Introduction

The overall prevalence of endometriosis in reproductive aged women has been suggested to be as high as 11% (Buck Louis, 2011). (1) Extra pelvic endometriosis, although often asymptomatic, should be suspected when associated with cyclical symptoms of pain or a palpable mass occur outside the pelvis. Endometriosis involving the intestinal tract (especially colon and rectum) is the most common site of extra pelvic disease and may cause abdominal and back pain, abdominal distention, cyclic rectal bleeding, constipation, and obstruction. Ureteral involvement can lead to obstruction and result in cyclic pain, dysuria, and hematuria. Pulmonary endometriosis can manifest as pneumo / hemothorax, or hemoptysis during menstruation. Umbilical endometriosis should be suspected when a patient has a palpable mass and cyclic pain in the umbilical area.

Umbilical endometriosis is rare, representing 0.5% to 1% of all endometriosis cases. Antonio Carlos Pereira Júnior was the first Brazilian to describe the disease in 1970. This kind of endometriosis can be classified as primary, when it comes “de novo” or secondary, when it comes after surgical procedures. The umbilicus, considered a physiologic scar, could have tropism for endometrial cells, as seen in other scar tissues. (2) Even today, many aspects of the disease remain enigmatic. The designation secondary endometriosis can be used even when it is not located on surgical scars, such as on the umbilicus, but its onset occurs within 2 years after the procedure. Clinically it presents as a reddish-brown nodule, commonly painful, with cyclic variations in size, with or without bleeding, that may or may not coincide with the patient’s menstrual cycle. (3)
Mrs. SP, 43-year woman, para 2, presented at surgery clinic suffering from a slowly growing swelling in the umbilicus that she noticed 9 months ago, now having bleeding for last 3 months. Initially she received some antibiotics and analgesics from a surgeon without any improvement. As the symptoms had established relationship with menstruation, she had Obgyn consultation at OBGYN clinic Bolangir, Odisha. History revealed that umbilical pain was also associated with bleeding during menstruation for the last 3 months. She had no operations and did not receive any medication for menstrual disorder. She did not describe dysmenorrhea, abdominal pain or dyspareunia. She had regular menstrual cycles and did not use oral contraceptive. The physical examination showed a dark-color sensitive nodule of 24 ×10 mm in size localized in the umbilicus [Fig. 1]. Umbilical endometriosis was suspected based on her medical history and physical examination findings. Ultrasonography showed a hypoechoic superficial mass in the umbilicus and no signs of intra-abdominal endometriosis. Pelvic examination revealed a mobile, normal size, anteverted uterus. Fornices were free and non-tender.

The patient was counselled. She was informed about possible modalities of treatment both medical and surgical. She opted for surgery. Preoperative hemogram, biochemical and coagulation profile was normal. Laparoscopic examination didn’t show any pelvic endometriotic lesion in the pelvis. Excision of umbilicus was done [Fig. 1]. Postoperative period was uneventful. Histological study confirms diagnosis of umbilical endometriosis [Fig. 2].
Fig. 2 Spindle-shaped fibroblastic cells, numerous benign glandular endometrial glands

Discussion

Extra-pelvic locations of endometriosis have been described in almost every tissue and organ (i.e. gastro-intestinal apparatus, abdominal organs, skin, diaphragm, lung and even brain). Primary (spontaneous) umbilical endometriosis was first defined by Villar in 1886. It accounts for 0.5–1% of all cases of extra genital endometriosis [3]. The pathogenesis of endometriosis has been a much-debated issue. According to hypothesis proposed by Sampson in 1920, endometriosis was caused by retrograde menstruation passing thorough the fallopian tube in the pelvis [4]. However, there are other theories such as coelomic metaplasia, direct spread, iatrogenic spread, lymphatic or hematogenous spread [2]. The theory of lymphatic and hematogenic transplantation is suggested for cases of umbilical endometriosis with pelvic endometriosis. But the disease may occur through metaplasia of urachus residues in a case of isolated umbilical endometriosis [2]. Secondary umbilical endometriosis may occur through iatrogenic spread of endometrial cells after operations such as cesarean section and laparoscopy [4]. In many cases of primary umbilical endometriosis, there is an umbilical nodule concurrent with menstruation, which causes periodic pain in the umbilicus and may have a bleeding tendency. There may be a constant pain rather than periodic pain. In absence of menstruation i.e. pregnancy and menopause diagnosis become more difficult. Spontaneous umbilical endometriosis is a rare disease that can worsen during pregnancy (1). The nodule can be a brown, blue or faint spot. The differential diagnosis for umbilical lesions includes pyogenic granuloma, umbilical polyp, melanocytic nevus, seborrheic keratosis, epithelial inclusion cyst, desmoid tumor, hemangioma and granular cell tumor (4). Other differential diagnoses can include umbilical hernia, omphalitis, keloid and foreign body granuloma (3). Malignant lesions such as melanoma, adenocarcinoma, squamous and basal cell carcinoma, and metastases from the gastrointestinal tract should be ruled out (2). Endometriosis, therefore, is not frequently high on the plastic surgeon’s differential diagnosis for solitary umbilical nodules.
Sensitivity and specificity of ultrasonography, computerized tomography and magnetic resonance are low in the diagnosis of umbilical endometriosis. None of these imaging techniques exhibit any pathognomonic sign of umbilical endometriosis. These can provide some information on the size of the nodule and its adherence to adjacent tissues [4]. Treatment of umbilical endometriosis has not been standardized yet, due to limited number of cases. The medical treatment including progesterone, danazol, nor-ethyolsterone, and GNRH analogues have not provided reliable results. However, some authors have reported success in reducing size of nodule and improvement of symptoms [2, 4]. In the review by Victory et al., almost 70% of patients required surgical treatment [4]. Surgical choices include either total umbilical resection with or without repair of underneath fascia and peritoneum, or local excision of endometrial nodule with preserving umbilicus. Total resection of umbilicus is mostly preferred. Local excision of endometrial lesion should be performed by achieving adequate edge of the surrounding normal tissue in order to avoid local recurrence. Several authors suggested total umbilical excision independent of the size of endometrial nodule.

Conclusion

Primary umbilical endometriosis should always be considered as a potential diagnosis in women who have painful, sometimes discolored umbilical swelling, and then only it can be identified earlier and treated optimally. Total umbilical resection should be preferred to avoid local recurrence.

Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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References


