Endometriosis of the appendix: A diagnostic dilemma

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Received: 30 August 2009 / Accepted: 7 January 2010
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Abstract The incidence of endometriosis of the appendix is reported to be just <1% of the total endometriosis cases. Laparoscopic visualization and diagnostic yield are of profound significance to women in whom gynecologic diseases such as endometriosis, pelvic inflammatory disease, ruptured ovarian follicles or tubal pregnancy may be the source of pain and mimic appendicitis. Therefore, lower abdominal pain in reproductive age group female must invite the attention of the surgeon or gynecologist to entertain the possibility of endometriosis since appendiceal endometriosis, while relatively uncommon in patients with endometriosis, is rare in the general population.

Keywords Endometriosis · Appendix · Lower abdominal pain · Reproductive age group female

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 [1]. The incidence of endometriosis of the appendix is reported to be just <1% of the total endometriosis cases [2].

Because of its propensity to manifest with a wide spectrum of site-specific presentations, appendiceal endometriosis should be a diagnostic consideration in the evaluation of pre menopausal women with gastrointestinal complaints [3].

Case report

A 20-year-old married, nulliparous female presented in the emergency with complains of acute lower abdominal pain of 5 days duration. The pain was dull, continuous and was localized more towards the right iliac fossa. Her menstrual period was regular, of 28–30 days duration with no history of dysmenorrhoea. The patient had her period 5 days before the onset of abdominal pain. There was no history of rectal bleeding, vaginal discharge, fever or vomiting. Per abdominal examination showed tenderness in the lower abdomen with no rigidity, guarding, or any palpable mass. Her routine haematological investigations were unremarkable except for mild leucocytosis and slightly raised C-Reactive Protein. Keeping in mind the clinical presentation of the patient, a preoperative provisional diagnosis of acute appendicitis or right tubo-ovarian mass was made and abdominal ultrasound was suggested which was unremarkable. The patient underwent diagnostic laparoscopy, which revealed adhesion of the tip of the appendix to the right adnexae along with minimal

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hemorrhagic fluid over the broad ligament. An occasional punctate hemorrhagic foci involving the serosal surface of the right ovary along with a ruptured luteal cyst was noted. Since the patient was nulliparous and had no history of menstrual irregularities, appendectomy was performed and the ovary was preserved. The postoperative period was uneventful and the patient was discharged on the 3rd postoperative day in a stable condition.

Histopathological findings

The resected appendix measured 6 cm in length and the wall was slightly edematous. Microscopically, besides lymphoid hyperplasia, a few groups of endometrial glands along with endometrial stroma were found embedded in the muscularis propria of the tip of the appendix (Figs. 1 and 2).

Fig. 1 Endometrial glands along with compact endometrial stroma embedded in the muscularis propria of the appendicular wall (H and E, 400X)

Fig. 2 The appendicular wall showing endometrial glands with stroma within the muscularis propria (H and E, 40X)

After the diagnosis of appendiceal endometriosis was established, patient was referred to gynaecology OPD for opinion and further management.

Discussion

Endometriosis of the appendix is asymptomatic and often found incidentally during operative treatment for pelvic endometriosis. It may present as a cause of lower abdominal pain and affects about 15% of females in the reproductive age [4]. The clinical diagnosis of intestinal endometriosis may be difficult because often symptoms are non specific and there is no relationship between the symptoms and the menstrual cycle [5]. The correct diagnosis is often delayed as it may present as appendicitis, mucocoele, or appendicular mass mimicking a neoplasm [3].

The most widely accepted mechanism for development of endometriosis is the retrograde menstruation through the fallopian tube into the peritoneum, with subsequent growth of the displaced cells. Involvement of distant sites such as the pericardium or pleura is best explained by the vascular dissemination of endometrial cells [6].

Endometriosis of the gastrointestinal tract can be localized by the frequency of occurrence to the recto sigmoid (72%), recto vaginal septum (13%), small bowel (7%), caecum (4%), appendix (3%) and other intestinal sites (0.5%) [7].

Mittal et al. found that endometriosis of the appendix involved the body in 56% and the tip in 44% cases. They also noted that muscular and sero-muscular involvement occurred in two-thirds of patients, while the serosal surface was involved in only one-third of patients [8]. The present case also corroborated well with their findings as endometriosis involved the tip of the appendix with involvement of muscularis propria.

There are no sufficiently sensitive and specific signs and symptoms or diagnostic tests for the clinical diagnosis of endometriosis. An important component of the evaluation is a careful pelvic examination including combined rectovaginal palpation which may reveal tender nodules or irregularities in the cul-de-sac and it should be done immediately before or after the menstruation. Since patient was nulliparous and endometriosis was not among the provisional diagnosis, a rectovaginal examination was not performed [5].

The preferred method for the diagnosis of endometriosis is direct visualization of ectopic endometrial lesions (usually via laparoscopy) accompanied by histological confirmation of the presence of at least two of the following features: Hemosiderin laden macrophages or endometrial glands with or without stroma [9]. Laparoscopic visualization and diagnostic yield are of profound significance to women in whom gynecologic diseases such as endometriosis, pelvic inflammatory disease, ruptured ovarian follicles or tubal pregnancy may be the source of pain and mimic appendicitis

[6]. This case also underwent diagnostic laparoscopy followed by histopathological examination of the specimen leading to confirmatory diagnosis of endometriosis of the appendix. Retrograde menstruation or a ruptured luteal cyst might be the reason for the presence of minimal hemorrhagic fluid seen over the broad ligament.

Cyclic response of endometrial tissue to hormones may lead to serosal irritation and progressive invasion of intestinal muscle with fibrosis and muscle hypertrophy. Pain may arise from nerve impingement or serosal inflammation whereas obstruction may occur from luminal narrowing or intestinal kinking [5]. As with endometriosis elsewhere in the intestinal tract, endometriosis of the appendix may result in perforation or intussusception [6]. Treatment may be expectant, or a patient may choose either medical or surgical options. Surgical treatment is the preferred approach in patients with advanced endometriosis. The benefit of surgery in these patients may be due entirely to the mechanical clearance of adhesions and obstructive lesions [10].

To conclude, lower abdominal pain in reproductive age group female must invite the attention of the surgeon or gynecologist to entertain the possibility of endometriosis since appendiceal endometriosis, while relatively uncommon in patients with endometriosis, is rare in the general population.

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