A multilobulated asymptomatic umbilical nodule revealing endometriosis

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Abstract
Primary umbilical endometriosis is unusual clinical presentation of endometriosis. Its diagnosis can be challenging due to lack of knowledge. This condition should be listed in the differential diagnosis of umbilical disorders.

KEYWORDS
cutaneous endometriosis, extrapelvic, nodule, umbilicus

1 | INTRODUCTION
Cutaneous endometriosis is a rare disorder characterized by the presence of ectopic functional endometrial tissues in the abdominal wall. Primary umbilical endometriosis is unusual clinical presentation often misdiagnosed due to its rarity and lack of knowledge.

Endometriosis is a chronic rare disorder characterized by the presence of ectopic functional endometrial tissues outside the uterine cavity, such as lung, genital and urinary tracts, surgical scars, or umbilicus. This condition affects 6%-10% of women in the reproductive age.1 Umbilical endometriosis, also known as Villar’s nodule, is quite rare and usually occurs secondary to surgical scars. Primary umbilical endometriosis is, however, unusual clinical presentation and often misdiagnosed due to its rarity and lack of knowledge.

2 | CASE REPORT
A 33-year-old woman was referred to our department for a painless and progressively enlarging nodule of the umbilicus of 3 years’ duration. She had no past history of any surgery or trauma of the abdomen. She did not use oral contraceptives and had regular menstrual cycles with normal flow. The patient complained of dysmenorrhea since the appearance of the nodule. She did not describe other symptoms such as menorrhagia, hematuria, pelvic pain, or dyspareunia. Besides, there was no periodic bleeding from the umbilicus. Physical examination revealed a firm, red to violaceous

FIGURE 1 A multilobulated red to violaceous nodule of the umbilicus
multilobulated nodule of 20 × 25 mm in size protruding from her umbilicus (Figure 1). A biopsy of the nodule revealed the presence of multiple endometrial glandular tubes surrounded by an endometrial-type stroma without signs of malignancy (Figure 2A,B). Thus, the diagnosis of primary umbilical endometriosis was assessed. Surgical resection was planned for the patient.

3 | DISCUSSION

Cutaneous endometriosis is the most common extrapelvic endometriosis. It was first described by Rokitansky in 1860 and defined by the presence of an extruterine functional endometrial tissue formation in the skin. It is classified in a primary and secondary form. Secondary endometriosis is the most common and occurs often after surgery on abdominal and pelvic scars such as cesarean section, hysterectomy, and laparoscopy.

Primary umbilical endometriosis (PUE) occurs spontaneously and is a very rare benign condition. The incidence of this disease is about 0.5 to 1% of extrapelvic endometriosis. It mostly occurs in female of reproductive age and is usually characterized by a discolored painful mass of the umbilicus with cyclic bleeding and/or swelling according to the menstrual cycle. In the study of Santos and coworkers, the mean age was 33 years; all patients had a complaint from pain and bleeding in the menstruation period. However, asymptomatic PUE is much less common, as reported in our patient. Hence, it may go unrecognized leading to a delayed diagnosis.

In the literature, the size of the lesion ranges from 0.5 to 4 cm. The nodule may be brownish, violaceous, dark bluish, or flesh colored, depending on the amount of hemorrhage and depth of penetration of ectopic endometrial tissue. In the review of Victory et al, the majority of patients presented these lesions colors: brown, blue, purple, black, and red. And in the study of Santos, 83% of lesions were violaceous and erythematous-red in 16%.

Differential diagnoses should include keloid, umbilical hernia, granuloma, sebaceous cyst, and urachus anomaly. Malignant diseases such as nodular melanoma, lymphoma, and Sister Marie Joseph's nodule should also be considered. Histopathological findings remain the gold standard for the definitive assessment of PUE and to exclude malignancy. It shows dilated intradermal endometrial glands surrounded by cellular endometrial-type stroma. Magnetic resonance imaging can be helpful, although no imaging tools are necessary to retain the diagnosis.

Dermoscopy features are controversial but could be helpful to distinguish from malignant Sister Mary Joseph nodule. Surgical excision remains the most effective treatment to reduce the risk of malignant transformation and to avoid recurrence. Medical treatment with oral contraceptives, such as progesterone, Danazol, norethisterone, or GnRH analogs, may help in reducing the size of the lesion and is often used as a diagnostic tool itself (anne 2017).

Pathogenesis of primary cutaneous endometriosis is still unclear. Endometrial cells might migrate to the umbilicus through the abdomen, the lymphatic system, and/or remnants of embryonic cells in the umbilical fold.

In summary, our case describes an unusual location of endometriosis. This condition should be listed in the differential diagnosis of umbilical disorders.

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CONFLICT OF INTEREST
None declared.

AUTHOR CONTRIBUTIONS
Drs Amal Chamli and Asmahene Souissi: contributed to the first draft of the manuscript. Drs Amal Chamli, Asmahene Souissi, Fatima Aloui, Ines Chelly, and Dalenda Eleuch: contributed to the literature search, analysis, and interpretation of the data. Dr Mourad Mokni: revised the manuscript and
gave final approval. All authors: read and approved the final manuscript and agree to be finally accountable for ensuring the integrity of and accuracy of the work.

ETHICAL APPROVAL
Appropriate consent has been obtained, prior to submission, for the publication of images and data.

DATA AVAILABILITY STATEMENT
Data sharing was not applicable to this article as no datasets were generated or analyzed during the current study.

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