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Case Report

Menstruating umbilicus: secondary subcutaneous umbilical endometriosis: a rare case report

Rakesh A. Hasabe^{1*}, Neha S. Tripathi², Prajakta Kesarkodi¹

¹B. K. L. Walawalkar Rural Medical College and Hospital, Ratnagiri, Maharashtra, India

²Wingate University, Wingate, NC, USA

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***Correspondence:**

Dr. Rakesh A. Hasabe,

E-mail: rakesh.hasabe@gmail.com

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ABSTRACT

This paper described a rare case of secondary umbilical endometriosis in a young nulliparous female and its effective diagnosis and treatment. A 29-year-old unmarried female presented with complaints of bleeding during menses from an umbilical nodule that had developed at the port insertion site of a previous diagnostic hystero-laparoscopy. Ultrasound and Doppler examinations confirmed the presence of a hypoechoic mass in the umbilicus with no associated blood vessels. Deep surgical excision of the nodule with a rim of macroscopic normal skin of 0.5 cm all around was performed under local anesthesia and the umbilicus was reconstructed. This case reinforced the concept that umbilical endometriosis should be considered in the list of differential diagnoses of umbilical disorders, even in young nulliparous women with no typical symptoms of pelvic endometriosis. Additionally, it emphasized the importance of early diagnosis of UE in order to avoid extensive abdominal wall surgery.

Keywords: Umbilical endometriosis, Menstruating umbilicus, Subcutaneous umbilical endometriosis

INTRODUCTION

When endometrial tissue is found to exist anywhere outside the uterus, the condition is defined as endometriosis. Existing literature reports findings of endometrial tissue in almost every type of extra-pelvic tissue and organ, including but not limited to the abdomen, skin, lung, diaphragm, brain and gastrointestinal system.^{2,3} Endometriosis is theorized to be caused due to direct, lymphatic or hematogenic spread as well as by retrograde menstruation, embryonal rest or coelomic metaplasia. The exact pathogenesis remains unknown. An extremely rare type of endometriosis, umbilical endometriosis (UE) occurs in about 0.5% to 1% of all diagnosed cases of extragenital endometriosis.^{1,4,5}

Depending on etiology, UE is usually classified as primary or secondary. Villar first described primary UE in 1886, because of which this type of UE is also named Villar's

nodule. Primary UE develops spontaneously and is the more common type with an incidence of 75% of all diagnosed cases of UE, although its pathogenesis is not entirely known.⁶ Secondary UE, on the other hand, is usually a rare complication that occurs after laparoscopic or open procedures that involve the umbilicus or immediate surrounding area. Primary and secondary UE may be cutaneous or subcutaneous.⁷

CASE REPORT

A 29 year old married nulligravida female presented at our outpatient clinic with a 3-year history of an umbilical nodule. She stated that the nodule had slowly increased in size and had started to bleed concomitantly with menstrual periods in the previous 2 years. Her medical history was unremarkable but she reported symptoms indicative of pelvic endometriosis such as dysmenorrhea and abdominal pain. She had undergone diagnostic hystero-laparoscopy

as part of an infertility workup around 4 years ago, which consisted of primary 10 mm port insertion at the umbilicus at the exact same site where she later developed the nodule. Physical examination revealed a brown, moderately tender nodule of about 2×2 cm in diameter located deep in the umbilical fold.



Figure 1: UE nodule.

On the basis of history and clinical findings, secondary umbilical endometriosis was suspected and the patient was asked to return for further examination during her menstrual period, which occurred after three weeks. At this follow up, the umbilical nodule appeared more tender, showing signs of recent bleeding. An ultrasound confirmed the presence of a hypoechoic mass of 20 mm in the umbilicus with no associated blood vessels confirmed by Doppler examination. Thus, surgical removal of the umbilical nodule was proposed and the patient was informed about the risk of local recurrence. The patient underwent excision of the nodule under local anesthesia. The lesion was entirely excised deep to the fascia, together with a rim of macroscopic normal skin of 0.5 cm all around. There was no evidence of connection with the peritoneal cavity and the umbilicus was reconstructed with discontinuous suture using non-absorbable stitches.

On gross examination of the excised specimen, a nodular lesion of 2×2 cm covered by normal skin was appreciable. For light microscopic examination, the specimen was fixed in 10% buffered formalin and embedded in paraffin. Microscopically, histologic sections revealed a glandular proliferation of mono-layered endometrial epithelium surrounded by a cytotroma with extravasated erythrocytes. On immuno-histochemistry, the epithelial and stromal cells showed a nuclear immuno-reactivity for ER and PR. All these features were consistent with the diagnosis of UE.

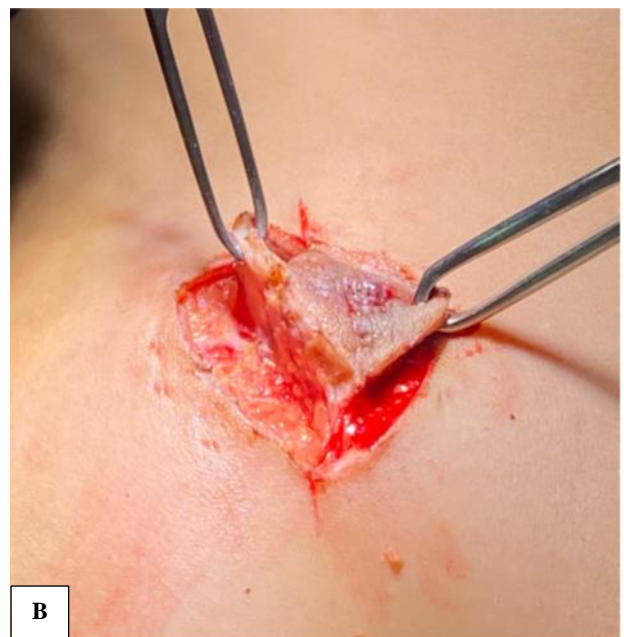


Figure 2: (A and B) Surgical excision of endometriotic nodule.

The patient's post-operative recovery was uneventful. Following the operation, the patient was started on GnRH agonists for three cycles. After 1.5 years of regular follow ups, there were neither any signs of local relapse nor other clinical or ultrasonographic signs of endometriosis.

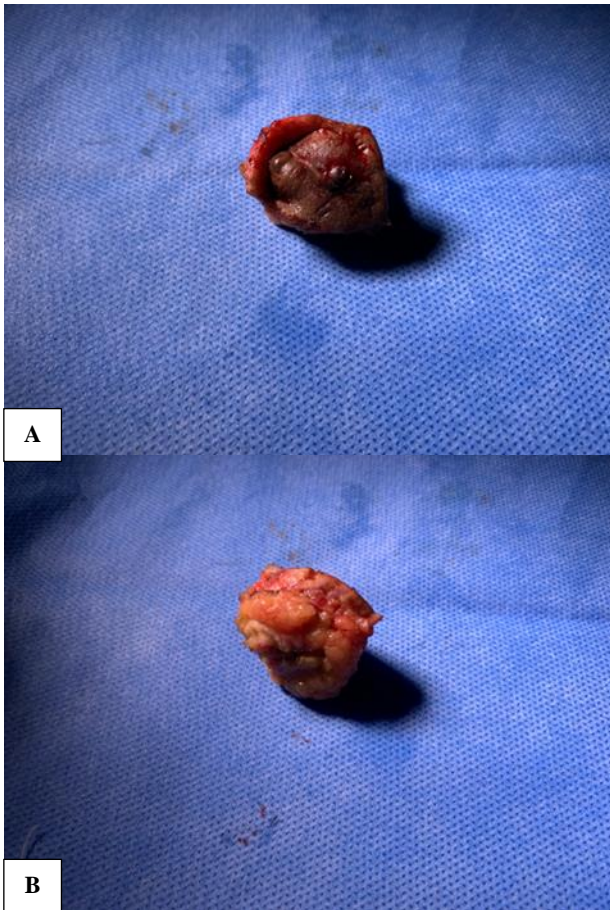


Figure 3: (A and B) Excised surgical nodule with free margins.

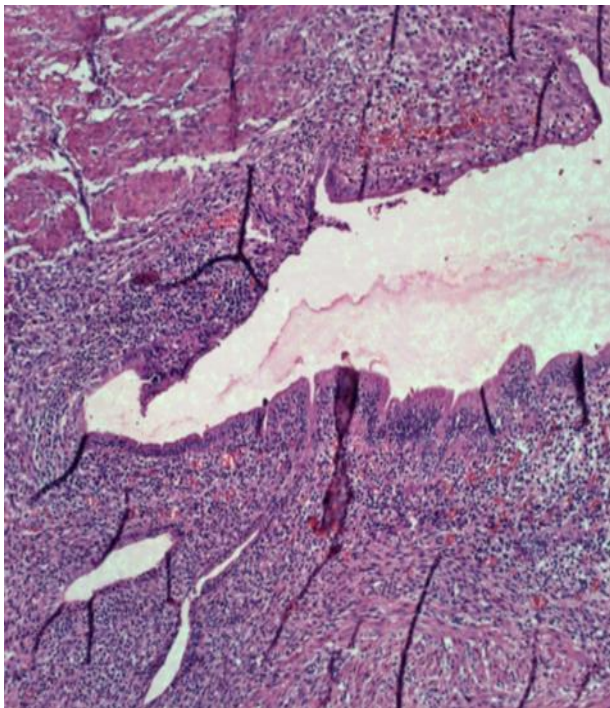


Figure 4: Histopathological slide showing endometrial glands and stroma.



Figure 5: Reconstructed umbilicus.

DISCUSSION

Primary UE is reported fairly commonly in 30-40% of all cutaneous forms of extragenital endometriosis.⁸ The pathogenesis of primary UE had been explained by various theories in literature, the retrograde menstruation theory, the embryonic remnant theory, the celomic metaplasia theory, the migration theory or a combination of them.⁹ Sampson's theory theorized that retrograde menstruation caused migration of endometrial tissue through the Fallopian tubes into the abdomen giving rise to primary UE. The embryonic rest theory assumed that cell rested developing from the Wolffian and Müllerian duct system undergo differentiation resulting in primary UE. The celomic metaplasia theory believed in the process of metaplastic conversion of celomic mesothelium-based peritoneal cells into endometrial tissue under the influence of outside stimuli. The migration theory proposed the dissemination of endometrial cells via vascular and lymphatic passages into remote, extra-pelvic and remote locations, where implantation and proliferation occurred.¹⁰ Primary UE had been theorized to be estrogen-dependent, occurring more commonly after C-sections. Literature theorized the migration of endometrial cells into the umbilical area via the lymphatic system, abdominal route, umbilical blood vessels or the urachus.¹¹ While the etiology of primary UE remained unknown, secondary UE was established to arise from post-surgical scar tissue. Malignant transformation of UE had only been documented in two cases in literature thus far, Lauslahti in 1972 and Obata et al in 2013, leading to the conclusion that the risk of malignant transformation from umbilical endometriosis was very low.^{12,13}

In most cases, UE clinically presented with umbilical swelling (90% of cases), often associated with cyclical pain (81.5%) and bleeding (49.2%). The differential diagnoses may include granuloma, umbilical polyps, hemangioma, melanocytic nevus, seborrheic keratosis, granular cell tumor and umbilical hernia. Suspected UE may be investigated with an ultrasound or MRI to evaluate echogenicity and vascular involvement. Medically,

hormonal treatments such with contraceptive pills or GnRH analogues may alleviate symptoms temporarily; however symptom recurrence was commonly seen after hormonal treatment ceases.¹⁴ The definitive management in UE was thus surgery, varying from superficial diathermy to laparoscopic omphalectomy.

Due to the extent and size of the endometriosis nodule, total removal of the umbilicus was often the choice of surgical intervention, even at the cost of poor cosmetic results due to mesh placement for wall reconstruction, especially in long standing cases of umbilical endometriosis, when adjacent abdominal structures may be involved or when UE was associated with umbilical hernias.¹⁵ Local endometriosis lesion excision with an adequate rim of normal tissue was recommended, in order to avoid recurrence.¹⁵

In our opinion, the case described in this paper was of significance on various fronts. To our knowledge, this patient was one of the youngest women with secondary UE reported in literature. This case reinforced the concept that umbilical endometriosis should be considered in the list of differential diagnoses of umbilical disorders, even in young nulliparous women with no typical symptoms of pelvic endometriosis. Moreover, it emphasized the importance of early diagnosis of UE in order to avoid extensive abdominal wall surgery.

CONCLUSION

Based on our case study and literature review, we believe that for small primary or secondary endometriotic nodules without involvement of the musculo-aponeurotic plane and other endometriotic localizations, partial/complete omphalectomy (local excision of the umbilical endometrial nodule) with a 3-5 mm free border, even without adjuvant hormonal treatment, could ensure adequate and effective treatment with favorable long-term outcomes.

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