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Umbilical Endometriosis Coexisting with Umbilical Hernia

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ABSTRACT

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Endometriosis, though most often pelvic in site can have multiple presentations including the rare umbilical endometriosis, which accounts for 0.5-1% of all extragenital locations of endometriotic lesions. It can be primary or secondary in origin and can have varied clinical features ranging from a painless, discoloured nodule to cyclical pain and bleeding coinciding with menstruation. Though different radiological investigations have been suggested, histopathological examination after surgical excision of the lesion is the best method to confirm the diagnosis.

Keywords: Umbilical Endometriosis, Umbilical Hernia, Extragenital

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INTRODUCTION

Endometriosis is the presence of functional endometrial tissue outside the uterine cavity. Though benign, it can invade the normal surrounding tissue. Most common sites are ovary, uterosacral ligaments, broad ligaments, fallopian tubes, uterovesical fold, round ligaments, rectovaginal septum. Less commonly, it can also be found in sigmoid colon, appendix, ureter, urinary bladder, caecum, ileum, lungs, nasal mucosa, eyes, brain, umbilicus. Umbilical endometriosis or Villar's nodule [named after Villar, the first to report the condition, in 1886] can be a rare occurrence but is one of the most common extrapelvic sites. It can be primary, if appearing spontaneously or secondary if it occurs after a surgical procedure involving the umbilicus [like a laparoscopic port insertion], on the scar. The term secondary endometriosis can be used even when it is not located on the exact surgical scars, [such as endometriotic implant on the umbilicus in a patient with the history of a previous caesarean section], but only if the onset is within 2 years after the procedure. It can present with a painless nodule or one that undergoes periodic changes like enlargement / bleeding during menstrual cycles. It could be brown, reddish or violaceous in colour. 75% patients may have temporal changes associated with menstruation, helping the physician rule this rare possibility in.

CASE REPORT

49 year old P3L3 woman came with painful umbilical swelling since 1 year and intermittent dark reddish discharge since 3 months coinciding with her menstrual cycles, usually from the third day. The swelling had been constant in size and appearance since she first noticed it with no enlargement or notable tenderness during days of menstrual flow. She had no constitutional symptoms, dysuria, dyschezia, dyspareunia, pelvic pain, severe dysmenorrhea, haematuria or altered bowel habits. Her menstrual cycles are regular with scantier flow than in the past since the last few months. She has had no history of subfertility. She has had a caesarean section 11 years ago. She is not on any contraceptives. She has no known medical comorbidities and has not been subjected to any other surgeries. She reports no history of trauma. Review of systems was normal.

On physical examination, a nontender, hyperpigmented, irreducible swelling of ~ 2x2 cm size in the periumbilical region was found. No discharge or local rise in temperature was noted and cough impulse was negative. Abdomen was soft, non tender and no masses/organomegaly was detected. Other systems were normal.

Differential diagnoses included umbilical endometriosis, incarcerated umbilical hernia, umbilical granuloma, keloid, melanocytic naevus, malignant lesions like adenocarcinoma, malignant melanoma, metastatic Sister Mary Joseph nodule etc. Umbilical endometriosis had a clear edge over the others due to the temporal association of the discharge with her menstrual cycles **(Figure 1)**.

The patient was scheduled for an excision under local anaesthesia. Preoperative haemogram, biochemical and coagulation profile were normal. Patient was put on

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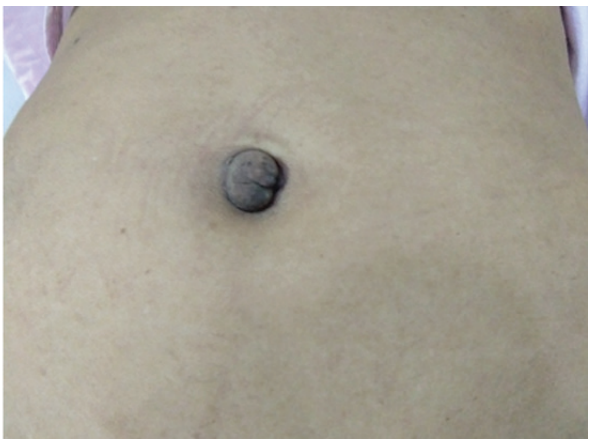


Figure 1. **Umbilical endometriosis**

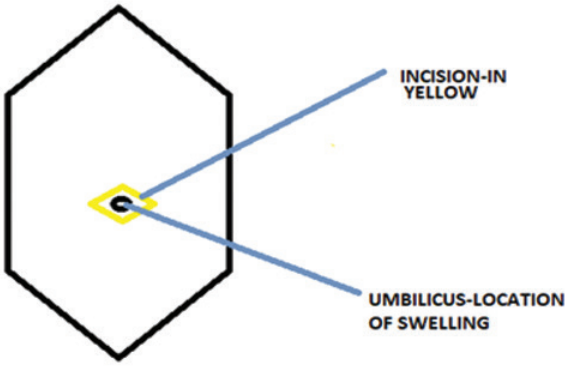


Figure 2. **Elliptical incision around the umbilicus**

Midazolam 2 mg iv, Inj. Fortwin 30 mg and Inj Phenergan 25 mg. Umbilical swelling and the region around it were painted with povidone-iodine and draped under sterile conditions. Local anaesthesia was given with 2% lignocaine + adrenaline. Elliptical incision was put around the umbilicus and the swelling was excised with a safe margin **(Figure 2)**. A hernia sac with bowel content found was dissected and the defect was repaired with prolene mesh. Wound was closed in layers, skin was sutured with absorbable sutures and umbilicus was reconstructed. Haemostasis was achieved. Excised segment was sent for biopsy.

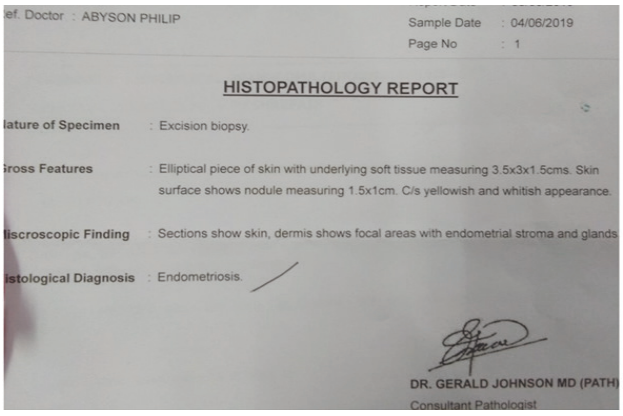
Histopathological examination of the excised region showed skin and dermis with focal areas of endometriotic stroma and glands **(Figure 4)**, which was compatible with the clinical diagnosis of umbilical endometriosis **(Figure 3)**.

Patient returned a week after the surgery for follow-up. The wound was healing well with no post-operative complications.

DISCUSSION

Endometriosis is an estrogen-dependent condition. Our patient has had a past caesarean section and literature shows nearly 1% of such women can have secondary endometriosis in the umbilicus. It is not necessary that the surgery should directly involve the umbilical region. But another school of thought exists that the remoteness of the endometriotic lesion [in the umbilicus] from the operative scar and the fact that the onset of her condition occurred more than 2 years after the surgery [11 years in this case] qualify this for being treated as a case of primary umbilical endometriosis. Though there is no formal criterion to decide the category, review of existing literature supports the latter view. Umbilicus is the commonest extragenital site of endometriosis. It is postulated that it acts as a physiologic scar exhibiting a predilection for ectopic endometrial implants which may have been activated by an extended exposure to hormonal, metaplastic and environmental factors, which might explain why a lesion might develop de novo in primary umbilical endometriosis.

Many theories have been propounded to explain its etiopathogenesis. Secondary umbilical endometriosis, with a surgical history can be explained to an extent by iatrogenic dissemination of cells, the *direct transplantation theory*. Primary umbilical endometriosis is thought to be caused by lymphatic or haematogenous spread of endometrial tissue from the pelvic cavity. Lymphatic vessels connect the peritoneal cavity to the umbilicus along the obliterated umbilical vessels. Scott proposed migration of pelvic contents to the umbilicus through this channel. Other studies have reported the presence of endometrial tissue in the periumbilical lymphatic tissue. Alternatively, endometriosis in this particular site may also develop through metaplasia of the embryonic remnants in the umbilical fold such as urachus or the umbilical vessels. Genetic predisposition and alterations in cell-mediated and humoral immunity may also favour spontaneous development of this condition. Our patient is a 49 year old woman, in her perimenopausal phase. Theoretically, with dwindling estrogen levels ascribed to this phase, an estrogen dependent condition like endometriosis



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HISTOPATHOLOGY REPORT

lature of Specimen : Excision biopsy.

Gross Features : Elliptical piece of skin with underlying soft tissue measuring 3.5x3x1.5cms. Skin surface shows nodule measuring 1.5x1cm. C/s yellowish and whitish appearance.

liscroscopic Finding : Sections show skin, dermis shows focal areas with endometrial stroma and glands.

istological Diagnosis : Endometriosis.

DR. GERALD JOHNSON MD (PATH)
Consultant Pathologist

Figure 3. **Histopathology report of Sample**

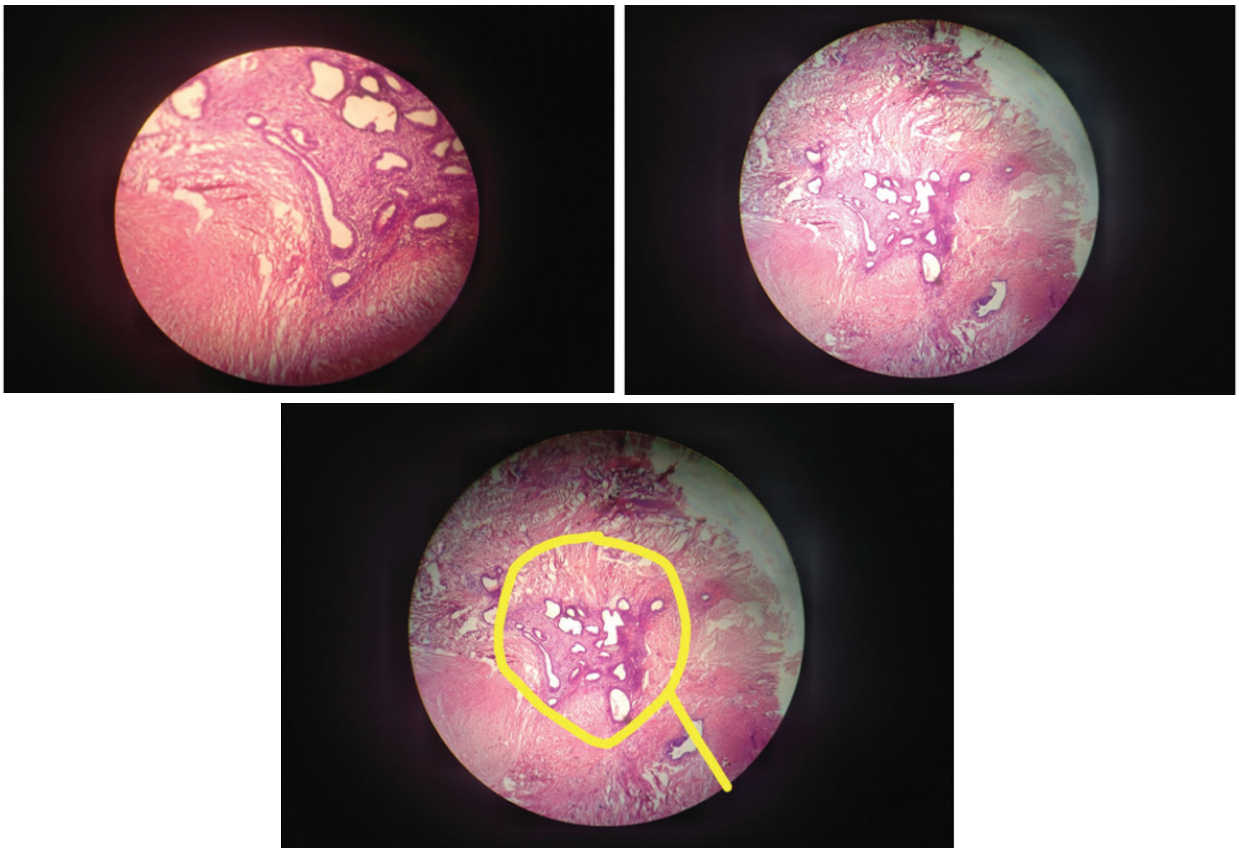

Figure 4. (a,b,c). **Circled in Yellow – Endometriotic glands and stroma [all images 20x]**

should cease to be a problem. But several studies have reported that early perimenopause could be a period of relative hyperestrogenemia contributed to by the luteal phase insufficiency. Quoting a study on perimenopause published in 2007, "*In many women, the circulating levels of estrogen in the perimenopause may be higher than at any other time of her reproductive life when she was not pregnant*". Ectopic endometrial cells, being estrogen-dependent could have been activated as a result of this favorable hormonal milieu. Besides, her relatively higher age [Mean age at presentation of umbilical endometriosis -37 years] might have been marked by prolonged exposure to metaplastic and environmental factors causing the endometriotic implants to be more functional at this stage.

Different case reports in the past have documented different approaches to manage this condition. Radiological investigations like ultrasonogram, computed tomography, MRI have all been attempted to study the lesion and is quite helpful in identifying various features like consistency, site, extent, adhesion to underlying structures. But none of these can diagnose a lesion as cutaneous endometriosis with clear pathognomonic features and are hence eclipsed in their clinical value by histopathological examination after surgical excision. Occasionally, observing the nodule during different phases of the menstrual cycle to note its changes and prescribing hormonal contraceptives, progestins, GnRH analogues to reduce the swelling, pain or bleeding have been attempted but results were inconsistent. Medical treatment is not a reliable curative method apart from offering symptomatic relief in some cases, especially prior to the surgery. Besides, though rare, malignant transformation of the umbilical endometriosis is a possibility and it can be confirmed only by excision of the lesion with a margin and its subsequent histopathological examination. Hence total umbilical excision and reconstruction of umbilicus or local excision of the nodule preserving the umbilicus is the safest and most definitive option to manage an umbilical nodule.

Gynaecological referral and evaluation for pelvic endometriosis is another cause for concern in such cases. Literature shows that it would be wise to base that decision on the individual merits of a case. A perimenopausal woman such as our patient with absolutely no clinical symptoms suggestive of pelvic endometriosis may not need an extensive evaluation. This approach is evidence-based as nearly 73% of women with umbilical endometriosis have no pelvic endometriotic implants at all. Hence, in our case, an excision without any prior imaging proved to be a very fast, effective and economical management strategy to reduce the patient's suffering. Even in cases where there may not be symptoms associated with menstruation, like just a painful or painless nodule, excision and biopsy would give definitive answers, though radiological clues can be complementary.

CONCLUSION

Umbilical endometriosis must be considered as a differential diagnosis in cases of an umbilical nodule. There have been cases where discharges from such nodules were attributed to an infective pathology and treated with antibiotics without improvement. It may not always present with the telltale signs of changes associated with menstruation or a past surgical history. Excision and subsequent histopathological assessment is the gold standard to confirm the diagnosis and effectively manage the condition.

END NOTE

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