What is your diagnosis?

A 38-year-old lady presented with a painful swelling in the umbilicus, together with a history of increased pain and bleeding from the swelling at the time of menstruation for the last seven months. Her menstrual cycles were regular, with average flow and no dysmenorrhea. She had two living children, both were delivered vaginally. There was no history of pelvic pain, infertility, treatment for infertility, pelvic/abdominal surgery, or caesarean section. Examination revealed a 1.0x0.5 cm firm, tender, reddish-blue colored nodular swelling in the abdominal wall, located just inferior to the umbilical ring with well-defined margins and a regular surface (Figure 1). Pelvic examination was essentially normal with a multiparous-sized uterus that was anteverted, mobile, and non-tender. Both fornices were free and non-tender. The rectovaginal septum was free and there were no nodules in the pouch of Douglas.

Ultrasound revealed a well-defined, hetero-echoic lesion with a peripheral rim of colour lying infra-umbilically, superficial to the rectus sheath. The same lesion appeared hyperintense on T1/T2 magnetic resonance imaging (MRI) with post-contrast enhancement. The abdomen and pelvis were found to be normal on MRI.

The patient was taken up for surgical excision of the nodule. Radical omphalectomy was performed. A peri-umbilical incision was made. Umbilicus, underlying nodule, and the surrounding area of fibrosis were dissected with a 5 mm clear margin using diathermy (Figure 2). The patient is on follow-up and is free of the disease at 18 months after surgery.

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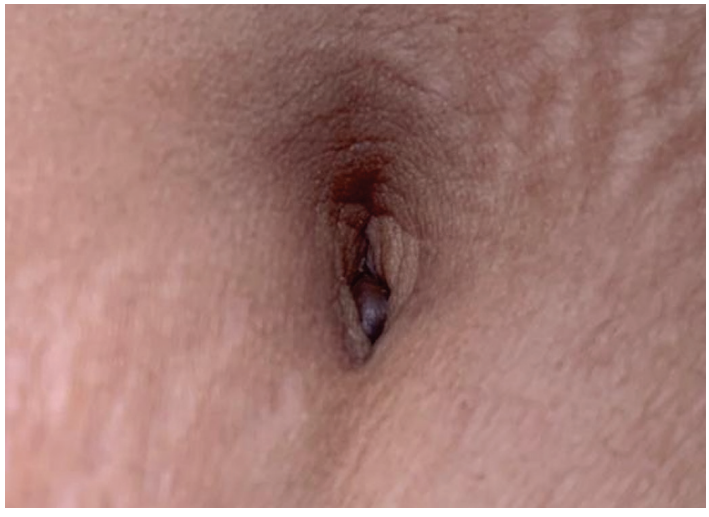

Figure 1. Clinical picture of umbilical endometriosis: bluish-purple firm umbilical swelling or nodule with associated cyclical pain

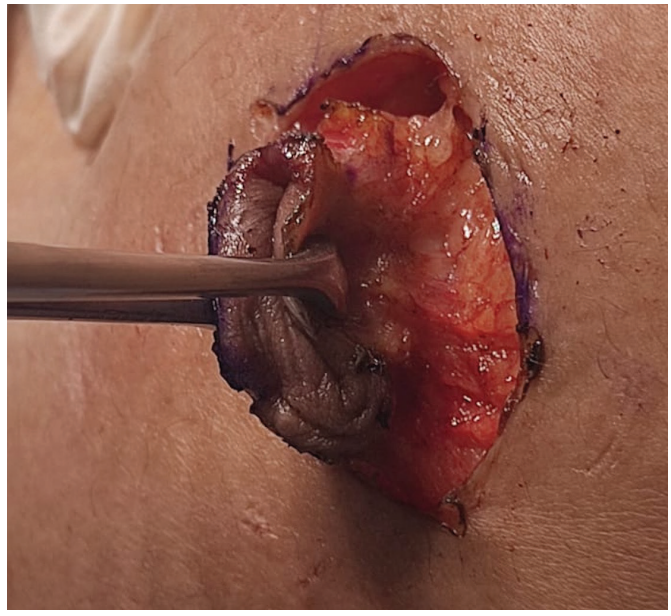

Figure 2. Radical omphalectomy

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Answer

Histopathological examination of the umbilical nodule revealed plenty of endometrial glands surrounded by compact stroma and intervening hemorrhage, confirming the diagnosis of umbilical endometriosis (PUE) (Figure 3). This was a rare case of Villar's nodule or PUE without concomitant pelvic endometriosis and in the absence of previous pelvic surgery.

PUE is extremely rare, making up 0.5-1% of all ectopic endometriosis cases (1). Extension of the endometrial cells to the umbilicus via the round ligament or the omphalo-mesenteric canal may explain the occurrence of PUE. Hematogenous or lymphatic transport of the endometrial cells is another possible mechanism that supports the existence of PUE (2).

PUE usually presents around 35-40 years of age. Clinical presentation includes a bluish-purple, firm, umbilical swelling or nodule with associated cyclical pain. Patients can even show catamenial bleeding from the umbilicus concomitantly with the menstrual cycle (3). Diagnosis is suspected when there is a typical history of an increase in pain/bleeding through the nodule at the time of menstruation. However, diagnosis can be confirmed only by histopathological examination (1,2).

Imaging modalities, such as ultrasound, MRI, or computed tomography scan are not superior in terms of sensitivity to the clinical scenario and examination findings (4). However, imaging can help assess the anatomical relationship of the nodule with the surrounding tissues and to rule out other differential diagnosis of umbilical lesions, like a desmoid tumor, lipoma, Sister Mary Joseph's nodule, teratoma, trichobezoars, umbilical concretions, and hernia, for example (5). Imaging can also help to investigate the anatomical relationships of the nodule with the surrounding tissues (5).

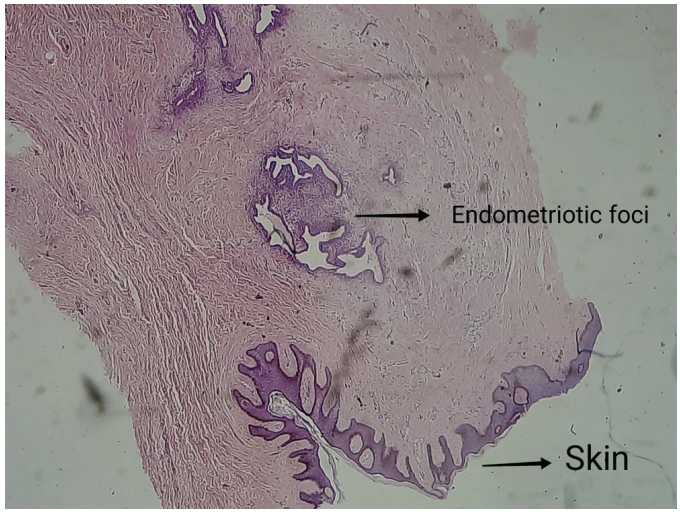

Figure 3. Sections from umbilical mass showing stratification squamous epithelium with the subepithelium showing endometrial glands and stroma

Surgery remains the mainstay of treatment. Medical management using progestins, danazol, and/or gonadotrophin releasing hormone agonists may be tried, but recurrence rates are high (5). Radical omphalectomy is the most frequently performed surgery for umbilical endometriosis. This involves the removal of the umbilicus with the nodule along with plastic reconstruction. Partial omphalectomy is local resection of the endometrial nodule with umbilical sparing. It is important to ensure disease-free margins of at least 3 mm to prevent local recurrences. Sometimes a deep-seated nodule may also necessitate removal of the underlying rectus sheath, which may require anatomical repair or mesh placement.

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