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ENDOMETRIOSIS IN UNICORNUATE UTERUS WITH NON-COMMUNICATING RUDIMENTARY HORNE

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ENDOMETRIOSIS IN UNICORNUATE UTERUS WITH NON-COMMUNICATING RUDIMENTARY HORN

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SUMMARY

Endometriosis, defined as the presence of endometrial tissue outside the uterine cavity, is itself an enigmatic and multifaceted pathology, a puzzle whose manifold pieces remain largely disconnected despite some decades of investigation. Its medical history and clinical presentation is heterogeneous. Endometriosis affects approximately 10% of women of reproductive age, given its high prevalence and that women with endometriosis may have severe pelvic pain and infertility, endometriosis poses a significant health problem. Evaluating and understanding the pathogenesis, clinical presentation, diagnosis of the endometriosis is critical in the management of this debilitating disease. A rare case of endometriosis in a patient with congenital anomaly- Unicornuate uterus with one unconnected rudimentary horn is presented.

INTRODUCTION

Endometriosis is considered to be a chronic, estrogen dependant inflammatory condition with a complex pathophysiology and a highly variable clinical presentation. The true prevalence of endometriosis Worldwide remains obscure, and little is known about its distribution in the population and its risk factors (1,2,3). Endometriosis affects approximately 10% of women of reproductive age, given its high prevalence and that women with endometriosis may have severe pelvic pain and infertility, endometriosis poses a significant health problem (2, 3, 5). Although the prevalence of endometriosis is well documented in the developed world, studies on the prevalence of this disease among African population are still scarce (4). Although, the African American has typical implantation sites of endometriosis, they have a higher preponderance of atypical sites including Uterus (6).

A unifying theory regarding the origin of endometriosis has remained mystifying elusive and several theories have been suggested (7). Endometriosis, however, has probably multifactorial in origin.

Retrograde Menstruation; Initial proposed by Sampson in 1927, the theory of retrograde implantation is both intuitively attractive and supported by multiple

lines of scientific evidence. The theory proposes that endometriosis occurs due to sloughed endometrial cells debris via the fallopian tubes into the pelvic cavity during menstruation (8,9).

Endometriosis is more common in women with autoimmune disease supports the possibility that pathogenesis of endometriosis may involve a defective immune response in these clients (10,11). Coelomic metaplasia theory postulates that endometriosis originates from the metaplastic specialised cells that are present in the mesothelium lining of the visceral abdominal peritoneum (12). Hormonal or immunological factors are thought to stimulate the transformation of normal peritoneum tissues/cells into endometrium-like tissue (12). Studies indicating that 7-9% of endometriosis patients' first-degree female relatives (sisters etc.) are diagnosed with the disease compared to the control rate of 1-2% suggest a genetic influence in the aetiology of the disease (13,14).

Environment factors such as early marriage, frequent pregnancies, breastfeeding and high incidence of pelvic inflammatory disease have been explained for the low incidence of endometriosis in Africans (15). With the change of lifestyle and social economic status among the Africans, the incidence of endometriosis is expected to increase. The objective of this paper is to describe the clinical presentation, diagnostic challenges, benefits and drawback for both

surgical and medical management of endometriosis.

CASE REPORT

Twenty one years nulliparous who was admitted with acute on chronic pain on the left ilio-lumbar region and severe dysmenorrhoea for the last four months. There was no history of pelvic discharge, ordyspareunia. The past medical history was insignificant and there was no history of contraceptive use. On examination, she was distressed, not pale, and afebrile. She had tenderness with guarding on the left ilio-lumbar region but no mass was felt. On pelvic examination, the uterus was normal size, right adnexal was normal, the left adnexal was tender with a mass and cervical excitation test was positive.

Investigations:

Haemoglobin: 12.4gm/dl, White blood cell: 8.0
Serum Beta HCG: Negative
CEA: 0.74
CA125: 16.4
FSH: 4.96

Transvaginal Ultrasonography: Uterus was normal size, endometrial stripe was normal, a solid left adnexal mass measuring 35x27x25mm = 12 grams was found. The mass was vascular on colour Doppler. Bilaterally the ovaries appeared normal. The conclusion was left adnexal mass aggravating to 12 grams.

Laparoscopy surgery: Laparoscopy was performed under general anaesthesia. Mini-lap incision was made intra-umbilical 10 mm trocar introduced; insufflation was done with carbon dioxide and pneumoperitoneum maintained at a pressure of 15mmHg. The patient was placed in trendelenberg position and the 2 lateral ancillary 5 mm trocars introduced.

Findings: Unirconuate uterus with left unconnected rudimentary horn, right ovary and fallopian tube was normal. The left ovary had an endometrioma. An endometriotic nodule was found at the sigmoid epiplo and superficial endometriosis on the left rudimentary umbilical artery. There was no endometriosis visualised on the right side of the pelvis. The endometriosis was graded as stage 3.

Done: Excision of the sigmoid epiplo nodular, left ovarian endometrioma cystectomy and coagulation of the endometriosis on the left rudimentary umbilical artery. The standard management of the rudimentary

horn is excision to avoid future complications, however this was not done due to lack of the patients consent. The tissue excised was taken for histology. There was no complication during the laparoscopic surgery. Post-operative the patient had administration of analgesia and antibiotics. The patient had fully recovery.

Histology results:

Left Ovary: Endometriotic foci.

Sigmoid epiplo: Endometriotic nodule.

Medical treatment: She was prescribed for Dionogest 2mg back to back treatment for six months. Subsequently, she was to conceive as she had wished.

Figure 1

Endometriotic nodule on the sigmoid epiplo



Figure 2

Endometriosis on the left rudimentary umbilical artery



Figure 3

Uterus unicornuate with left unconnected rudimentary horn



DISCUSSION

Endometriosis affects women of the reproductive age during the prime years of their lives – almost two thirds of the patients are younger than 30 years of age at their first consultation and its impact is complex, multidimensional and pervasive (16,17). The symptoms include dysmenorrhoea, deep dyspareunia, and chronic pelvic pain. The clinical signs of endometriosis include tender lower abdomen and localised dark blue nodules if external. Pelvic examination reveals tender nodules in the cul-de-sac and along the uterosacral ligaments. The uterus is often fixed in the retroverted position (due to adhesion) while the ovaries may be enlarged (due to endometriomas) or not felt at all owing to distorted pelvic anatomy. Generalised pelvic tenderness with positive cervical excitation test mimics findings on patients with pelvic inflammatory disease. The patient presented was 21 years with history of severe pelvic pain and dysmenorrhoea, clinically she had presented with pelvic tenderness with guarding and per vagina examination, revealed cervical motion tenderness and left adnexal mass.

The diagnosis is based on the patient's comprehensive history, physical examination and surgery (laparoscopy or laparotomy). In addition magnetic resonance imaging (MRI), ultrasonography (particularly transvaginal) and other ancillary diagnostic procedures unique to the clinical presentation may be used to diagnose the disease. However, lack of appropriate non-invasive tool is the main limiting factor for the diagnosis, monitoring the disease progression, predicting clinical outcome, and evaluating therapeutic effects even when medical/expectant management has been decided (18). One of the principle challenges actually involved in managing endometriosis is obtaining information that might help decision-making and allow gynaecologist

to select the optimal surgical strategy, even before surgery.

Endometriosis is a complex disease and treatment decisions are multifactorial. Factors such as age, history, desire for pregnancy and treatment preferences are likely to influence treatment decisions. There are benefit and drawback for both surgical and medical management of endometriosis. Treatment of endometriosis should be individualised, and may include surgery, the medical approach or both.

The transvaginal ultrasonography in this case was not conclusive in making diagnosis. The diagnosis was made by laparoscopy, which revealed unicornuate uterus with non-communicating rudimentary horn and with superficial endometriosis, left ovarian endometrioma and sigmoid apiplo endometrial nodule. Laparoscopic visualisation remains the gold standard for the diagnosis of the endometriosis. The use of narrow band imaging at laparoscopy has been found to be beneficial (19).

Unicornuate uterus is formed when one mullerian ducts completely or incompletely fails to elongate while the other develops normally. Unicornuate uterus accounts for approximately 2.4-13% of all mullerian anomalies. The unicornuate may occur alone, but it is frequently associated with a rudimentary horn. Non-communicating accessory horns that have an endometrial activity are the most common unicornuate subtype that is most clinically significant. Ectopic pregnancy, with a rate of 2.7%, has been found as a complication of unicornuate uterus. When the accessory horn becomes obstructed, severe complications, such as hematometra and endometriosis occur. The endometriosis in this case was as a result of retrograde menstruation from the non-communicating rudimentary horn as proposed by Sampson (19). The patient had presented with chronic pelvic pain and dysmenorrhoea, consisted with endometriosis. Endometriosis should be ruled out in adolescent and young women who present with congenital genital anomaly such as unicornuate uterus with rudimentary horn.

Surgical removal of lesions is effective at reducing pain associated with endometriosis and consequently increases quality of life (20-23) Additionally, surgical removal of endometriosis lesions in severe forms of the disease may associated with increased chances of fertility, important for those wishing to conceive (24). However, surgery is associated with increased risk of reduced ovarian function and in cases of deep infiltrating endometriosis with morbidity and complications such as bladder, intestinal and sexual dysfunctions, and fistula (25,26). The patient presented had excision of sigmoid epiplo nodule, ovarian endometrioma cystectomy and superficial endometriosis coagulation. However, a consistent body of literature has demonstrated that repetitive surgical procedures are associated with an increased

risk of complications and little improvement of symptoms and fertility. Long-term medical treatment is advocated.

Although definitive diagnosis of endometriosis requires a laparoscopy, empiric medical treatment has been advocated to not only treat symptoms but also to make a presumptive diagnosis of endometriosis (27). Multiple medical treatment options exist, non specific treatment are combined oral contraceptive (COCs) and anti-inflammatory agents, while specific options are progestin's and Gonadotropins releasing hormone analogues. The use of COCs is well established as an endometriosis treatment option, described in several guidelines. However, it should be noted that these medications do not have an approved indication for that purpose. COCs are not the choice for management of patients with endometriosis associated with pain (28,29).

Progestin's, such as dienogest, act to reduce serum estrogens levels, cause desidualisation and atrophy of endometrial tissue and have anti-inflammatory and analgesic effect, making them an effective treatment option for managing endometriosis symptoms (30-33).

The patient presented was commenced on dienogest 2 mg for six months and subsequently for conception as per her desire.

In conclusion, endometriosis is being increasingly diagnosed amongst the younger African population. In view of the unicornuate uterus with non-communicating rudimentary horn that was found in this patient, it was our conclusion that congenital genital tract anomalies need to be ruled out in young patients suspected to have endometriosis and that there's need to take consent for possible removal of rudimentary horn at surgery.

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