Primary umbilical endometriosis

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Summary

A case of primary solitary umbilical endometriosis is presented. The presentation, differential diagnosis and work-up is discussed and the literature is reviewed.

Key words: Primary; Umbilical endometriosis; Solitary; Therapy.

Introduction

Endometriosis is the presence of endometrial tissue outside the uterine cavity. It affects about 20-25% of women who are diagnosed with infertility. The most common location is in the pelvis, affecting the ovaries, the uterine suspensory ligaments, the rectovaginal septum, and the pelvic peritoneum in decreasing order of frequency [1]. However, other locations can be involved such as the skin, the lungs, the gastrointestinal tract, and the bladder. These lesions are most often associated with pelvic disease. We report a case of primary solitary umbilical endometrioisis.

Case Report

Mrs. PS, a 37-year-old, gravida 2, para 2 woman, presented because of a two-month history of an umbilical "draining sinus". She reported pain at that site, cyclic swelling and occasional drainage of brownish material. The patient was otherwise healthy. Her last delivery was 14 years prior to presentation. Physical exam revealed a 2.5 cm, tender, and indurated mass in the umbilicus. The lesion was excised including the underlying fascia. It consisted of several small cysts filled with old blood. Histologically, inclusions were made of endometrial mucosa with cystic glands filled with blood, and endometrial stroma exhibiting numerous siderophages (Figure 1). The patient had a smooth postoperative course. Later on she underwent diagnostic laparoscopy which revealed no evidence of pelvic endometriosis. One year later, she remains healthy on no treatment and shows no evidence of recurrence.

Discussion

Endometriosis is usually a pathologic diagnosis which depends on the presence of endometrial-like glands, endometrial stroma, and hemosiderin pigment in an ectopic location outside the uterus. In the pelvis, the appearance is so typical that it can be identified grossly. Endometriosis occurs in the umbilicus in 0.5 -1% of all cases [2]. It affects women of the reproductive age group. It usually presents itself as a single, solid, brownish or bluish nodule ranging in size from a few millimeters to 6 cm in diameter. The lesions can be painful or tender, especially before the onset of menstruation. They might also bleed cyclically, because they do respond to ovarian hormones like ectopic endometrium. The differential diagnosis of such lesions includes: hernia, primary neoplasms, metastasis from an intra-abdominal tumor (Sister Marie-Joseph nodule), embryological residual masses (urachal cysts), or obviously endometriosis. The recommended treatment is complete excision, with excellent reported results and no recurrence [3]. The peculiarity of our case of umbilical endometriosis was that it was both primary and solitary. We had documented that the pelvis was free of disease. Steck and Helwig have reported 21 cases of umbilical endometriosis; only seven of them were explored, four of which turned out to have pelvic endometriosis [4]. On the other hand, Scott and TeLinde reported on four such cases, with only one having pelvic endometriosis [5]. The majority of the other cases were either secondary to a scar, were associated with other types of endometriosis, or had no documentation of the

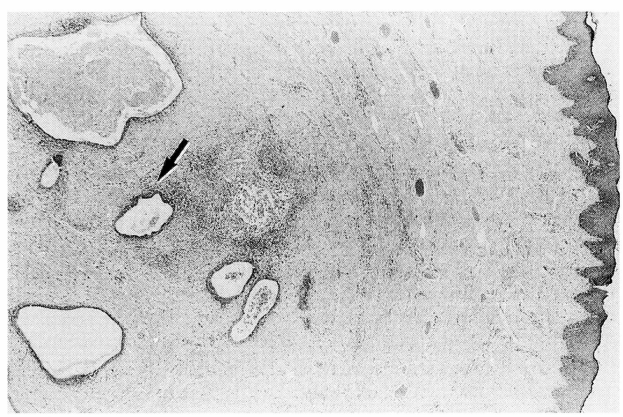

Figure 1. — Low magnification showing the skin surface with dilated glands in the deep (arrow). (H&E stain, 20 x magnification).

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status of the pelvis. This case and others pose a challenge to the accepted theories in the pathogenesis of this disease. The Sampson theory assumes that retrograde menstruation mechanically transports the endometrial fragments into the pelvic cavity through the fallopian tubes. These fragments will form the seeds for future implants. According to the metaplasia theory, all tissues of coelomic origin including the female genital tract and the peritoneum retain their potential capacity to differentiate into endometrial tissue and will do so in the proper microenvironment. One or both theories may explain our case [4]. Endometrial tissue can be transported to the umbilicus mechanically or through the lymphatics, and will not be cleared from that site because of a defect in the natural killer cells [6]. This ectopic endometrium will induce an inflammatory reaction with the release of several cytokines and growth factors [6]. Substances such as IL-8, VEGF, and integrins will induce adherence, angiogenesis and growth of these implants, as well as enhancement of the inflammation. High concentrations of these factors have been demonstrated in peritoneal fluid [7].

References

[1] Chun J. T., Nelson H. S. Jr., Maull K. I.: "Endometriosis of the abdominal wall". *South Med. J.*, 1990, *83*, 1491, 2.

[2] Singh K. K., Lessells A. M., Adam D. J. *et al.*: "Presentation of endometriosis to general surgeons: a 10-year experience". *Br. J. Surg.*, 1995, *82*, 1349, 1351.

[3] Igawa H. H., Ohura T., Sugihara T., Hosokawa M., Kawamura K., Kaneko Y.: "Umbilical endometriosis". *Ann. Plast. Surg.*, 1992, *29*, 266, 8.

[4] Steck W. D., Helwig E. B.: "Cutaneous endometriosis". *Clin. Obstet. Gynecol.*, 1966, *9*, 373, 383.

[5] Scott R. B., TeLinde R. W.: "External endometriosis-the scourge of the private patient". *Ann. Surg.*, 1958, *13*, 697, 720.

[6] Nisolle M., Donnez J.: "Peritoneal endometriosis, ovarian endometriosis, and adenomyotic nodules of the rectovaginal septum are three different entities". *Fertil. Steril.*, 1997, *68*, 585, 596.

[7] Ryan P. I., Taylor N. R.: "Endometriosis and infertility: New concepts". *Obs. Gyn. Surv.*, 1997, *52*, 365, 371.

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