

Case Report



Primary Umbilical Endometriosis (Villar's Nodule): A Rare Symptomatic Umbilical Pathology in An Adult Woman

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Abstract

A 35-year old married non-diabetic, non-hypertensive, non-alcoholic, non-smoking and non-betel-nut-chewing poor housewife having average body build and body weight hailing from Shahjadpur of Sirajgonj district got herself admitted with intermittent pain and 2.5x1.75 cm sized discolored swelling in the umbilical region for one year. The dull aching non-radiating pain aggravated by moving was not associated with fever, chills and rigor. The pain was distinctly remarkable during regular normal menstrual flow. It was responding variably to traditional analgesic and nonspecific antimicrobial drugs as advised by local village doctors. But she was not cured. Her bowel and bladder habits were normal. She had no abdominal pain, distension, chest pain, bone pain, dyspnea, cough, jaundice, umbilical discharge of faeces, blood or urine. Her two children were born normally (i.e., by NVD: Normal Vaginal Delivery). She had no previous history of any form of surgery. Her rt-PCR for Covid-19 test was negative with normal liver and kidney function tests and normal sonographic findings of the whole abdomen. Our clinical diagnosis of primary umbilical endometriosis was supported by FNAC from discolored umbilical swelling. After having an informed written consent we excised the umbilicus. The histopathological report had confirmed our preoperative diagnosis. The post-operative period was smooth and uneventful. We like to share our experience of diagnosing and treating this very rare disease of primary umbilical endometriosis.

Key words: Umbilical cyclic pain, Discoloration, Haemorrhage, Swelling.

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Introduction

The 'umbilicus' (also known as navel or belly button) in adulthood is composed of scar tissue located at the midpoint of anterior abdominal wall. It is covered by infolded and puckered skin having crevices inside. It marks the area of passage of two umbilical arteries, one umbilical vein, the allantois and the omphalomesenteric (vitelline) duct within the wharton's jelly during intrauterine life.¹⁻³ In adults, it may appear as a protruded or hollowed or flat area of variable dimension in the abdominal wall denoting the position of the attachment of the fetal umbilical cord.⁴ Various pathological conditions of adult umbilicus are well known. They include umbilical hernia, umbilical sinuses, umbilical fistulas (through which urinary or fecal or menstrual

discharge can flow), umbilical endometriosis, umbilical sepsis, urachal cysts or sinuses, vitelline cysts or sinuses, hair tufts with or without infections, pilonidal sinuses, foreign bodies (FBs), Omphalitis (inflammation of the umbilicus), umbilical infections and its such complications as sepsis, cellulitis, portal pyemia, PVT (Portal Vein Thrombosis), hepatitis, liver abscess, septicemia, septic shock, MODS (MultiOrgan Dysfunction Syndrome), MSOF (MultiSystem Organ Failure) etc., Omphalophobia (psychiatric) etc.⁷⁻⁹ Persistent urachus (allantois) or persistent vitelline duct may cause umbilical troubles at any age.³⁻⁵ Several other benign and malignant diseases of the umbilicus include keloids, dermatofibroma, cholesteatoma, melanoma, primary umbilical cancers and umbilical secondaries (the

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well-known Sister Mary Joseph metastatic nodule).^{6,10,11} umbolith or Omphalolith (also known omphalokeratolith, omphalith, inspissated umbilical bolus or simply umbilical bolus) is also seen as an uncommon disease.^{2,12,13} Rarely Umbilical endometriosis, also known as Villar's nodule, is found in women which may be primary or secondary characterized by presence of ectopic endometrial tissue in the umbilicus, that can present as a painful, discolored mass or swelling in the umbilicus or as umbilical discharge during menstrual period. Umbilical endometriosis though rare, is the commonest form of cutaneous endometriosis.^{10,14,15}

It is categorized as primary umbilical endometriosis (75% of all umbilical endometriosis) when it occurs spontaneously (without a known aetio-pathogenesis, i.e. no cause is discernible) and secondary umbilical endometriosis when it occurs following a surgical incision from a previous open gynaecological or abdominal surgery, such as caesarean section, hysterectomy, or laparoscopic surgery, the latter being more common (thought to occur following iatrogenic seeding of umbilical tissue). Primary umbilical endometriosis was first described by Mr. Villar in 1886, that is why it is also known as Villar's nodule.^{16,17,18}

Endometriosis is the presence of endometrial tissue outside the uterine cavity. These lesions are typically found in the pelvic cavity but can occur in other extrapelvic areas including skin. Umbilical endometriosis (also known as Villar's nodule) is a rare disease comprising 0.5–1% of all extrapelvic endometriosis, thereby causing a dilemma in diagnosis and evaluation of umbilical hernias. It commonly presents with cyclical pain and bleeding from an umbilical (endometriotic) nodule.^{1,10,18}

Case Report

Mrs Kajulin Khatun, age 35 years, a married non-diabetic, non-hypertensive, non-alcoholic, non-smoking and non-betel-nut-chewing housewife of poor class with average body build and body weight hailing from Shahjadpur of Sirajgonj district got herself admitted to the female surgery ward with intermittent pain and 2.5x1.75 cm sized dusky colored swelling in the umbilical region for one year. The pain initially was mild, dull aching in nature, non-radiating, aggravated by moving and not associated with fever, chills and rigor. The pain was distinctly remarkable mostly during menstruation. Gradually it was getting worse day by day, with increasing intensity, responding variably to traditional OTC (Over-the-counter) analgesic and antimicrobial drugs that were prescribed by village doctors. She failed to specify the name of the drugs she used for her illness. The umbilical pain was accompanied with gradual and fluctuating umbilical swelling and the covering skin was getting discolored. Her bowel and bladder habits were normal with no complaints related to the alimentary, the hepato-biliary, the urinary, the respiratory and the nervous systems. Her menstrual cycles were regular with normal menstrual flow. She had no abdominal pain, no abdominal distension, no chest pain, no bone pain, no dyspnea, no cough, no yellow coloration of eye and skin, no umbilical discharge of faeces, blood or urine. She has a son and a daughter. The age of her last child was five years. Both of her children were born normally (i.e., by NVD: Normal Vaginal Delivery) without any

instrumentation. She had not been operated previously for any disease. None of her family members suffered from such sort of illness. During her childhood, she was immunized as per the then EPI (Expanded Program of Immunization) schedule against the then prevailing common infectious diseases. She was not known to be allergic to any diet, drug, pollen or allergen. She was psychiatrically and psychologically healthy. He gave no other notable past event of any disease.

The swollen umbilicus was found dusky colored (not red of acute inflammation), soft and tender. Local temperature was not notably elevated. The swollen parts seemed to be non-neoplastic, irreducible, non-pulsatile, non-compressible and not trans-illuminating. There was no visible and palpable impulse on coughing. (Fig. 1).



Fig. 1: Showing the swollen discolored endometriotic umbilicus.

She was mildly anaemic (which was not abnormal for an average Bangladeshi woman of poor class) with no jaundice, no cyanosis, no clubbing, no koilonychia, no leuconychia, no dehydration, no oedema, no ascites. Her hernial orifices were intact. Her neck veins were not engorged. No abnormality was detected in her heart, lungs, liver, spleen, kidneys, other non-umbilical skin, superficial veins, thyroid gland, breasts and accessible lymph nodes. Her pulse was 78/min, BP 130/80 mm Hg, respiratory rate 14/min, temperature 98 degree Fahrenheit. Her abdomen was scaphoid, moving up and down characterizing normal thoraco-abdominal respiration, with centrally placed, everted, tender, soft, swollen umbilicus. Her abdomen had no scar mark, no scratch mark, no muscle guard, no hyper

aesthesia. She had no palpable abdominal lump. Percussion note was tympanic with normal bowel sounds. No abnormality was detected in her all other systems.

Her total leucocyte count was 5,000/microL with polymorphs 70%, Hb 10.4 gm/ dl, ESR 14 mm in the 1st hour, HbA1C 5.5, RBG (Random Blood Glucose) 5.5 mmol/L, Serum creatinine 63 micromol/L, ECG normal. She had rt-PCR for Covid-19 test negative, normal chest x-ray, Serum bilirubin 8.88 micromol/L with other normal liver function tests and normal sonographic findings of the whole abdomen. FNAC from discolored umbilical swelling revealed it as a case of umbilical endometriosis.

After having a informed written consent and a shared decision with the patient and her husband, with all aseptic precautions, under general anaesthesia, through an elliptical incision centering the umbilicus, umbilectomy was done, (Figure 2). As peritoneal cavity was opened, the defect was repaired by double breasting technique as if it were an umbilical hernia. Further the abdominal wall was strengthened by fixing an onlay prolene mesh. Keeping a drain inside over the mesh, skin incision wound was closed with cosmetic stitches using prolene. The excised umbilical specimen was sent for histopathological examination where the report had confirmed our preoperative diagnosis of umbilical endometriosis, (Figure 3). Histopathological examination identified endometrial glands and endometrial stroma, focal areas of hemorrhage, chronic inflammation with chronic inflammatory cells plus hemosiderin laden macrophages.



Fig. 2: Showing umbilicus is being excised through an elliptical incision.

Umb endometriosis

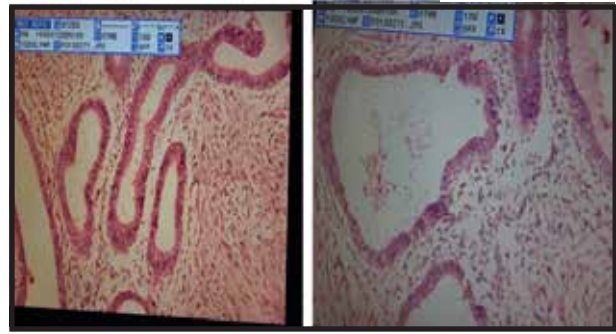


Fig. 3: Histopathology showing endometrial tissue in excised umbilicus.

Discussion

Endometriosis classically occurs women of reproductive child bearing age. It occurs mostly in the lower abdomen on pelvic organs mostly implicating the ovaries, the utero-sacral ligaments, the ovarian fossa, the pelvic cul-de-sac and bladder in that sequence.³ Extrapelvic endometriosis is less common. The extrapelvic sites may be the diaphragm, the respiratory system, the urinary tract, the alimentary tract, the hepatobiliary system, the brain and the skin. Umbilical endometriosis is the commonest type of cutaneous endometriosis.⁴ Theories on umbilical endometriosis: the exact cause of umbilical endometriosis is unknown. It is assumed that the normal physiological scar umbilicus is a preferred site for endometriosis.^{1,12} Lymphatic and hematogenous access to the umbilicus and direct spread of endometrial cells through the uterine round ligaments or the omphalo-mesenteric remnants are possible theories to explain the aetiolo-pathology of umbilical endometriosis.^{7,10,18} Discrete umbilical endometriosis may develop from metaplasia of tissue of urachal remnants.^{3,17}

D Makena, T Obura and S Mutiso et al in 2020 described five patients aging between 31 and 47 years with umbilical endometriosis, who had presented with umbilical pain and 1.6 cm to 4 cm sized swelling for 3 and 60 months.¹⁰ One had umbilical discharge and not hemorrhage These were diagnosed clinically and confirmed by histopathology (identifying endometrial glands and/or stroma and recent hemorrhages) that excluded cancerous lesion also. Two patients were nulliparous, and the rest three had previous one to six deliveries None was having miscarriage. One had two caesarean deliveries, whereas the others had (NVDs) Normal Vaginal Deliveries without instrumentation. None had reported any other form of previous abdominal surgeries. The presentations had been almost identical, having an umbilical swelling and cyclical pain, with or without discharge or bleeding. Three patients had such other symptoms as severe dysmenorrhea, with one having heavy bleeding. On preoperative MRI (Magnetic Resonance Imaging), one had shown a 1.6 cm umbilical pathology indicative of endometriosis and normal abdominal findings. On preoperative USG (UltraSonoGraphy), one patient had shown to have many intramural uterine fibroids inside uterine and a right ovarian simple cyst. . All other patients had not been submitted for any imaging study. Laparoscopy and hysteroscopy were done in one patient for dysmenorrhea, and the findings

were quite normal. Laparoscopy was done in one patient who had presented with subfertility, dysmenorrhea, and heavy cyclical bleeding, where endometriosis was detected on the uterosacral ligaments, the right ovarian fossa and the posterior uterine wall, that were treated at the same time. Laparoscopic hysterectomy was also performed. Laparoscopy was also offered to another patient where superficial endometriotic deposits on the anterior and the posterior cul-de-sac ablated in accordance to the informed written consent taken at admission for total management. All patients had uneventful postoperative recovery and no recurrence on follow-up.

Our this patient of case report presented at the age of 35 years which is consistent with all reported literatures, that indicates that endometriosis is an active hormone-responsive benign disease implicative women of reproductive age prior to menopause. The duration of the symptoms in our patient was only one year, which is consistent with any reported literature. Umbilical endometriosis may be primary if it occurs spontaneously without any previous known aetiopathogenesis or secondary that follows past surgical events like laparotomy, laparoscopy, caesarean or other instrumental deliveries etc., especially laparoscopic one with periumbilical or umbilical port entries. Our case had no previous surgical or instrumental event. Thus it was a case of primary umbilical endometriosis, as described in literatures. We diagnosed it mainly on clinical impression, which was consistent with any reported literatures. Our FNAC findings were consistent with our clinical impression that was subsequently confirmed by histopathological examination. We had done USG of whole abdomen including the pelvis to detect other pathological lesions in other parts if any. We didn't need any other imaging procedures like local USG of the umbilicus or MRI of umbilicus and pelvis. Our patient had umbilical swelling with dusky discoloration of overlying skin and cyclical. In accordance to Victory et al⁴, umbilical swelling was present in almost 90% of patients with about 80% having umbilical pain. Pain is generated because of inflammation, distention and cyclical fluctuations. The size of the lesion is about 2.5 x 1.75 cm, with color changes ranging. Discoloration occurs as a result of cyclical hemorrhages and hemosiderin pigment deposition into the lesion as seen on histopathological examination. However, USG and MRI can be used to assess the nodule size and involvement of other tissues plus to assess and evaluate other pelvic pathologies (like uterine fibroids, ovarian cysts, other endometriotic sites, granulomas, miscellaneous and cancerous lesions as Sister Mary Joseph nodule etc.), thus helping in planning the surgical treatment. Our patient had no such additional pathologies. About 25% of umbilical endometriosis occurs along with simultaneous pelvic endometriosis. Infertility and Subfertility are common condition among patients with endometriosis, that may be as high as 50% of women with endometriosis¹. Our patient had neither of these two associations. Surgical excision or ablation is the only curative treatment of choice. Hormones may be used to ally symptoms. Hormones can also reduce the size of large lesions before surgery. But hormone therapy is associated with such side-effects such as amenorrhea.¹⁴ We treated our patient by traditional surgical excision and confirmed by histopathological examination. Umbilical endometriosis is said to have a very low risk of malignancy and recurrence.^{17,18}

Umbilical endometriosis is an uncommon disease, especially when it occurs primarily. The clinical presentation of umbilical swelling, cyclical pain, and occasional hemorrhagic discharge from the lesion are highly diagnostic. The treatment of choice is of course surgical excision, and the diagnosis is to be confirmed by histopathological examination, especially to exclude other diseases.^{10,11,18}

Conclusion

Umbilical endometriosis (primary or secondary) is a very uncommon benign disease that should be borne in mind as a differential diagnosis in women of reproductive age with any umbilical pathology. Diagnosis is mostly clinical by adequate history taking and careful physical examination. Most of the patients present with umbilical swelling, cyclical pain, discolored overlying skin with or without hemorrhagic discharge. FNAC can well aid in diagnosis. Imaging like USG or MRI has a valuable role for diagnosis and to exclude other pathologies. Histopathology is the final tool to arrive at a confirmatory diagnosis. Surgical excision or ablation is the only curative treatment of choice. It carries a very low risk of recurrence and malignant transformation.

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