

RESEARCH ARTICLE

UMBILICAL ENDOMETRIOSIS: A CASE REPORT

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Abstract

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Manuscript Info

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Introduction: We report a recently observed case of secondary umbilical endometriosis (UE), with the main aim to discuss diagnosis and the management of this rare condition.

Observation: A 41-year-old woman admitted to our facility with a nodular, bluish umbilical swelling, non- painful, gradually increasing in size, bleeding with her menstrual cycle. Ultrasonography did not reveal any images indicative of pelvic endometriotic lesions. A pelvic MRI was performed wich showed an ovoid formation at the umbilical level, suggestive of a UE implant. A biopsy of the lesion was also carried out, showing a morphological appearance compatible with parietal endometriosis. Histopathological analysis confirmed the diagnosis of UE. Neither symptoms or signs of local recurrence have been observed after 6 months.

Discussion: Extra-pelvic endometriosis sites are not common, especially the umbilicus. It usually occurs secondary to surgical scars, specifically after laparoscopy or open abdominal surgery. UE should be taken into accounting differential diagnosis of umbilical disorders even in young nulliparous women with no typical symptoms of pelvic endometriosis. UE can be treated medically or surgically. Surgical treatment options are either local excision of the lesion or removal of the whole umbilicus with or without laparoscopic exploration of the peritoneal cavity. The decision should be tailored for the individual patient, taking into consideration the size of the lesion, the duration of symptoms and the presence of possible pelvic endometriosis.

Conclusion: Local excision saving the umbilicus may be the treatment of choice in patients with small UE lesions.

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Introduction:-

Endometriosis is a pathology characterized by the presence, in an ectopic location, of endometrial structures exhibiting histological and biological characteristics of the endometrium but remaining anatomically separate [1]. Overall, the prevalence of the disease in the general population is estimated to be 5 to 10% [2]. Cutaneous involvement represents only 0.5 to 3.5%, primarily occurring in laparotomy scars, especially after hysterectomy [2]. However, isolated occurrences at the umbilicus have been reported but are exceptionally rare [2]. In the majority of cases (60%), umbilical endometriosis has been described in women who underwent laparoscopic surgery. It primarily affects women in their reproductive years [3]. We report a case of a patient admitted for the management of an umbilical nodule, with histological examination of the biopsy suggesting endometriosis.

Observation:-

We report the case of a 41-year-old married woman, in her fourth pregnancy with four deliveries, including three live births through vaginal delivery and the last one via cesarean section five years ago. She is being treated for type 2 diabetes with oral antidiabetic agents and hypertension under dietary control. The patient was admitted to our facility with a nodular, bluish umbilical swelling, non-painful, gradually increasing in size, reaching a diameter of 2 cm. It is mobile relative to the deep plane and is accompanied by bleeding at the umbilicus during menstruation. This condition has been evolving over the past four years.



Fig 1:- A 2 cm umbilical tumor.

The clinical examination reveals a patient in good general condition, stable on the hemodynamic and respiratory fronts, with a soft abdomen, a clean scar, and slight inflammation around the umbilicus. The breast examination was normal, with a normal-looking cervix and no visible lesions or bleeding. The diagnosis of endometriosis was suspected based on the bluish appearance and the cyclical nature of pain and bleeding.

A pelvic and endovaginally ultrasound did not reveal any images indicative of pelvic endometriotic lesions. The patient also underwent a pelvic MRI, which identified an ovoid formation at the umbilical level measuring 11 x 25 x 19 mm, suggestive of an umbilical endometriotic implant. Biopsy of the lesion exhibited a morphological appearance consistent with parietal endometriosis without histological signs of malignancy.



Fig 2:- Cytogenic stroma surrounding simple glandular structures lined with cub cylindrical epithelium from the endometrium, indicative of umbilical endometriosis.

The surgical intervention involved a wide excision, encompassing both the tumor and the umbilicus. Histological examination revealed a morphological and immunohistochemical profile consistent with subcutaneous umbilical endometriosis. As of six months postoperatively, no recurrence has been observed.



Fig 3:- The dermis and subcutaneous tissue exhibit multiple cystic glands lined by a regular, simple cub cylindrical epithelium, indicative of subcutaneous umbilical endometriosis.



Fig 4:- Endometriosis at a magnification of \times 100: Presence of dilated glandular structures forming cysts filled with secretions. Hematoxylin and eosin staining.

Discussion:-

Umbilical endometriosis, also known as Villar's nodule, is a rare condition, with an incidence of 0.5 to 1% among all extra-genital endometrioses. It is characterized by the presence of functional endometrial tissue in the umbilicus [4]. It is most commonly observed in women of reproductive age, with a peak incidence between 30 and 40 years, as seen in our patient; it is exceptionally rare before the age of 20 [5]. The typical clinical presentation involves sharp pain localized at the umbilicus, resembling a superficial burning sensation, which was not the case in our observation.

We distinguish between primary endometriosis, which occurs in women without a history of abdominal or pelvic surgery, and secondary endometriosis that develops on a scar following a gynecological or obstetric intervention or laparoscopy [6], as is the case with our patient.

In secondary forms, there is believed to be a graft of endometrial tissue at the surgical wound during the surgical intervention [6,7], which may manifest after a period ranging from 1 month to 13 years [6, 8].

Clinically, umbilical endometriosis presents as a firm swelling, sensitive to palpation, with variable volume depending on the menstrual cycle or may simply exude hemocytic serositis [9]. Perilesional sclerosis can lead to an overestimation of its size [10]. Bloody discharge synchronized with menstruation appears to be less frequent. The cyclical nature of this symptomatology, coinciding with menstruation, is crucial and sometimes sufficient for considering the diagnosis during the clinical examination [11]. However, this cyclical evolution is observed in only about half of the cases and is rarely complete [11]. In our case, the nodular swelling was painless with bloody discharge, all evolving in a cyclical pattern.

The main differential diagnoses include umbilical tumors such as hernias, granulomas, hematomas, neurinomas, epidermoid cysts, and more rarely malignant tumors (sarcoma, Sister Mary Joseph nodule revealing ovarian adenocarcinoma) [12].

The ultrasound appearance of umbilical endometriosis varies. It is most often a well-defined, tissue-like, hypoechoic mass. However, the lesion can be cystic, mixed, or solid. Color Doppler ultrasound shows a frequently highly vascularized mass with dilated afferent vessels [2]. MRI is the preferred diagnostic tool to confirm the diagnosis in case of uncertainty, being more sensitive for detecting small lesions [2].

The advantages of MRI to ultrasound makes it second line/problem solving modality. Tissue characterization of MRI adds specificity by identifying hemorrhage associated with endometriotic lesions. Superb soft tissue contrast resolution aids in better delineation between muscles and abdominal subcutaneous tissues and infiltration of abdominal wall structures [13].

Imaging can also guide in differentiating other causes of umbilical swelling such as hemangioma, granuloma, melanocytic nevus, seborrheic keratosis, granular cell tumor, umbilical hernia and umbilical polyps [14].

Umbilical endometriosis can only be diagnosed through the pathological examination of the lesion. Macroscopically, it appears as a poorly defined, bluish, nodular, or cystic tumor that, when cut, releases a chocolate-colored fluid or even blood. Sometimes it presents as a small tumor surrounded by intense sclerosis without a clear cleavage plane, and the multiplicity of tumor foci is a strong argument in favor of endometriosis [10]. In our case, the macroscopic appearance was that of a nodular tumor without the presence of chocolate-colored fluid upon cutting. Microscopically, the appearance is similar to that of ectopic endometrium, particularly the coexistence of endometrial glands lined with cylindrical epithelium and endometrial stroma composed of small round cells and an extensive vascular network (Fig. 2). From a functional perspective, ectopic endometrium is hormonally dependent, as evidenced by the cyclical nature of symptoms based on the menstrual cycle, changes throughout reproductive life, worsening under estrogenic treatment, and decidualization of lesions during pregnancy. However, these cyclical modifications are common, minor, and partial [10, 15, 16].

The consensus about standard management of primary umbilical endometriosis is not developed due to the rarity of the disease [14, 17].

Medical management widely used hormonal therapy like oral contraceptive pills, dienogest and GnRH antagonist have been mentioned in literature and have been effective in relieving the symptoms but disease progression is not halted by medical therapy so recurrence of symptoms and growth of lesions is the major drawback [18].

Surgery remains the only curative treatment through extensive surgical excision, removing the entire lesion and extending at least 5 mm into healthy tissue to prevent recurrence. A parietal prosthesis can be used to close the aponeurotic defect [12, 19]. In our case, the patient underwent complete excision of the nodule with margins closest to 1mm. Histological examination confirmed endometriosis. Other therapies based on selective estrogen and progesterone receptor modulators are currently under evaluation and appear promising [20].

Conclusion:-

Umbilical endometriosis is a rare condition, and its physio pathogenic explanation remains controversial. Neither clinical presentation nor imaging through ultrasound or MRI is specific to this pathology. Its diagnosis should be considered in the presence of a painful, well-defined, solid or cystic nodule in the umbilical fossa, accompanied by brownish discharge, in a woman with or without a history of abdominal surgery. Surgical excision is the preferred treatment to prevent rare recurrences. A definitive diagnosis is only obtained through histological examination.

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