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Case Report

Primary umbilical endometrioma: Analyzing the pathogenesis of endometriosis from an unusual localization

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ABSTRACT

Objective: This report presents a rare case of symptomatic primary umbilical endometriosis and reviews the literature on the topic with the aim to clarify some questions on the origin of endometriosis.
Case report: A 33-year-old woman with cyclic umbilical bleeding was found to have umbilical endometriosis. She had no history of pelvic or abdominal surgery. There was no past history of endometriosis or endometriosis-associated symptoms. An omphalectomy was performed after explorative laparoscopy to carefully inspect the abdominopelvic cavity and assess any coexisting pelvic endometriotic lesions. Histological examination confirmed the diagnosis of umbilical endometriosis.
Conclusion: Umbilical endometriosis is a rare but under-recognized phenomenon. Primary lesions are difficult to recognize, but probably represent an independent nosological entity. The possibility of endometriosis must be considered during the evaluation of an umbilical mass despite the absence of previous surgery. Complete excision and successive histology are highly recommended.

Introduction

Endometriosis is an elusive chronic disease caused by the growth of functional endometrial-like tissue outside the uterus, which in turn causes infertility and pelvic pain, and affects up to 10% of women of reproductive age [1]. Most commonly, the condition affects pelvic organs, however, among all diagnosed endometriosis, 12% are encountered at extragenital sites, such as the lungs, diaphragm, or umbilicus [2,3]. Umbilical endometriosis is a rare entity, first described in 1886; since then, approximately 100 cases have been reported in the medical literature [4]. It has an estimated incidence of 0.5–1% of all patients with extragenital endometriosis [5], and this percentage includes both secondary scar-related and spontaneous primary forms. The former, usually associated with pelvic endometriosis, is most often found in old surgical scars with direct seeding after laparotomy or laparoscopy, which has been proposed as the pathogenesis. By contrast, the spontaneous primary umbilical variety, considered much less common than the iatrogenic form, is not associated with previous abdominal or uterine surgery, and, to date, has an unclear origin [6].

We present an uncommon case of spontaneous umbilical endometriosis without peritoneal involvement, treated with laparoscopy-assisted radical surgery. In addition, a review of the literature on the topic is offered.

Case report

A 33-year-old woman presented with a 6-month history of spontaneous catamenial bleeding from the umbilicus. The patient had no history of abdominal or uterine surgery or trauma to her umbilicus, and had a spontaneous vaginal delivery. There was no past history of endometriosis or other endometriosis-associated symptoms. She did not use any oral contraception and had a regular menstrual cycle. On physical examination, conducted in the premenstrual period, there was a 1-cm solid, painful, reddish nodule at the surface of her umbilicus (Fig. 1A). There were no signs of infection. Preoperative workup included a transvaginal

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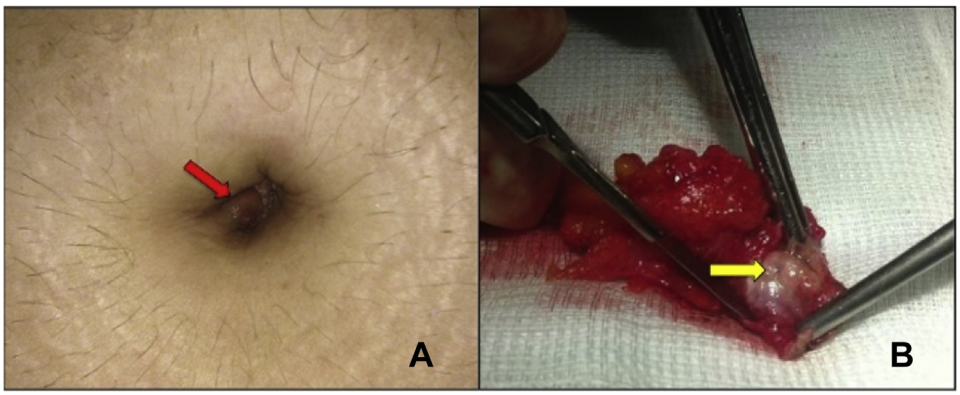

Fig. 1. Endometriosis of the umbilicus: (A) preoperative appearance of the umbilical endometriotic nodule (red arrow) and (B) excised tissue; yellow arrow shows a focal pigmented area.

ultrasound scan (US), and abdominopelvic and abdominal wall magnetic resonance imaging (MRI). At transvaginal US, the uterus and both ovaries appeared normal. Transcutaneous US showed a 0.8-cm well-defined, oval-shaped hypoechogenic area, apparently not involving the entire umbilicus but extending 2 mm below the skin surface. An initial differential diagnosis included umbilical granuloma, simple inclusion cyst, or benign or malignant neoplasm of the umbilicus. MRI confirmed the normality of the genital tract and abdominal organs, and revealed a small, poorly defined hypervascular umbilical nodule, in both T1- and T2-weighted sequences. Tumor marker cancer antigen 125 was negative. A lesion biopsy confirmed the suspected diagnosis of umbilical endometriosis. Therefore, it was decided to perform surgical excision after explorative laparoscopy, to carefully inspect the abdominopelvic cavity, and assess and treat any coexisting pelvic endometriotic lesions. The patient consented to surgical resection of the umbilical endometriosis after being fully informed of the associated risks and complications.

Surgical technique

A Veress needle was inserted along the left midclavicular line, and adequate pneumoperitoneum was obtained. A 5-mm trocar was inserted in the same site and, with a 5-mm optics, the pelvis and the upper abdomen were examined. No suspicious lesions, endometriotic or otherwise, were identified. Following the laparoscopic procedure, an omphalectomy was performed; the navel was excised *en bloc* including the nodule, fascia, and peritoneum (Fig. 1B). The peritoneum and fascia were then sutured, prior to fixation of the periumbilical skin to the latter. The skin was closed using interrupted absorbable sutures.

Histological examination confirmed the diagnosis of umbilical endometriosis. Microscopically, a typical area of endometriosis consisting of endometrial-type glands and stroma was seen. Additional immunohistological stains were performed (CK7+, CK20−, CD10+, ER+, and PR+) to support the histological finding of endometriosis (Figs. 2–4). Follow up consisted of assessment at 1 month and re-examination at 6 months. No relapses of the umbilical lesion have occurred, and, to date, the patient is asymptomatic with a normal umbilicus. Considering the risk of recurrence, she was invited to undergo periodic follow up.

Literature review

A review of the medical literature on spontaneous umbilical endometriosis from 1990 to 2013 was carried out. To this end, a PubMed electronic database search was initiated, using the following key words: endometriosis, umbilical nodule, umbilical endometriosis, primary umbilical endometriosis, and spontaneous umbilical endometriosis. All pertinent English-language articles

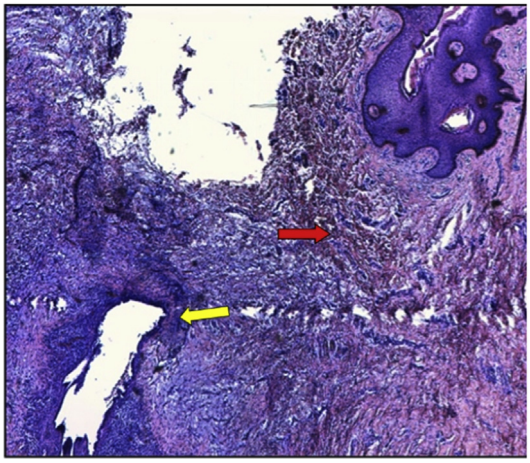

Fig. 2. Histological examination demonstrated endometrial gland surrounded by endometrial stroma (yellow arrow) beneath the squamous epithelium of the skin (red arrow) (hematoxylin and eosin, 4×).

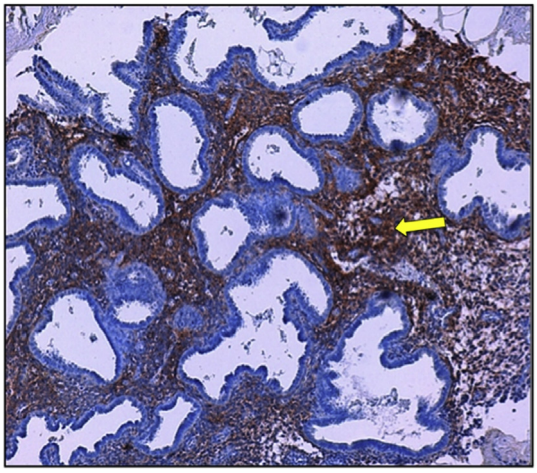

Fig. 3. Endometrial stroma diffusely positive for CD10 (yellow arrow) (immunohistochemical, 10×).