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Case Report

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Umbilical Endometriosis: A Case Report

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Abstract

We report the case of a 31 year old female patient with no history of abdominal-pelvic surgery or trauma to the umbilicus, consulted for a painful umbilical swelling measuring approximately 1 to 2 cm diagnosed with umbilical endometriosis.

Introduction

Endometriosis is a condition characterised by the presence of ectopic endometrial structures which have the histological and biological characteristics of the endometrium, but which remain anatomically separate from it. It affects women during their genital period. [1] Cutaneous involvement is very rare and represents only 0.5 to 1% in the various series.

It is defined by the presence of functional endometrial tissue in the umbilicus. Two types of umbilical endometriosis can be distinguished, the pathophysiology of which still remains a challenge for research. The primary form appears on an abdomen unaffected by any surgical intervention and the secondary form develops on a scar following a gyneco-obstetric intervention or laparoscopy. Its clinical diagnosis is difficult but it should be suspected in the presence of any bluish, painful umbilical nodule, sometimes with a brownish discharge, the evolution of which is regulated by the menstrual cycle. Ultrasound of the abdominal wall points to the diagnosis of umbilical endometriosis despite the absence of characteristic signs on imaging. Wide surgical excision is the treatment of choice because of its resemblance to a primary tumour or metastasis. We report a case of umbilical endometriosis explored by ultrasound. Through the analysis of this rare observation, we highlight the difficulties related to clinical and ultrasound diagnosis and advanced theories of the pathophysiology of umbilical endometriosis [3].

Case report

A 31-year-old woman, multiparous, with no history of abdominal-pelvic surgery or trauma to the umbilicus, consulted for a painful umbilical swelling measuring approximately 1 to 2 cm, which had been evolving for two years, becoming purplish and sensitive at the beginning of menstruation (figure 2), and causing a minimal discharge of brownish fluid, thick at the end of menstruation. She had been experiencing mild dysmenorrhoea for several years, with inconstant deep dyspareunia.

On clinical examination, a bluish nodule about 1 cm in diameter was found at the bottom of the umbilical depression, which was tender to palpation (Figure 1). The rest of the abdominal examination was normal.

The gynaecological examination was normal. The cyclical nature of the bleeding and the symptomatology, which was accompanied by menstruation, made us suspect a primary umbilical endometriosis. Ultrasound of the abdominal wall: skin thickening at the level of the umbilicus with a 5 mm umbilical nodule, well limited and finely echogenic, suggesting an endometrial nodule (Figure 3). Pelvic MRI scan did not identify any lesions of pelvic endometriosis.

Skin histology showed papillomatous acanthosic epidermis with orthokeratosis. The dermis has an interstitial infiltrate with siderophages and haemosiderin deposits. With the presence of a glandular structure partially represented as a regular cubocylindrical epithelium without atypia (figure 4 to 7). The diagnosis of primary umbilical endometriosis is retained.

As a result, treatment with leuprolide acetate was instituted and surgical removal was recommended as a secondary procedure. After one month of leuprolide (LHRH analogue), the patient noted a disappearance of menstrual symptoms (pain, bleeding and abdominal swelling) and she is awaiting surgery after two months of treatment.

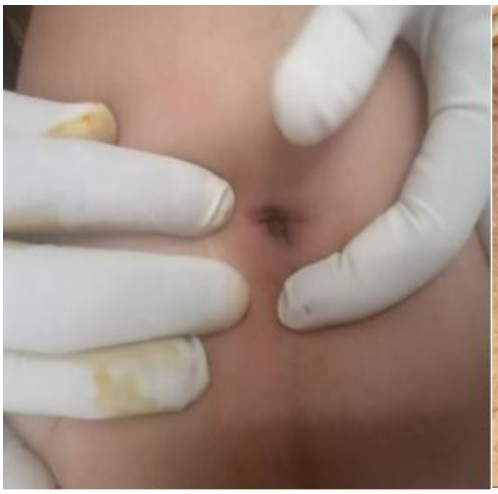

Figure 1

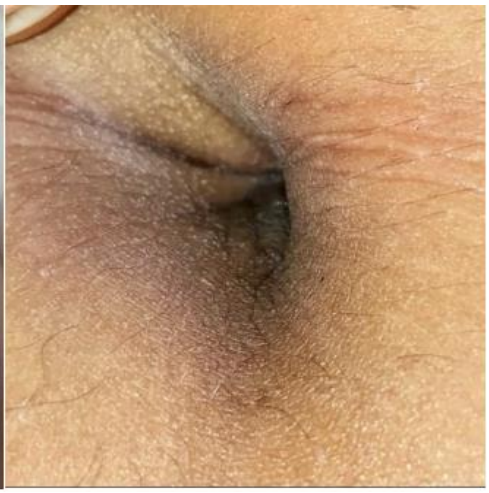

Figure 2

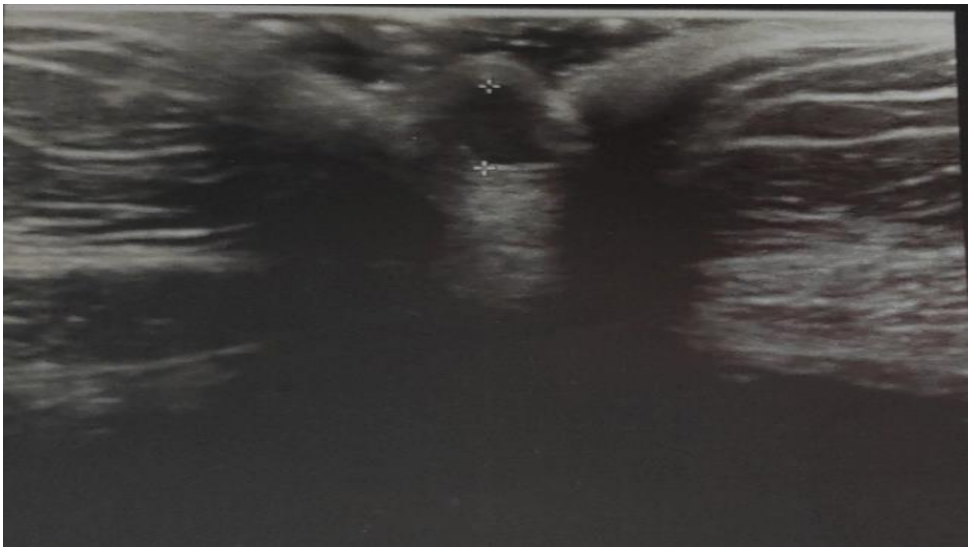

Figure 3

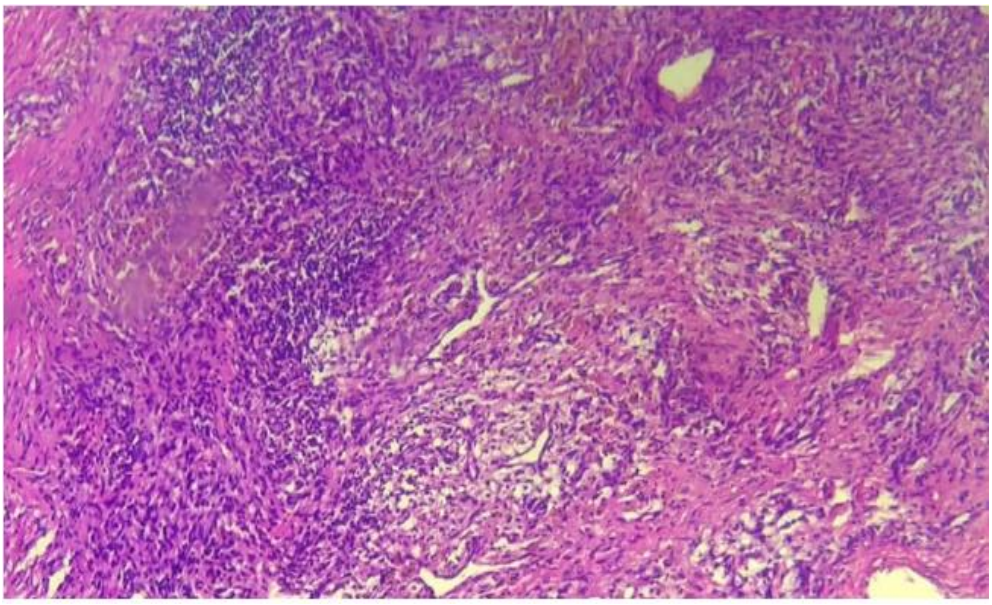

Figure 4: Présence d'infiltrat inflammatoire interstitiel avec des sidérophages et des dépôts d'hémosidérine.

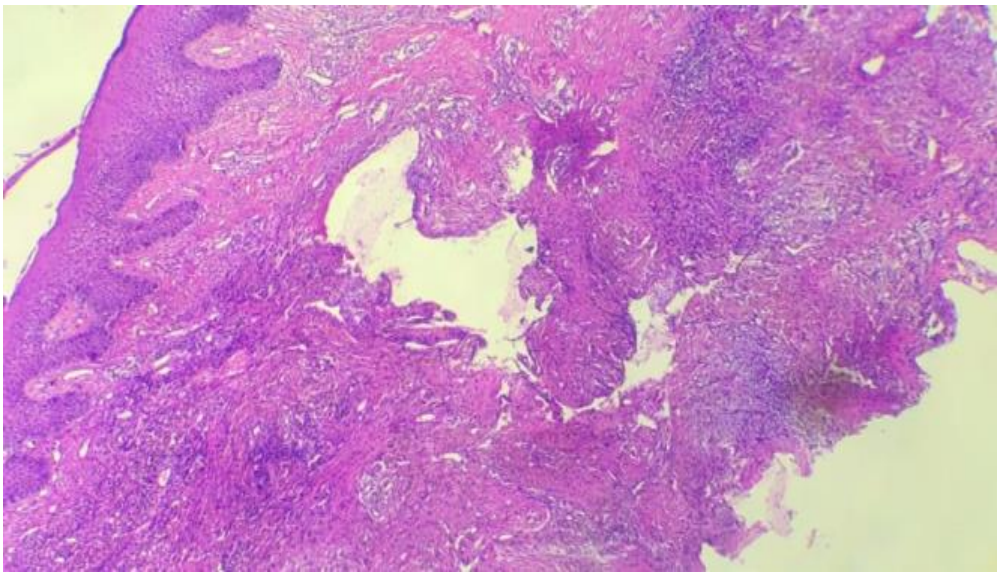

Figure5 : Épiderme hyperplasie papillomateux surmonté d'une orthokeratose

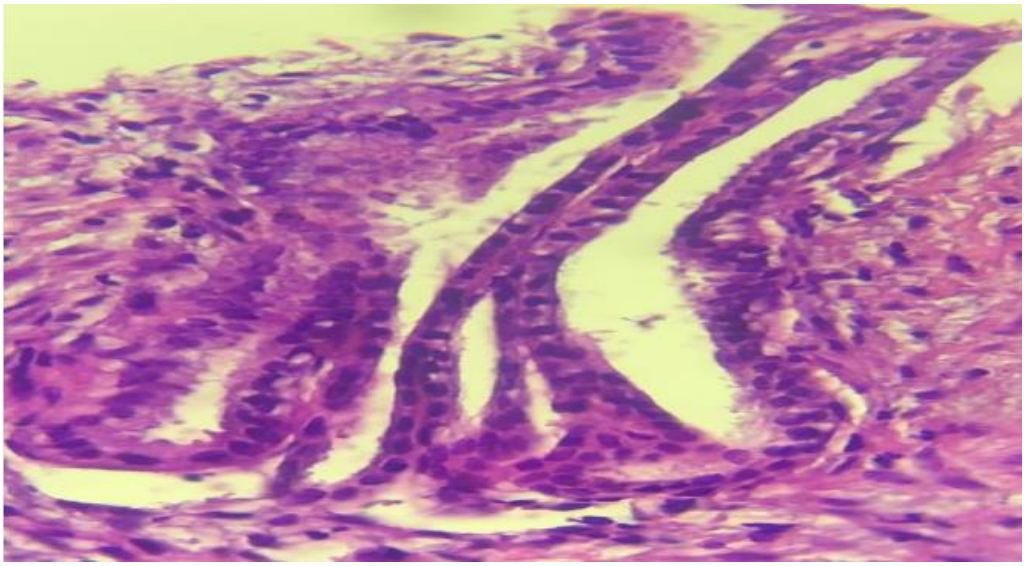

Figure 6 : Présence de quelques glandes bordées d'un épithelium cubocylindrique régulier sans atypies

Discussion

Endometriosis is characterised by the presence of functional endometrial tissue outside the uterine cavity, and skin involvement is very rare, representing only 0.5 to 1% in the various series. It affects 15% of women in the genitally active period between the ages of 20 and 40 and 5% of cases are discovered in postmenopausal women. The usual locations of endometriosis are ovarian, peritoneal, in the partition separating the bladder, the uterus and the vagina or the rectum (3)

A distinction is made between internal endometriosis or adenomyosis, which is the implantation of endometrial cells in the myometrium, and external endometriosis, which is the location of endometriotic tissue outside the myometrium.
endometriotic tissue outside the uterine cavity and myometrium.

Extra-genital locations are thought to represent 5% of lesions and seem to be underestimated in the literature. The existence of these lesions often calls for different physiopathological theories to explain their extra-pelvic location [1]. The lesions may be multiple or single, with a wide variety of sites. Diagnosis can be difficult due to the atypical and unexpected symptoms described by patients. However, the catamenial nature of the pain or symptoms is a characteristic and highly suggestive feature regardless of location. It is essential to make the diagnosis, as these conditions can also have a real impact on the patient's health and quality of life (4).

Endometriosis of the abdominal wall, particularly the umbilical wall, is a rare condition, representing only 0.03 to 2% of extra-genital endometriosis. It is associated with pelvic endometriosis in 26% of cases

Two types of umbilical endometriosis can be distinguished: primary endometriosis, which is exceptional, occurs in women with no history of abdominal surgery, and secondary endometriosis, which is uncommon, appears on a scar following a gynaeco-obstetric operation or on the site where the laparoscopic trocar passes (5)

The umbilical location could also be explained by the venous or lymphatic metastatic theory, when endometriotic cells migrate to the umbilicus through the periumbilical venous network. Finally, according to a third theory, that of metaplasia, cells derived from the coelomic epithelium undergo metaplasia towards endometrial cells, under the effect of various infectious, toxic or hormonal factors (6)

The most common differential diagnoses with umbilical endometriosis include umbilical hernia, primary malignancy such as melanoma or metastasis, lipoma, inflammatory or infectious granuloma, complicating cyst, congenital cyst of the urachus and endometriosis [18]. Umbilical hernia is a real dilemma in acute abdominal symptomatology¨, as in the case of one of our patients. Ultrasound of the abdominal wall with a high frequency probe is the initial, easily accessible examination that points to the diagnosis of umbilical endometriosis, but it is not pathognomonic; it confirms the presence of an umbilical nodule, specifies its size, its contours, its limits with the adjacent superficial and deep structures, its homo or heterogeneous content, its solid or cystic nature (7).

Endometriosis of the abdominal wall, particularly the umbilical wall, is a rare condition, representing only 0.03 to 2% of extra-genital endometriosis. It is associated with pelvic endometriosis in 26% of cases

Percutaneous biopsy of the nodule evokes the diagnosis by showing a cylindrical glandular epithelium surrounded by a stroma. A study by Catalina-Fernandez et al. showed that in cytological smears of cutaneous endometriosis there is a high cellularity containing macrophages loaded with haemosiderin and epithelial cells on old bleeding; it is still contraindicated by some authors because of the increased risk of dissemination in cases of suspected endometriosis (8).

Finally, the formal diagnosis of umbilical endometriosis is only obtained with the help of histological examination. The presence of endometrial glands (epithelial cells) and a cytogenic chorion in the ectopic endometrial tissue is necessary to establish the histological diagnosis. It consists of small foci of inflamed endometrial tissue and hemosiderin-laden macrophages secondary to acute and chronic bleeding [24]. The typical histological appearance of endometriosis excludes a primary malignant tumour, umbilical metastasis or a benign lesion (4).

The treatment of choice for umbilical endometriosis is wide surgical excision because of its similarity to malignant tumours and to avoid recurrence. Malignant transformation into carcinoma of endometriotic nodules is rare.

In general, medical treatment with danazol, norethisterone or LHRH analogues is recommended before surgery. It would allow a reduction in the size of the endometriotic nodules (9).

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