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ENDGAMES

CASE REPORT

An umbilical nodule with cyclical changes

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A 47 year old non-parous woman who had never had abdominal surgery was admitted with a four year history of an intermittently palpable nodule in the umbilical region. Initially, she noticed an increase in lesional size and pressure pain, which resolved within a few days. Six months before admission, blue-black discoloration appeared in the umbilical area; this vanished within a week but reappeared cyclically, along with an increase in size of the nodule and localised discomfort.

Macroscopic inspection during menstruation showed a sharply confined blue-black macule within the umbilical area. Lesional palpation identified a circumscribed, firm, well relocatable pea sized nodule just below the umbilical skin. Dermoscopy of the area showed features corresponding to a cutaneous haematoma.

Questions

- 1 What is the likely diagnosis?
- 2 Which differential diagnoses have to be considered?
- 3 Which diagnostic procedure(s) would be appropriate?
- 4 What kind of treatment would you recommend?

Answers

1 What is the likely diagnosis?

Short answer

Umbilical endometriosis.

Long answer

Figure 1 shows the umbilical area with a discrete central blue-black macule. The occurrence of this visible and palpable lesion with menses is characteristic of cutaneous umbilical endometriosis.



Fig 1 Abdominal region with umbilical haematoma during menses

Endometriosis is defined as the presence of endometrial tissue outside the uterine cavity. It occurs in 6-10% of women in the reproductive age group.¹ Regions with endometrial tissue include pelvic organs—especially the ovaries, the uterosacral ligaments, pouch of Douglas, bowel, bladder, diaphragm, pleura, pericardium, brain, and skin.²⁻⁷

The most pronounced symptoms of endometriosis are dysmenorrhoea, menorrhagia, pelvic pain, dyspareunia, infertility, and bowel irregularities.¹ Depression occurs in 23.5-86% of patients with endometriosis and pelvic pain (13-38% in those without pain).⁸⁻¹⁰

With regard to cutaneous lesions, endometriosis occurs most commonly in scars after surgical interventions.¹¹ Umbilical endometriosis is rare, being seen in only 0.5-1.0% of patients with endometriosis.¹²⁻¹⁴ Patients may report cyclical localised swelling, pain, discharge, or bleeding from the umbilicus. The skin usually shows bluish-black discoloration. Umbilical endometriosis is seen in about 15% of patients with intraperitoneal endometriosis.^{15 16}

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Cutaneous endometriosis develops into a malignant tumour only rarely (0.3-1% in surgical scars compared with ~2.5% in ovarian endometriosis).^{17 18}

2 Which differential diagnoses have to be considered?

Short answer

Primary or metastatic malignant umbilical tumour, lipoma, lipoleiomyoma, abscess, cyst, hernia, granuloma, keloid, or a congenital malformation of the omphalomesenteric duct or urachus.

Long answer

Umbilical endometriosis must be differentiated from several disorders. Primary malignant umbilical tumours (such as melanoma, basal cell carcinoma, squamous cell carcinoma, myosarcoma) are rare, representing only about 20% of all malignant umbilical tumours, with the remainder being metastatic.¹⁹ Most often, filiae of an adenocarcinoma are found, but metastases from sarcoma, mesothelioma, melanoma, and others have also been detected.²⁰

Non-malignant umbilical tumours include lipoma, lipoleiomyoma, abscess, cyst, hernia, granuloma, and keloid.^{21 22} Congenital malformations of the omphalomesenteric duct or urachus must also be considered.¹⁹

3 Which diagnostic procedure(s) would be appropriate?

Short answer

Inspection, palpation, ultrasound, computed tomography, magnetic resonance imaging, and biopsy with histopathological analysis.

Long answer

In addition to medical history, macroscopic inspection, and palpation, the following examinations are appropriate^{16 23}:

- Magnetic resonance imaging
- Computed tomography
- Ultrasound scans
- Histopathological analysis of lesional tissue (fig 2).



Fig 2 Histopathology of endometriosis (haematoxylin and eosin stain; magnification ×20). Courtesy of Doris Mayr, Institute of Pathology, Ludwig-Maximilians University, Munich, Germany)

A single dermoscopy study on cutaneous endometriosis mentions the pathognomonic sign of "red atolls," which was not present in our patient.²⁴

If pelvic manifestations of endometriosis are suspected, gynaecological examination, vaginal ultrasound, and laparoscopy may help to exclude the diagnosis or locate and remove tissue.^{1 16}

The value of the serum CA125 concentration in the diagnosis of endometriosis has been debated.²⁵

4 What kind of treatment would you recommend?

Short answer

Surgical excision and medical treatment.

Long answer

In umbilical endometriosis, a complete wide local excision is the treatment of choice.^{2 3 14} In severe cases or in the presence of pelvic endometriosis, hormonal treatment aims to reduce the symptoms of endometriosis by suppression of luteinising hormone and follicle stimulating hormone.³ In this context, gonadotrophin releasing hormone agonists and other agents (including oral contraceptive pills, progestins, and synthetic androgens) have been used to induce atrophy of endometrial tissue.^{1 14} ¹⁶ ^{26 27}

Patient outcome

Excision of the lesion with accompanying umbilical reconstruction proved successful. We did not start the patient on systemic treatment with gonadotrophin releasing hormone agonists or other agents for suppression of luteinising hormone and follicle stimulating hormone. Although local recurrence is uncommon, she was informed of the risk of endometriosis around the scar. Perioperative screening for depression was negative.^{28 29} She is being followed up at the gynaecology department.

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Patient consent obtained.

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