

Villar's Nodule with Multifocal Endometriosis: Case Report

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Abstract

Umbilical endometriosis which was first described by Villar in 1886 is a rare clinical entity with a frequency of only 0.5-1%. The overall prevalence rate of endometriosis among women in their reproductive age is put at 5-10%. This is a descriptive presentation of a Villar's nodule with concurrent multifocal presentations paradoxically affecting the pelvic structures and the peritoneal wall in a patient with no prior history of a diagnosis or treatment for endometriosis. It is worth considering of the diagnosis in any given patient with nodules, cysts, bleeding and pelvic and/or abdominal pain in order to overcome the delayed and/or under diagnosis of the entity. To the best of our knowledge, this is the first ever reported enigmatic case from Ethiopia. It is intended to highlight aspects of issues pertaining to endometriosis and amplify its importance for academicians and practicing physicians in Ethiopia as an increasingly interesting phenomenon; and finally, to contribute to the world literature from our geographic location.

Keywords: Endometriosis, Umbilicus, Multifocal, Addis Ababa, Ethiopia

Introduction

The normal endometrial lining with its glands and stroma within the endometrial cavity proper and outside (endometriosis) is under constant ovarian hormonal influence during the reproductive years with cyclical and normative proliferative and secretory phases and shedding with its withdrawals resulting in the onset of menstruation as a reflection of the intactness of the hypothalamic-pituitary-ovarian-uterine and vaginal axis. Endometriosis is a benign condition that undergoes regressive changes following menopause and rarely evolves into an endometrial cancer. The pathology is basically a chronic gynecological disorder with marked aseptic inflammatory reactions as a result of overproduction of the tissue hormone prostaglandins and with the involvement of cytokine macrophages, Tumor Necrosis Factor (TNF) and many other factors resulting in adhesion, fibrosis, scarring and anatomical distortions manifesting in painful conditions. It may be running within the family members subsequent to genetic predisposition [1-3].

The endometrium-like tissue that may co-exist outside the endometrial cavity can be found within the myometrium (adenomyosis genitalis interna), fallopian tubes, ovaries, pouch of Douglas, uterosacral and round ligaments, cervix, vagina, vulva, ureter,

spleen, appendix and a group of distant extragenital occurrences involving the central nervous system, nose, pericardium, pleura and lung. A severe form of an inward continuous Deep Infiltrating Endometriosis (DIE) with a persistent and intense chronic abdominopelvic pain could also be a feature of endometriosis. Furthermore, Villar's nodule, a cutaneous umbilical type and the spontaneous or iatrogenic postoperative or ectopic implantations that are well documented could be regarded as similar variants affecting the bladder, rectum, vagina, vulva and the skin. The Villar's nodule is one of the rare and atypical types (Villar, 1886) and the list cannot be claimed to be exhaustive. Hence, the signs and symptoms could equally be diverse and very much dependent on the degree of involvement of the respective anatomical locations as it hardly spares any of the body organs [3].

The frequency of endometriosis, in as much as its etiology, is not exactly determined and it occurs more commonly among women in childbearing age of 15-44 years. In general, 6-10% of all women in their reproductive age develop endometriosis. Nevertheless, the relative distributions indicate that Villar's nodule is put at 0.5-1%, and 20% with pelvic pain, 25-50% with infertility and 75-85% with chronic pelvic pain and the extragenital accounting for 5%, respectively [4-8].

History and physical examination, as important as they are FNAC and histopathological studies of Villar's nodule, cutaneous endometriosis, easily accessible lower genital parts that include the vulva, vagina and cervix remain the corner stone for the establishment of the ultimate diagnosis in conjunction with laparoscopy or laparotomy for the internal body parts. In addition to abdominopelvic, transrectal, transvaginal sonographic evaluations including pelvic MRI with contrast, non-specific but yet contributory laboratory investigations may encompass the determination of ESR, CA125 and CEA [9].

The treatment provisions, though invariably are hormonal, may comprehensively embrace medical treatments like NSAIDs, danazol (antiprogestone), GnRh analogs and agonists, progestines, progesterone medicated Mirena devices, injectable like DMPA, combined oral contraceptive pills or postoperative corticosteroid and antihistamine administrations. Furthermore, minimally invasive laparoscopic, robot-assisted laparoscopic or invasive laparotomy designed broader choices need to be considered on individual and technical capabilities as deemed necessary [10].

This case report will eventually provide an up-to-date realistic narrative over endometriosis, appreciate the complex challenges it poses and promote compassionate right oriented approach and illustrate its holistic and individual management opportunities.

Case Description

The current case index was registered at our Medical Center as "EH" under card number 203/15. She is a divorcee with one vaginally delivered child of 11 years of age. She presented with a pur-

plish, brownish, puckered umbilical swelling and an abdominopelvic pain which was ill-characterized except as unceasing and with painful menstrual bleeding. The menstrual bleeding was darkish and relatively heavier. Same session clinical and ultrasound examinations lead us to the tentative clinical diagnosis of Villar's nodule and bilateral ovarian cysts with debris which was considered suggestive of a chocolate cyst.

From rights perspective, she was elaborated upon the treatment options all along her work up so that she can finally and freely understand the magnitude of her gynecological condition and establish consent of her choice at the end. She declined medical 'treatment only' offer and insisted and literally cried for a quick intervention at our disposal which was for the surgical alternative with a backup medical treatment. Accordingly, explorative laparotomy was performed. The findings demonstrated the fact that the pathology has massively involved the pelvic organ structures and its surroundings. The entered documents noted a big right ovarian and smaller multiple bilateral ovarian chocolate cyst, extensive deposits on the peritoneal surfaces and supporting ligaments with obliteration of the uterovaginal and rectovaginal septa. In conjunction with these findings, the surrounding structures were embedded within adhesions and making further dissection quite impossible; and the deeper structures inaccessible.

In fulfillment of the continual request of the patient for an urgent intervention and also understanding the severity of symptom complexes, the treatment modalities finally favored and concluded with simultaneous surgical and complementary six months course of dienogest, a synthetic progesterone administration.

Imaging Reports of Radiologists



Figure 1: There is minimal free fluid noted within the endometrial cavity with uterine dimensions of 8.90 cm length, height of 5.87cm and width of 3.70cm and uterine volume of 101.18ml. No free fluid seen in the pouch of Douglas.



Figure 2: There is a solitary intramural fibroid (F) at the fundus measuring 3.24cm x2.43cm.



Figure 3: Note the myoma, the endometrial cavity, and the ovarian cysts along with the urinary bladder.

The Complex Ovarian Cysts with Internal Echoes



Figure 4: Right ovarian cyst measures 7.6 x 4.3cm. There is tenderness on probe pressure application. The echocomplex is described as predominantly cystic adnexal mass with multiple septa and internal flow on color Doppler interrogation is quite remarkable. Ovarian cysts filled with blood forming the so-called chocolate cyst.

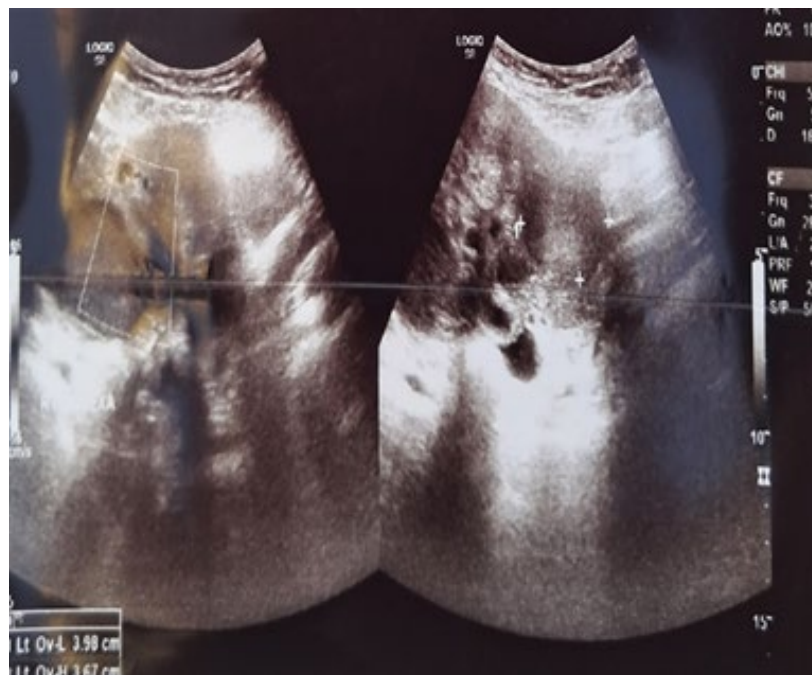


Figure 5: Left ovarian cyst measures 44.1mm x 36.7mm x 39.8mm (33.76cc) Normal arterial flow is demonstrated in Doppler interrogations.

Impressions based on Ultrasonography Reporting of Radiologists:

1. Complex bilateral ovarian cysts
2. Free fluid within the endometrial cavity is suspicious of inflammatory process
3. Solitary intramural myoma

Villar'S (periumbilical) Nodule



Figure 6:

Clinical note: Periumbilical dark-brown cystic lesion of 2.8cm x 4.1cm

FNA biopsy smears from periumbilical nodule disclosed cytologic features of cutaneous endometriosis (Villar's nodule) confirmed.

Conclusion: Periumbilical-Villar's nodule (Cutaneous endometriosis)

Clinical note: Periumbilical dark-Brown cystic lesion

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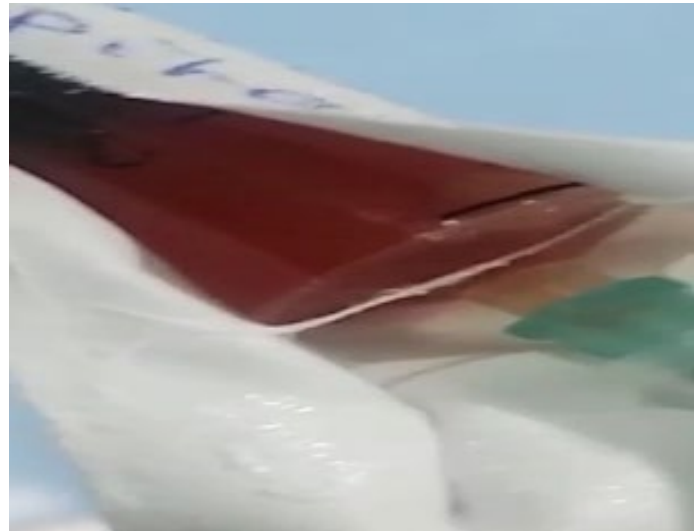


Figure 7: Surgical Findings

- right adnexal mass
- primipara, CPP and persistent right adnexal mass
- Intra-op - dark, cystic multiple masses with frozen pelvis
- CA 125 – 44
- biopsy taken from the cyst (ruptured) to r/o endometriosis, pelvic TB

Gross Finding:

- multiple gray-brown membranous tissue fragments, 3 cm in aggregate.
- cyst fluid sent for cytology – negative

Microscopy:

Section from the adnexal cystic lesion shows fibrotic and hemorrhagic cyst walls containing several glandular structures surrounded by endometrial-type stroma consistent with endometriosis.

Conclusion: Adnexa/ovary – endometriosis

Discussion

The current case under discussion that attracted our attention presented with umbilical endometriosis which remarkably mimics physiological or keloid scar, umbilical hernia, malignant melanoma and Sister Mary Joseph's nodule secondary to malignant intra-abdominal malignancy. Moreover, a preoperative concomitant ovarian (chocolate) cyst was entertained; and upon laparotomy multifocal other peritoneal seedlings involving the peritoneal wall, rectovaginal septum, vesicouterine space, uterosacral and round ligaments were discovered. Therefore, this atypical clinical presentation deservingly entails serious consideration and worth to be documented [11-16].

In the light of a multitude of signs and symptoms of endometriosis in general and pelvic pain in particular, one should always suspect this clinical entity as an invaluable differential diagnosis imagining also of the overlap with chronic pelvic inflammatory disease, infertility, types of ovarian tumors, adenomyosis genitalis interna and clinical manifestations of dyschezia, rectorrhagia, dyspareunia, dysmenorrhea, cyclical hematuria, nasal, rectal, respiratory and pericardial bleeding among many others. Thus, the hallmark of the triad of signs and symptoms of endometriosis unremittingly tend to present with mass, pain and bleeding internally or externally thereby emphatically influencing the quality of the life of the

affected patients, as in our case, with convincingly strong implication message [5,17,18].

No single theory of the postulated pathogenetic mechanisms can explain the clinical scenario of the patient. Hence it can only be the effect of the interplay or cumulative multi-theory operational phenomenon which include Sampson's theory of retrograde menstruation and implantation; colon coelomic metaplasia of multipotential cells transforming and developing into endometrial tissue within the peritoneal cavity according to Meyers theory; lymphatic, vascular and canalicular dissemination of endometrial cells impacting the external types (Halban's theory), the basal layer growth downwards into the myometrium after Cullen's theory as well as Mullerian remnants of dominant embryonic cells, genetic and immunological factors which can be pooled within the conundrum of the clinical pathology [19-23]. Thus, it is to be stressed that none of them is outmoded or disregarded as obsolete but collectively and constructively operational and academically merit-oriented as demonstrated in this report.

We exercised no critical appraisal of the many existing or proposed classifications and/or staging based on clinical or surgical as well as anatomical tangible findings because of personal and inter-personal variations in interpretation. The simplicity, applicability, reproducibility, lack of consensual comprehensive agreement or uniformity in the diagnosis, investigations and management modalities in accordance with specific type of endometriosis and its prognosis remarkably pause a great challenge. In the absence of conventionally accepted approach, the commonly applied classifications, to name several of the many, include rASRM, rAFS, EFI, AAGL, ESHRE and ENZIAN [24-26]. In our case, we have applied the four-stage classification system adopted by The American Society of Reproductive Medicine based on the massive adhesions and fibrosis, bilateral large ovarian cysts and multiple nearby and distant organ involvements whereby it was perceived as severe and Stage IV.

The easily accessible but non-specific diagnostic auxiliary measures include ESR, CA125, CEA and hysterosalpingography. The sensitivity and specificity of CA125 apparently is not indicative of the severity of the disease condition, its progress or response to treatment as much as it does in ovarian or endometrial cancers though widely used. More importantly Fine Needle Aspiration Cytology and tissue histopathological scrutiny directly from cutaneous or Villar's nodule or the retrieved tissue during laparoscopy or laparotomy will eventually pin down the ultimate diagnosis. Almost all of these complementary workups were of relevance and carried out in our case [27-30].

Endometriosis resembles a wide spectrum of clinical entities. The diagnosis, impact of treatment and prognosis should be gauged according to the intensity and persistence of the debilitating chronic pelvic pain, mass and cyclical bleeding from unusual anatomical sites. Hence, we emphasize that one should always think of en-

dometriosis and should not be merely forsaken for a diagnosis by exclusion!.

Therefore, the understanding of the incessant severity of the symptom complex of the patient, the treatment modalities favored in this particular case was simultaneous surgical and medical treatment with progesterone. We exercised caution in avoiding GnRH agonists and antagonists because of her age, induced menopausal complications and the need to supplementary add-back therapy with preparations containing estrogen and progesterone. It is ingenious to withhold estrogen therapy as it may exacerbate the clinical condition which is commensurate with the cyclicity of the menstrual changes [31-33]. En passé, we would like to remark that bilateral oophorectomy, though effective and attractive in managing endometriosis, should be absolutely withheld unless strongly indicated in association with BRCA mutation related ovarian, breast and endometrial cancers. Such prophylactic procedures to reduce the risks may be considered as tantamount to castration in women of such a reproductive age.

In general, endometriosis is a chronic gynecological disorder with debilitating mental, physical and social impacts and with immensely diverse clinical signs and symptoms affecting a significant segment of women in their reproductive ages. There is a strong need for early diagnosis with the intent of implementing timely and complete, effective and available treatment [34-36]. The presentation should be considered an eye opener with a window of hope to conduct a large scale, long term and prospective study on endometriosis under the watchful eyes of practitioners and physicians alike clinically and during laparoscopy or laparotomy.

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Abbreviations:

AAGL : American Association of Gynecological Laparoscopists
CA125 : Cancer Antigen
CEA : Carcino Embryogenic Antigen
DIE : Deep Infiltrating Endometriosis
DMPA : Depot Medroxy Progesterone Acetate
EFI : Endometriosis Fertility Index

ENZIAN : Endometriosis Classification
 ESHRE : European Society of Human Reproduction and Embryology
 ESR : Erythrocyte Sedimentation Rate
 FNAC : Fine Needle Aspiration Cytology
 GnRH : Gonadotropic Releasing Hormones
 MRI : Magnetic Resonance Imaging
 NSAIDs : Non-Steroidal Anti-inflammatory Drugs
 rAFS : Revised American Fertility Society
 rASRM : Revised American Society for Reproductive Medicine
 TNF : Tumor Necrosis Factor

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