Endometriosis of the appendix presenting as acute appendicitis

J J Khoo, M S A Ismail, C C Tiu

ABSTRACT

While endometriosis is fairly common, endometriosis of the appendix is a very rare occurrence. Correct pre-operative diagnosis is uncommon and definitive diagnosis is established by histology of the appendix. We present a case of endometriosis of the appendix that manifested as acute appendicitis in a 40-year-old woman.

Keywords: acute appendicitis, appendix, endometriosis

INTRODUCTION

Endometriosis is defined as the presence of ectopic endometrial tissue outside the lining of the uterine cavity and is fairly common in childbearing women. However, involvement of the gastrointestinal tract is uncommon and endometriosis of the appendix is an even rarer occurrence. Endometriosis of the appendix manifesting as acute appendicitis is exceedingly rare. We report a case of appendiceal endometriosis clinically presenting as acute appendicitis, and aim to highlight this rare entity.

CASE REPORT

A 40-year-old Malay woman was admitted with a 5-day history of right iliac fossa pain. She had no associated symptoms of fever, nausea, vomiting or anorexia. She denied any unusual vaginal discharge. Her menses had been irregular and heavy, with occasional dysmenorrhoea. Her last menstruation was three weeks before admission. She was seen regularly in the gynaecology clinic for 2.5 years for primary infertility. Two years ago, she was operated for an intramural leiomyoma and a left ovarian epithelial cyst, with no evidence of endometriosis. She had received hormonal therapy for infertility. Two months prior to admission, investigations showed that she had bilateral hydrosalphinx.

On admission, she had a low-grade fever of 37.9°C. Pain was localised to the McBurney’s point, with tenderness, guarding and rigidity. Vaginal examination did not show any tenderness when the cervix was rocked, nor was there any adnexal mass or pain at the fornices. The white blood cell count was 15,400/mm³ with 73% segmented neutrophils. Urine analysis was normal and urine pregnancy test was negative. The tentative diagnosis was acute appendicitis and an appendectomy was done.

At surgery, the peritoneal cavity was relatively clean with minimal fluid in the cavity. Intra-operatively, the appendix appeared mildly congested. The appendix measured 4.5 x 0.5 cm at the widest diameter. It was grossly normal and the excised section showed intact mucosal epithelium and a patent lumen (Fig. 1). Histological examination showed several ectopic endometrial glands with stroma in the muscular propria and subserosa of the appendix (Fig. 2).

Department of Pathology
Sultanah Aminah Hospital
Jalan Skudai
80100, Johor Bahru
Johor, Malaysia
J J Khoo, MBBS, MPath
Consultant Pathologist
M S A Ismail, BMedSc, MBBS
Medical Officer

Department of Surgery
C C Tiu, MBchBAO, MS
Clinical Specialist

Correspondence to:
Dr Khoo Joon Joon
Tel: (607) 223 1803
Fax: (607) 223 3303
Email: jjoon60@hotmail.com
appendix (Fig. 2). Some of the endometrial glands were dilated. Minimal fresh haemorrhage was noted with moderate lymphoid hyperplasia in the lamina propria, but no significant acute inflammation was seen in the appendix. The histological diagnosis was appendiceal endometriosis with no microscopical evidence of acute appendicitis.

Post-operatively, the patient recovered with no residual pain. However, six weeks later she returned with vague lower abdominal pain. Ultrasonography showed multiple small cystic lesions in both ovaries which were not seen previously. She was advised for admission, but the patient refused and defaulted on her follow-up.

**DISCUSSION**

Isolated endometriosis of appendix is very rare. It is usually asymptomatic, but occasionally causes symptoms such as appendicitis(1-3), perforation(4-5), intussusception(6-7) or acute lower gastrointestinal bleeding(8). While it may have various clinical presentations, there is no means of pre-operative recognition of appendiceal endometriosis. Our patient presented clinically as acute appendicitis with a raised leucocyte count. Although she was seen frequently in the gynaecology clinic for infertility with bilateral hydrosalphinx, appendiceal endometriosis was never suspected pre-operatively.

Endometriosis of the appendix is reported to have a high incidence of association with leiomyoma of the uterus and menstrual abnormalities(12-13). Some authors even reported symptoms of abdominal pain with menstruation(14). Our patient had an abnormal menstrual pattern and had an intramural leiomyoma resected previously. However, she menstruated only five days after the appendectomy. Her symptoms did not coincide with her menstruation.

Gross inspection of the appendix does not give any hint of the disease. The appendix may appear grossly normal(15). The diagnosis of appendiceal endometriosis is based on the histological presence of endometrial glands and stroma, with or without haemorrhage. The presence of recent haemorrhage was thought to be responsible for the acute symptoms. In our patient, there was minimal haemorrhage in the lamina propria of the appendix and this could account for pain in the right iliac fossa.

In their series, Mittal et al(15) found that 56% of endometriosis of the appendix involved the body of the appendix, compared to 44% at the tip. The base was not involved in any of their cases. They also noted that muscular and seromuscular involvement occurred in two-thirds of patients, while the serosal surface was involved in only one-third of patients. Mucosa was not involved in their series. However, Langman et al(15) found that the submucosa was involved in one-third of patients with endometriosis of the appendix. In their series, the endometriotic foci were also found in the muscle, serosa and subserosa. There was no correlation between the histological location of the endometriotic foci and the patients’ symptoms(2-10).

All patients with appendiceal endometriosis who had a preoperative diagnosis of acute appendicitis recovered after the appendectomy(1,3). In our patient, the acute abdominal symptoms disappeared completely, but lower abdominal pain recurred. This was probably due to associated pelvic (ovarian) endometriosis. A gynaecological assessment should be done to gauge the extent of endometriosis. Post-operative follow-up is mandatory for appendiceal endometriosis. In cases where associated extraintestinal endometriosis is proven or suspected, hormonal manipulation (whether medical or surgical) may be necessary to treat the symptoms.

In conclusion, appendiceal endometriosis is rare and almost never correctly diagnosed pre-operatively. It may be suspected when associated with obvious pelvic endometriosis. Definitive diagnosis is only established by microscopical examination of the appendix. Appendectomy cures the acute symptoms.

Post-operative follow-up with referral to the gynaecologist may be necessary.

**REFERENCES**