Case report
Spontaneous umbilical endometriosis:

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Case presentation

- **A 32-year-old married lady** visited our outpatient clinic after she presented **with umbilical swelling of 1 year duration**. Associated to this she has **a cyclical pain along with her menses and a bluish dark discoloration of the umbilicus** (Fig. 1).
- She had one pregnancy loss after 3 months of pregnancy.
- She has regular menstrual cycle which comes every month and stays 3–4 days.
- Otherwise, she has no previous abdominal surgery, dyspareunia, bowel habit change, or weight loss.
- For the above compliant she has been treated with antibiotics and analgesics with the consideration of umbilical abscess several times but doesn't show improvement.
- On examination she has a 1 by 1 cm sized tender, firm, dark brownish umbilical nodule.

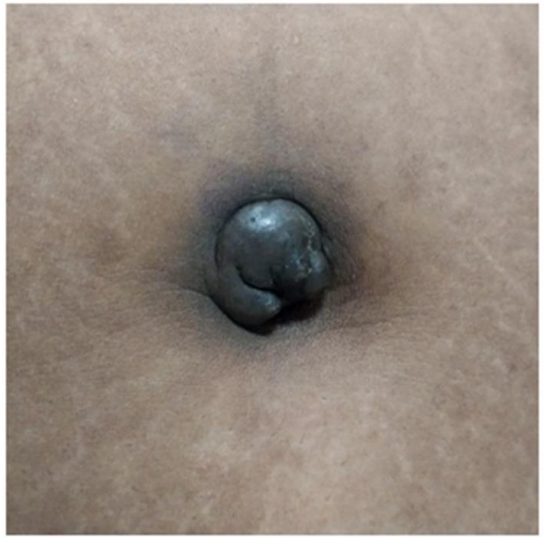
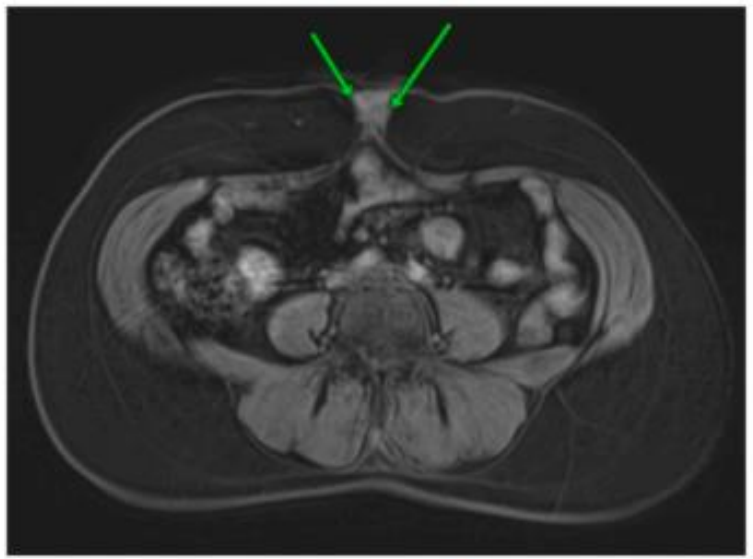
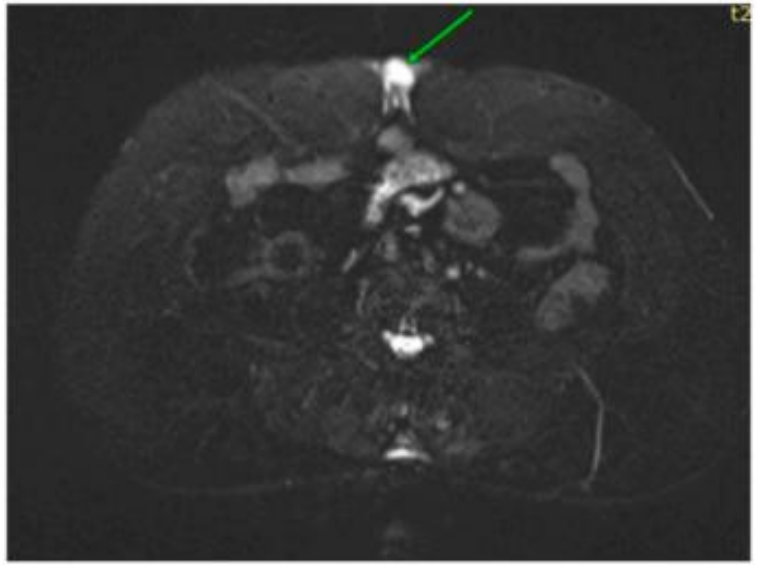


Fig. 1. A dark brownish umbilical nodule which measures 1 cm by 1 cm with a firm consistency.

- For the above compliant she was investigated with complete blood cell count (CBC), abdomino-pelvic ultrasound, Pelvic MRI and fine needle aspiration cytology (FNAC) was done.
- CBC showed normal cell lines. Abdominal ultrasound shows umbilical subcutaneous well defined hypoechoic lesion measuring 1.7 by 0.9 cm. Abdominopelvic MRI showed an umbilical subcutaneous well defined T1 hyperintense lesion measuring 2.3 by 2.1 cm (Fig. 2).
- FNAC smear show cellular aspirate of cohesive sheets of bland looking epithelial cells in a hemorrhagic background and hemosiderin laden macrophages seen.
- With all the clinical presentation of the patient and supportive diagnostic modalities, a diagnosis of primary umbilical endometriosis has been made.
- The patient started **on NSAIDs** but doesn't show improvement and **GNRH agonists** were not affordable.





Then umbilical excision with wide margin followed by reconstruction was done

Fig. 3. Excision of the umbilical nodule with negative margins.

- The excised sample was subjected for biopsy and microscopy showed keratinizing stratified squamous epithelium lined tissue with dermal based proliferation of endometrial type gland and stroma composed of tubular glands lined by Pseudostratified columnar cells along with areas of hemorrhage and hemosiderin confirming the diagnosis of umbilical endometriosis
- . No features of malignancy (Fig. 4).
- The patient was followed for six months postoperatively and has shown improvement from the symptoms

Discussion

- Umbilical endometriosis is the presence of endometrial glands and stroma in the umbilicus which is either localized to the umbilicus or extending to the peritoneum.
- It is also known as **Villar's nodule** named after the first physician describing the disease in 1886.
- It usually occurs either secondary to endometrial tissue implantation during **laparoscopic or open surgical** procedures also called scar endometriosis or as a **primary umbilical endometriosis (PUE**) which has low incidence compared to scar endometriosis .
- Our patient had history of neither laparoscopic nor open surgery previously. Up to 25 % of umbilical endometriosis occurs with concurrent pelvic endometriosis.
- Subfertility is a common condition among patients with endometriosis, occurring in up to 50 % of women with endometriosis .
- Patients with umbilical endometriosis might **be asymptomatic or typically present with a dark brown nodule in the umbilicus,** which may be swollen, painful, and sometimes bleed during menstrual periods .
- Our patient had been experiencing umbilical swelling for a year, accompanied by cyclical pain coinciding with her menstrual cycle, and her umbilicus had a bluish-dark coloured nodule (Fig. 1).
- This is a characteristic presentation for umbilical endometriosis. In addition to the typical clinical presentation, diagnosis of umbilical endometriosis is made based on imaging (ultrasound or MRI) and fine needle aspiration .
- Investigations employed to diagnose umbilical endometriosis include abdominal and/or transvaginal ultrasound and Magnetic resonance imaging (MRI).

- On imaging, we look for the umbilical connection with the peritoneum or fascia as well as the involvement of adjacent structures particularly the fascia which have a strong implication during resection and subsequent reconstruction.
- Imaging modalities can also help us to assess the presence of multifocal endometriosis.
- Our patient has only umbilical mass consistent with cutaneous endometriosis without adjacent structure involvement and no evidence of endometriosis on other abdominal organs.
- Histopathological tests like biopsy and fine-needle aspiration cytology are confirmatory for the diagnosis of umbilical endometriosis.
- The commonest differentials for umbilical nodule that mimic umbilical endometriosis are umbilical hernia, lipoma, pyogenic granuloma, amelanotic melanoma, Sister Mary Joseph metastatic nodule, primary umbilical malignancy, urachal residual or cyst, pemphigus vegetans, Desmoid tumour, and haemangioma .
- Our patient had a history **of peroral antibiotic prescription** by her primary physician because of the presence of tenderness and skin discoloration which mimics pyogenic granuloma.

- There are medical and Surgical management options for endometriosis.
- **The medical management of endometriosis** is targeted towards controlling pain and suppression of the hormonally active endometriotic tissue, while **the surgical management** deals with excision of the ectopic endometrial tissue.
- Options of medical management includes nonsteroidal anti-inflammatory **drugs(NSAIDs)** that controls pain during menstruation and hormonal therapies that rely on suppression of the endometriotic tissues **include combined oral contraceptives, progesterone only contraceptives, gonadotropin releasing hormone (GnRH) agonists, aromatase inhibitors and danazol .**
- A complete nodule excision procedure with free margin is recommended to avoid recurrence and a low risk of malignancy .
- Hormonal therapy with GnRH agonists and NSAIDs can be used preoperatively for relief of symptoms, but it is not curative.
- It can also be used to reduce the size of large lesions prior to surgery. However, it is associated with side effects such as amenorrhea .
- We did surgical excision of the umbilical nodule with free margin followed by umbilical reconstruction. The drawback of total umbilical excision is that it has cosmetically unacceptable scar.
- There is a modification to this technique where laparoscopically assisted excision of umbilical endometriosis is done with non-umbilical port of entry which have better aesthetic preservation of the umbilicus

Conclusion

- Umbilical endometriosisis a rare condition that should be considered as a differential diagnosis in women with umbilical lump, cyclical pain, and bleeding.
- The diagnosis is clinical and confirmed by histopathology.
- Imaging modalities are helpful to assess the extent of involvement of adjacent structures, rule out differential diagnosis, and evaluate for the presence of multifocal endometriosis.
- Surgical excision is the treatment of choice with a low risk of recurrence or malignancy.
- Hormone therapy and NSAIDs can be used preoperatively for relief of symptoms, but it is not curative.
- It can also be used to reduce the size of large lesions prior to surgery

- Endometriosis is an inflammatory disease associated with pelvic pain and infertility that is characterized by lesions of endometrial-like tissue outside of the uterus [1].
- It affects roughly 10 % (190 million) of reproductive age women and girls globally.
- In the field of endometriosis, more than 20 classifications, staging and descriptive systems have been developed, but none of the classification systems adequately classifies endometriosis [1,2].
- The well accepted classification system for endometriosis is the revised American Society for Reproductive Medicine (rASRM) classification (1997), which classifies endometriosis in the peritoneum, ovary and tubes [3].
- Endometriosis most commonly affects the pelvic organs, particularly the ovaries and fallopian tubes; however, extra-pelvic endometriosis affecting the bladder, kidney, bowel, omentum, lymph nodes, lungs, pleura, extremities, umbilicus, hernia sacs, abdominal wall, heart, and even the brain has been described.
- Extra- pelvic endometriosis is uncommon, accounting for just 12 % of documented instances.
- The gastrointestinal tract and the urinary system are the most common sites of the extra-pelvic endometriosis [4].
- Umbilical endometriosis is defined by the presence of endometrial glands and stroma outside the uterine cavity in the umbilicus [5].
- Umbilical endometriosis is a very rare finding, accounting for about 0.5–1 % of extra-pelvic endometriosis [6].
- It usually occurs either secondary to endometrial tissue implantation during laparoscopic or open surgical procedures also called scar endometriosis or as a primary umbilical endometriosis (PUE) [7].

- Primary umbilical endometriosis is a rare entity occurring spontaneously without any history of surgery.
- The exact pathogenesis in primary umbilical endometriosis remains unknown however retrograde menstrual flow, haematogenous and lymphatic transport of endometrial cells from the pelvis, and urachal remnant metaplasia are the proposed theories .
- Patients with umbilical endometriosis may not exhibit symptoms or present with various complaints.
- The commonest presenting symptoms are umbilical lump, cyclic pain, and bleeding from the umbilicus .
- Due to the varied presentations and rare incidence of endometriosis, it remains a diagnostic dilemma and challenge to treat it timely and properly.
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