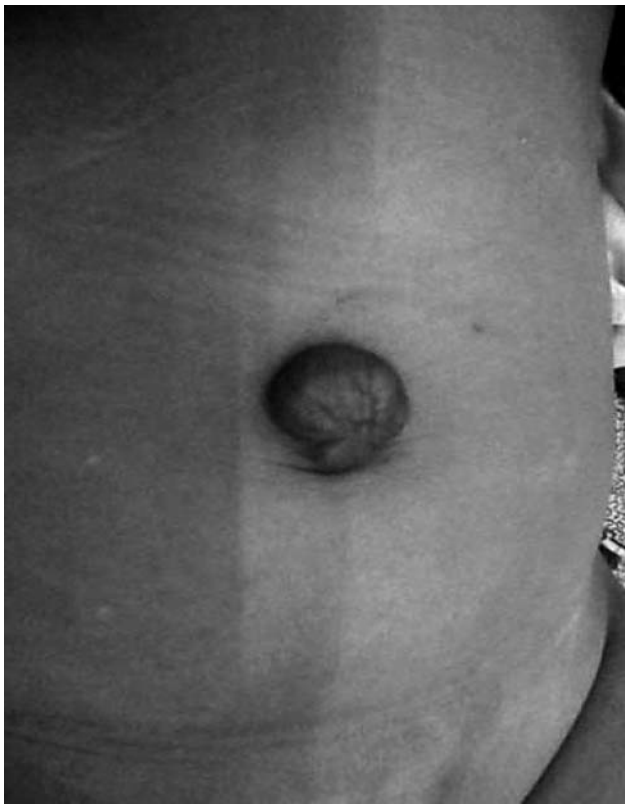

Figure 1. — Umbilical endometriosis nodule

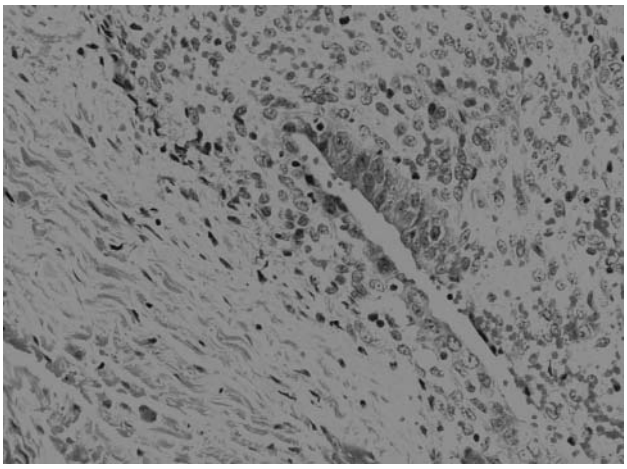

Figure 2. — Umbilical endometriosis: endometriotic glands with metaplasia

For Papavramidis *et al.*, the definition of wall endometriosis includes lesions that are not due to previous surgical procedures, and such cases are referred to as spontaneous abdominal wall endometriosis [4]. No large prospective or retrospective studies have investigated SUE [6]. However, Horton *et al.* found that it is less common than scar-related endometriosis, and represents only 20% of all the cases [9]. The most common locations of SUE appear to be the umbilicus and groin [4, 5].

There are several doubts concerning the etiopathogenesis of the condition during the decades [7]. Many theories have been put forward to explain pathogenesis of endometriosis [4, 9, 10]. They have been classified into three main categories, i.e., the embryonic rest theory, the coelomic metaplasia theory, and the migratory pathogenesis theory [4, 7]. The embryonic rest theory explains the pelvic endometriosis such as a stimulus to a Müllerian origin cell nest [4, 9-11]. The migratory pathogenesis by implantation or retrograde menstruation theory explains the implantation on surrounding pelvic structures [4, 9-11]. Direct transplantation can explain the endometriosis occurring on surgical scars, but does not explain the distant locations, therefore Halban advocated the dissemination theory of vascular migration through vascular or lymphatic channels and in also surgical procedures [5, 9]. Even if the actual mechanism of SUE remains unclear, none of the suggested theories should be excluded until convincing experimental data are obtained. Some authors advocate a combination of the aforementioned theories [9, 10]. Papavramidis *et al.* suggested that the dissemination theory through lymphatic or vascular spread can explain occurrence of spontaneous abdominal wall endometriosis [4] as in the case report herein.

As for many authors, extragenital endometriosis has various presentations and remains a difficult condition to diagnose and treat [4, 10]. Clinical diagnosis has varying features such as flesh colored nodule, black nodule, flesh colored bluish, and with a size range of up to several centimeters. Hence, malignant melanoma should be considered [7].

In primary cutaneous umbilical endometriosis, the chief symptom is usually a mass at the site of maximum tenderness, which varies in size following the menstrual cycle, while the typical characteristic is cyclic pain associated with menses [4, 10].

There are reports that the pain can be constantly present without any association with the menstrual cycle, but this is generally regarded as atypical, which may explain why umbilical endometriosis is often misdiagnosed clinically. In such atypical cases, and especially in cases of SUE, signs and symptoms may occur singly, which always hinders an accurate diagnosis. In published series, the reported preoperative diagnosis rate has varied between 20% and 50% [4].

This diagnostic failure could be due to general surgeons, who often make the diagnosis, not being sufficiently familiar with SUE. Another possible explanation is the atypical presentation of the disease along with the possible differential diagnoses, including lipoma, sarcoma, lymphoma, primary or metastatic cancer, cysts, and inguinal or incisional hernia [4].

Clinical diagnosis is often difficult and patients suffering from this condition are usually of reproductive age and often present with an umbilical mass associated with swelling, pain, discharge, or cyclical bleeding [5]. Cyclical pain in umbilicus with a palpable mass was most the presenting symp-

tom as in this case, as for many authors in literature review [1, 7]. Rare cases of cyclical bleeding discharge in umbilicus have been described from the umbilical mass during menstrual period [12]. The umbilical nodule has been described as being flesh-colored, brownish, dark-bluish, or simply a subcutaneous mass, with a size that typically varies from 0.5 cm to several centimeters, but can be enormous [4, 10]. There may be associated symptoms of coexistent pelvic endometriosis, although the incidence of pelvic disease in abdominal wall endometriosis is within the same range as the general population (8%–15%) [5, 9].

The possibility of co-existing genital-pelvic endometriosis should be excluded by ultrasonogram and or exploratory laparoscopy of abdominal cavity [7]. Due to the variable macroscopic appearance of umbilical endometriomas, the differential diagnosis of umbilical nodules includes: pyogenic granuloma, embryological rests, irreducible hernia, endometriosis, inclusion cysts, primary tumours or secondary metastatic tumours from intra-abdominal malignancy [7].

According to Catalina-Fernandez *et al*., dermoscopy can be helpful in cases of cutaneous or subcutaneous endometriosis, with cytologic smears revealing high cellularity with hemosiderin-laden macrophages and sheets of stromal and epithelial cells on a hemorrhagic background [10]. The histologic diagnosis of endometrioma requires two of the three following features: endometrial-like glands, endometrial stroma, or hemosiderin pigment [4]

Surgical excision is necessary for proper histological diagnosis as well as for therapeutic purpose. It is the preferred treatment in all cases of UE consisting in wide local excision of the mass.

In case, no additional of hormonal therapy was associated. Papavramedis *et al.* proposed it whenever severe pelvic disease is assumed or demonstrably present [4,]. Therefore, surgical resection of an umbilical endometrioma with safety and clear margins is the treatment of choice, and offers the highest probability of both a definitive diagnosis and a favorable outcome. Preservation of the umbilicus is preferred, but if the umbilicus has to be completely removed in order to achieve radical excision, certain methods can provide adequate reconstruction [11].

Complete excision of the umbilical lesion with partial resection of the underlying fascia is recommended, to avoid local recurrence [4, 10]. Therefore, wide excision with a margin of at least one cm is considered the treatment of choice, even for recurrent lesions [4, 10].

Several authors have advocated the use of hormonal therapy with a gonadotropin-releasing hormone analog (eg, danazol or progesterone), with the aim of decreasing the size of the mass and facilitating surgery. Furthermore, these hormones can be added to surgical treatment in cases of severe pelvic disease [5].

Medical management cannot be enthusiastically recommended, due to its reported success rate being low, with it offering only temporary alleviation of the symptoms, and serious adverse effects often being followed by recurrence after cessation of drug intake [1,6]. It is also known that both abdominal wall and scar endometriosis are less responsive to hormonal therapy [6].

Malignant transformation of abdominal wall endometriosis is a very rare complication (in 1% of cases) [6].

Spontaneous abdominal wall endometriosis is usually diagnosed by pathology, especially in cases without the typical triad of mass, pain, and cyclic symptomatology, as in the case report presented herein.

Conclusion

Primary umbilical endometriomas is a rare event. Careful history-taking and physical examination are essential to making the correct diagnosis, although this can be difficult in atypical presentations, and so other causes of umbilical lesions should be considered. In underdeveloped countries, complete excision and histology is highly recommended for obtaining a definitive diagnosis and to rule out malignancy. Radical surgical resection is the treatment of choice.

References

[1] Thapa A., Kumar A., Gupta S.: "Abdominal wall endometriosis: Report of a case and how much we know about it". *Internet J. Surg.*, 2007, *9*, 30.
[2] Catalina-Fernandez I., Lopez-Presa D., Saenz-Santamaria J.: "Fine needle aspiration cytology in cutaneous and subcutaneous endometriosis". *Acta Cytol.*, 2007, *51*, 380.
[3] Agarwal A., Fond Y.F.: "Cutaneous endometriosis". *Singapore Med. J.*, 2008, *49*, 704.
[4] Papavramidis T.S., Sapalidis K., Michalopoulos N., Karayanopoulou G., Raptou G., Tzioufa V., *et al.*: "Spontaneous abdominal wall endometriosis: a case report". *Acta Chir. Belg.*, 2009, *109*, 778.
[5] Pavalli V.B., Mamdouh M.G.: "Menstruating from the umbilicus as a rare case of primarily umbilical endometriosis: a case report". *J. Med. Case Rep.*, 2009, *3*, 9326.
[6] Efremidou EI, Kouklakis G, Mitrakas A, Liratzopoulos N, Polychronidis ACh.: "Primary umbilical endometrioma: a rare case of spontaneous abdominal wall endometriosi". *Int. J. Gen. Med.*, 2012, *5*, 999. doi: 10.2147/IJGM.S37302. Epub 2012 Dec 5.
[7] Singh A.: "Umbilical endometriosis mimicking as papilloma to general surgeons: a case report". *Australas Med. J.*, 2012, *5*, 272. doi: 10.4066/AMJ.2012.1198. Epub 2012 May 31.
[8] Kodandapani S., Pai M.V., Mathew M.: "Umbilical laparscopic scar endometriosis". *J. Hum. Reprod. Sci.*, 2011, *4*, 150. doi: 10.4103/0974-1208.92291.
[9] Horton J.D., DeZee K.J., Ahnfeldt E.P., Wagner M.: "Abdominal wall endometriosis: a surgeon's perspective and review of 445 cases". *Am. J. Surg.*, 2008, *196*, 207.
[10] Chatzikokkinou P., Thorfinn J., Angelidis I.K., Papa G., Trevisan G.: "Spontaneous endometriosis in an umbilical skin lesion". *Acta Dermatovenerol. Alp.Panonica Adriat.*, 2009, *18*, 126.
[11] Kyamidis K., Lora V., Kanitakis J.: "Spontaneous cutaneous abdominal endometriosis: report of new case with immunohistochemical study and literature review". *Dermatol. Online J.*, 2011, *17*, 5.

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