Appendiceal Endometriosis: A case Report and Literature Review

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SUMMARY

Appendiceal endometriosis is a very rare and usually asymptomatic condiction, but can result in severe complications such as intestinal perforation, massive gastrointestinal bleeding or intussusception. We report a case of endometriosis of the appendix presenting as acute appendicitis. The patient was a 36 year old nulliparous woman who was scheduled for myomectomy for 20 weeks sized uterine firbroids. She however developed a right ovarian chocolate cyst, pelvic endometrial deposits and multiple uterine fibroids. Myomectomy and right ovarian cystectomy was done in addition to appendectomy. Histological examination revealed endometrial deposits within an oedematous wall of the appendix and a diagnosis of endometroisis of the appendix causing acute appendicitis was made. We considered the relevant literature on appendiceal endometroisis and its association with endometrioma. We recommend routine evaluation of the appendix during pelvic surgery especially surgery for endometriosis.

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INTRODUCTION

Endometriosis is defined as the presence of ectopic endometrial tissue outside the lining of the uterine cavity and is a fairly common condition affecting women of the reproductive age group. It is more common in the industrialized countries affecting about 15% of fertile and 50% of infertile women[1,2]. However, endometriosis of the gastro intestinal tract is rare and appendiceal endometriosis presenting as acute appendicitis is even rarer[3]. It is reported to have a high incidence of association with leiomyomata of the uterus and pelvic endometriosis[3]. We report a case of endometriosis of the appendix presenting as appendicitis in association with pelvic endometriosis and leiomyomata of the uterus.

CASE REPORT

A 36 year old nulliparous woman presented to us with a 4-month history of lower abdominal mass and excessive menstrual flow. Her last menstrual period had been two weeks earlier but her menstrual cycle had been regular, though associated with dysmenorrhoea. Physical examination revealed a lower abdominal mass arising from the pelvis of about 20-weeks size, firm with irregular surface and non tender. Pelvic examination revealed a right cystic adnexal mass that was not tender. Cervical excitation tenderness was negative. An impression of uterine fibroids was made and an abdominopelvic ultrasound scan was done which revealed multiple uterine fibroids and chocolate cyst of the right ovary. She was counselled and booked for myomectomy.

However, five days later, she was admitted into the hospital because of increasing right lower abdominal pain associated with nausea and vomiting. Her temperature on admission was 38.4°C. Localized tenderness and rebound tenderness were detected over the right iliac fossa. Pelvic ultrasound did not show any new findings. Her white blood cell count was 12,300/mm^3. The routine serum biochemistry profile was normal. An impression of appendicitis was made. The patient had a laparotomy via a subumbilical midline incision. Appendectomy, myomectomy and right ovarian cystectomy were performed. Intra-operative findings included an inflamed appendix, multiple endometrial deposits within the pelvis, a chocolate cyst on the right ovary and multiple uterine fibroids.

Histological examination of the appendix revealed moderate oedema of the muscular wall of the proximal end with focus of endometrial tissue, including dilated endometrial glands and stroma (Figs. 1&2). Also seen were compact clusters of adipocytes. The histopathological diagnosis was endometriosis of the appendix causing appendicitis. She made an uneventful postoperative recovery and was discharged home one week after the surgery.

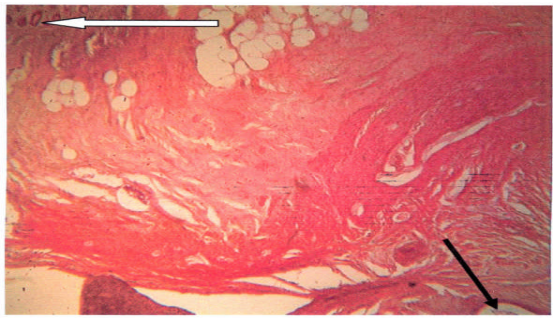

Figure 1: Picomicrograph showing focus of endometrial tissue (thin arrow) in an oedematous muscular wall of vermiform appendix. Thick arrow points at the appendiceal glandular epithelium; x l 00. Haematoxylin & Eosin stain used.

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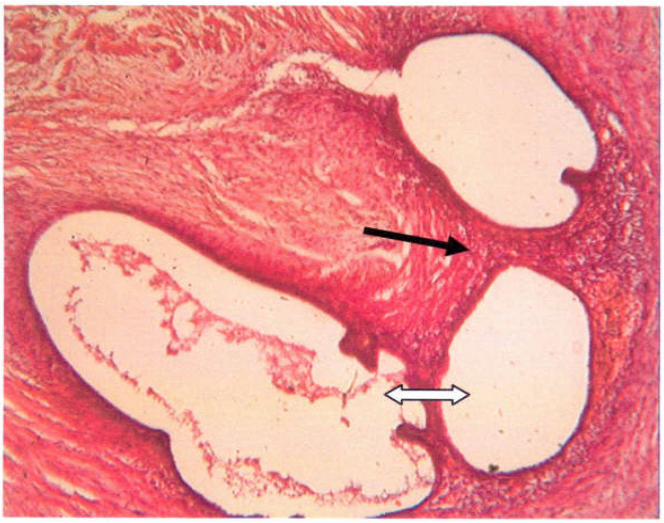

Figure 2. Picomicrograph shows lumin. of endometrial glands (thick double-headed arrow) and stroma (thin arrow); x400. Haematoxylin & Eosin stain used.

DISCUSSION

Endometriosis primarily affects the genital structures such as the ovaries, the uterosacral ligament, and the rectovaginal septum[4], but sometimes extragenital involvement of the gastrointestinal tract, the ureters, the pleura and lungs can occur with variable presentations [1,5,6]. We had previously reported a case of endometriosis causing pleural effusion and haemoperitoneum [5].

Endometriosis of the appendix is very uncommon accounting for only about I% of all cases of endometriosis [1,7]. However Douglas and Rotimi reported an incidence of 5.9% of all extragenital endometriosis cases[1]. It is usually asymptomatic but occasionally can present as appendicitis, intestinal perforations, intussusception or acute lower gastrointestinal bleeding [1-3,6,8-11].

On account of its asymptomatic nature, the possibility of pre operative diagnosis is very remote. The diagnosis is usually based on the histological finding of endometrial gland and stroma within either the wall or the lumen of the appendix. The only clinical feature that supports the suspicion of appendiceal endometriosis may be the long history of right lower abdominal pain with an intermittent course in a woman known to have endometriosis. Ovarian endometriosis has been shown to be a marker for the presence of an extensive pelvic and intestinal disease[4]. Therefore it is very important to critically examine the appendix during surgical treatment of such cases.

The place of routine removal of the appendix in the surgical treatment of endometriosis is still controversial. While many authors recognize the need for incidental appendectomy for a limited class of patients such as those with symptoms suggestive of appendiceal disease or those in whom the appendix is grossly involved in the disease process[12,13], others recommend routine appendectomy even when the appendix is grossly normal in appearance, arguing that the intraluminal contents can not be examined accurately without an appendectomy[14]. In a recent review, Hie Jong Wie et aI[15] reported that 34 out of 37 women with ovarian endometrioma who had grossly normal-appearing appendix, had abnormal histopathological findings including 12 (35.3%) women with appendiceal endometriosis.

The mechanism through which appendiceal endometriosis occurs is still unknown. In cases associated with pelvic endometriosis, as in our case, the conventionally held opinion that endometriosis of the appendix occurs as a direct extension of endometriosis of the right ovary seems reasonable but it cannot account for cases of isolated appendiceal endometriosis. In these cases the metaplasia theory (de-differentiation of the coelomic epithelial lining of the peritoneal cavity) or the systemic embolization theory seems more plausible.

The most important issue in the treatment of this rare condition is the gynaecological assessment and follow up. Appendectomy usually cures the acute symptoms but for the relief of long- time symptoms, drug therapy, particularly hormonal therapy may be considered.

In conclusion, appendiceal endometriosis is rare and can mimick acute appendicitis. Routine evaluation of the appendix is recommended during gynaecological surgeries especially surgeries for endometriosis.

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