CASE REPORT

Spontaneous Primary Umbilical Endometriosis Preceding Severe Pelvic Endometriosis: A Case Report

Sujata Datta[1], Ritisha Basu[2]

Abstract

Primary spontaneous umbilical endometriosis accounts for a minority of the cases of umbilical endometriosis, the incidence of which is 0.5–1% of all extragenital endometriosis. Cyclical umbilical bleeding without any accompanying symptom of pelvic endometriosis or any prior history of surgery is an extremely rare condition that throws up diagnostic challenges, in the absence of overt clinical signs. We present a case of solitary primary umbilical endometriosis, which progressed to severe grade 4 endometriosis over a span of 4 years for poor patient compliance. Imaging difficulties as well as laparoscopic challenges in therapy, especially with the insertion of the primary port in the presence of an umbilical endometriotic nodule, are also discussed.

Keywords: Cutaneous sinus, Umbilical bleeding, Umbilical endometriosis, Umbilical nodule.

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Introduction

Endometriosis presenting primarily as cyclical umbilical bleeding without any accompanying symptom of pelvic endometriosis or any prior history of surgery is an extremely rare condition. It throws up diagnostic challenges, in the absence of overt clinical signs and unsuccessful imaging in small lesions, especially during the intermenstrual period. It accounts for a minority of the cases of umbilical endometriosis, the incidence of which is 0.5–1% of all extragenital endometriosis.[1] Treatment includes wide local excision of endometriotic nodule and treatment of accompanying pelvic disease if present. Laparoscopic challenges are encountered during the insertion of the primary umbilical port in the presence of an endometriotic nodule and in ensuring complete excision to prevent future recurrence.

Case Report

A 41-year-old lady with two spontaneous vaginal deliveries in the past presented with a sudden onset of cyclical umbilical bleeding during menstruation for 4 months. On physical examination, there was no visible umbilical nodule. Her menstrual cycles were regular and painless. She had no prior history of abdominal surgery. Her pelvic ultrasound was normal. Her menstrual cycles were suppressed with a 3-month course of medroxyprogesterone acetate. The induced amenorrhea simultaneously suppressed umbilical blood loss, clinically clinching the diagnosis of primary umbilical endometriosis.

However, she was noncompliant with medication and was lost to follow-up. She presented 4 years later with severe dysmenorrhea and painful irregular umbilical blood loss. A nodule was now palpable at the base of the umbilicus during menstruation. A transvaginal ultrasound done again revealed severe pelvic endometriosis evidenced by bilateral chocolate cysts measuring approximately 5 cm each. Magnetic resonance imaging (MRI) of the umbilicus revealed a diffuse restricted nodular lesion of 1.2 cm at the base of the umbilicus, although an ultrasound between menstrual cycles failed to pick up the small lesion.

[1,2]Department of Obstetrics and Gynaecology, Fortis Hospital, Kolkata, West Bengal, India

Corresponding Author: Ritisha Basu, Department of Obstetrics and Gynaecology, Fortis Hospital, Kolkata, West Bengal, India, Phone: +91 8910560163, e-mail: basu.ritisha312@gmail.com

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At surgery, a deep nodule with a cutaneous sinus was found at the base of the umbilicus. She was treated with wide excision of the umbilical nodule using monopolar diathermy, with a 1 cm margin of dissection. The dissection was carried up to the rectus sheath and peritoneum.

Concomitant laparoscopy was performed through an umbilical port introduced at the lower end of the umbilicus, after dissecting the endometriotic nodule off the underlying rectus sheath. Port entry was done prior to complete excision of the nodule. A total laparoscopic hysterectomy with bilateral salpingo-oophorectomy was performed for grade 4 endometriosis with dissection of rectal adhesions in the pouch of Douglas. Complete excision of the umbilical endometriotic nodule (Fig. 1) was done under laparoscopic guidance to exclude peritoneal extension of the lesion. The rectus sheath at umbilicus was repaired with continuous Vicryl suture. Reconstruction of umbilicus was performed. Histopathological examination confirmed umbilical endometriosis (Fig. 2).

Discussion

Secondary endometriosis of the umbilical port site (scar endometriosis) following laparoscopic treatment of endometriotic disease is more common than spontaneous primary endometriosis of the umbilicus. Our case of spontaneous umbilical endometriosis

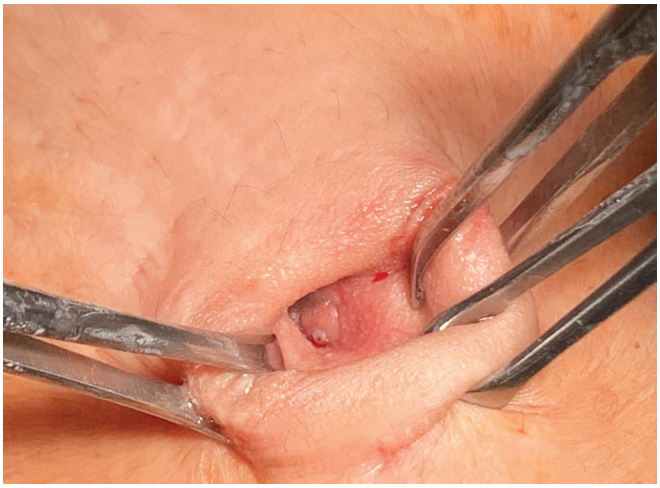

Fig. 1: Umbilical endometriotic nodule

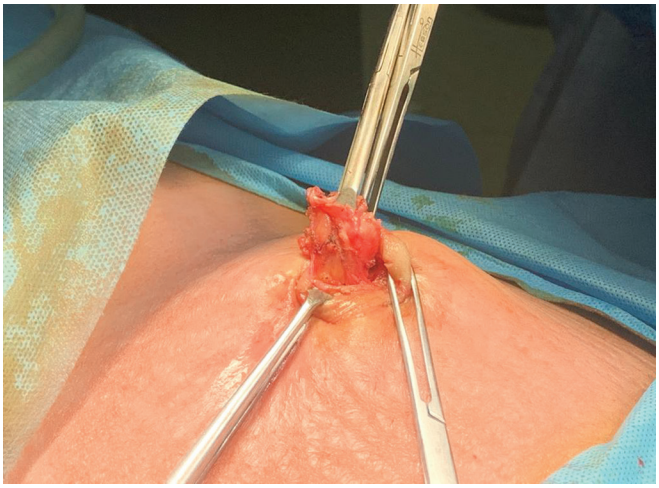

Fig. 2: Wide local excision of umbilical endometriotic nodule

can be considered as a scar endometriosis in a physiological scar[2] or metaplasia from the remnants of the urachus.[3]

The typical presentation of umbilical endometriosis is that of a painful umbilical nodule that bleeds simultaneously during menstruation. Diagnosis can be difficult in the absence of a visible or palpable umbilical nodule and a negative ultrasound in small lesions but can be made from a suggestive history, a trial of suppressive medical therapy and MRI if available.

Medical therapy with GnRH analogs, progesterone, and the oral contraceptive pill can be used for temporary disease suppression but is not curative. The definitive treatment is excision of the nodule with a wide margin to minimize recurrence.[4] If needed, an umbilectomy should be discussed preoperatively. Excision should be carried down to the rectus sheath to avoid residual disease.

Spontaneous primary umbilical endometriosis can also progress to involve pelvis. As in our case, an accompanying laparoscopy was used to treat the coexistent pelvic endometriosis.

Conclusion

Our case highlights diagnostic difficulty in evaluation of early spontaneous primary umbilical endometriosis, and we explain how a careful history with a simple trial of medical therapy can be helpful. The second point of importance is the description of the technique of wide excision and how a simultaneous laparoscopy following dissection of the endometriotic nodule off the rectus sheath is used to exclude peritoneal extension of the umbilical nodule, to guide complete excision, and to prevent recurrence.

Consent for Publication

Written informed consent was taken from patient for publication of the case and accompanying images.

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