Quiz

Correct answer to the quiz. Check your diagnosis

Case report

Intramural florid cystic endosalpingiosis of the uterus after menopause

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Endosalpingiosis is a benign condition characterized by the presence of tubal epithelium outside the fallopian tube and the absence of endometrial stroma. Florid cystic endosalpingiosis is a very rare form of endosalpingiosis that presents as a tumor-like lesion. We report the case of a 67-year-old woman who presented with a cystic lesion of the uterus. Macroscopically, a cut section revealed a multicystic, whitish mass in the myometrium of the fundus. Histologically, the lesion consisted of numerous variably sized glands that were lined with a single or stratified layer of ciliated columnar cells similar to tubal epithelium.

Key words: florid cystic endosalpingiosis, endosalpingiosis, after menopause, gynecological pathology, immunohistochemistry.

Introduction

Endosalpingiosis (ES) refers to the presence of benign glands lined by tubal-like epithelium. Endosalpingiosis appears to be a reasonably common condition and was reported to be found in 7% of woman undergoing laparoscopy [1]. The most common sites are the serosa of the uterus and the fallopian tubes [2]. Endosalpingiosis is mostly an asymptomatic lesion with no serious prognostic associations [3]. However, it may rarely present as a tumor-like lesion and can be confused with a neoplasm clinically. Florid cystic endosalpingiosis (FCE) is a rare form of ES that presents as a tumor-like lesion. FCE involving the uterus is rare, and only 18 cases have been reported in the English literature. According to its location, FCE can be categorized into two types, intramural or subserosal [4]. Six reported cases and the present case are classified as the intramural type of FCE (4–8). In addition, most cases of FCE are usually found in women of reproductive age of less than 55 years [4]. This report presents an extremely rare case of an intramural FCE that occurred after menopause.

Case report

A 67-year-old woman, gravida 2, para 2, with an endometriotic cyst had previously undergone

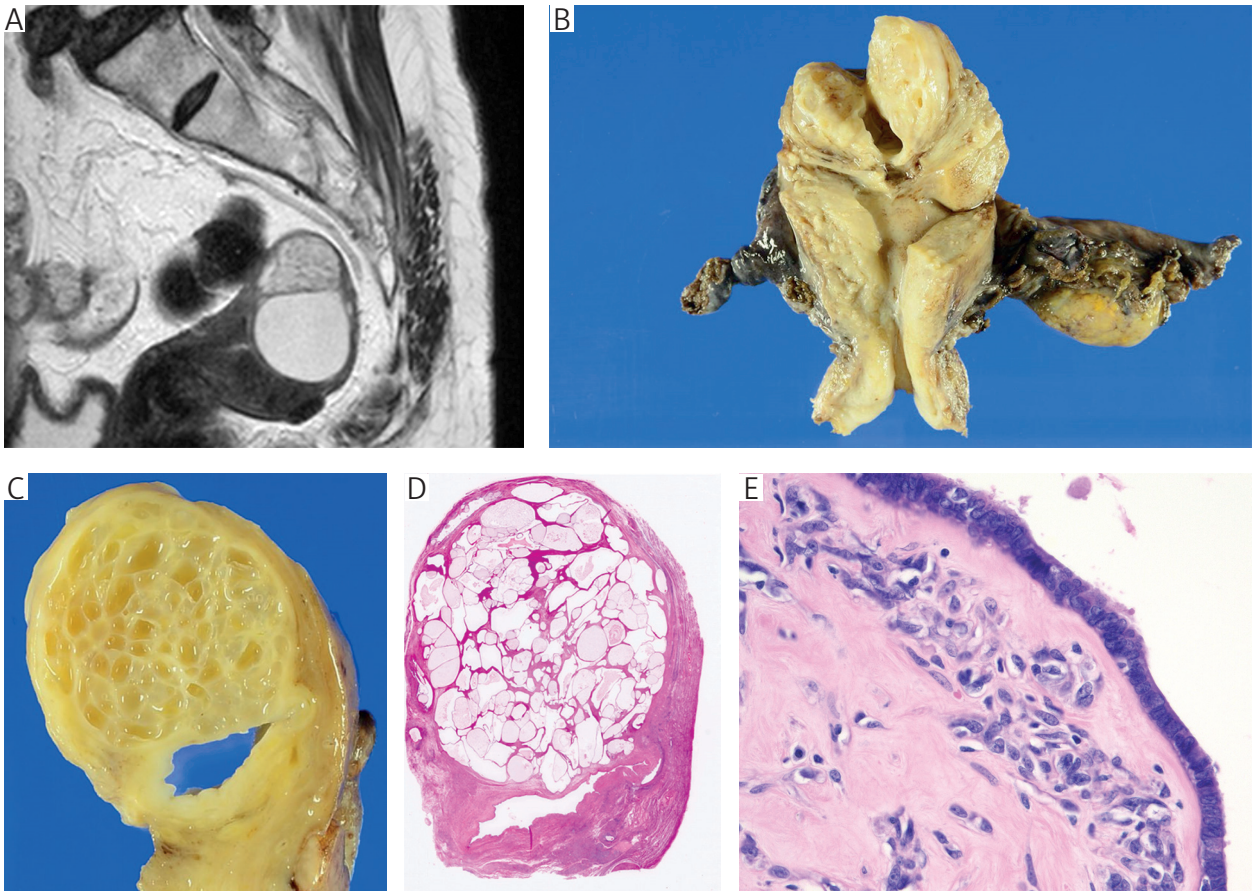

Fig. 1. A) Magnetic resonance imaging (MRI) revealed a 36-mm intramural hyperintense cystic lesion in the uterine fundus. B, C) Macroscopically, the cut section revealed a 30 × 25-mm multicystic, whitish mass in the myometrium of the fundus. D, E) Histological examinations showed multiple cysts that were lined with a single layer of ciliated columnar cells similar to tubal epithelium

a right salpingo-oophorectomy. She presented to our hospital with a history of pain in the lower abdomen for about 3 months. An ultrasound showed a 40–45-mm mass with multiple cysts in the left side of the uterine corpus. Magnetic resonance imaging revealed a 36-mm intramural hyperintense cystic lesion in the uterine fundus (Fig. 1A). Degenerated leiomyoma was suspected clinically, and the patient underwent total hysterectomy.

The resected specimen consisted of an atrophic uterus with attached fallopian tubes and left ovary. Macroscopically, a cut section revealed a 30 × 25-mm multicystic, whitish mass in the myometrium of the fundus (Fig. 1B, C). Multiple leiomyomas with a maximum size of 1.0 cm were also present in the uterine corpus.

Histological examinations showed multiple cysts that were lined with a single layer of ciliated columnar cells similar to tubal epithelium (Fig. 1D, E). The lack of cellular stratification and especially of any mitotic activity, and the slight nuclear atypia, displayed no evidence of malignancy. The differential diagnosis was adenomyosis/adenomyoma or adenomatoid tumor. However, the endometrial stroma was not accompanied by cysts. Immunohistochemical analysis revealed that the epithelial cells were positive for estrogen receptor (ER) (Fig. 2A), progesterone receptor (PgR) (Fig. 2B) and paired box gene 8 (PAX8) (Fig. 2C), and negative for calretinin. Mucin was present in the apical cytoplasm and was retained within the glandular lumens (Fig. 2D). The pathological diagnosis was FCE.

Discussion

Endosalpingiosis is typically found in women of reproductive age (mean age 29.7 years) [2], although it occasionally occurs after menopause. Endosalpingiosis appears to be a reasonably common condition, with one report suggesting that it affected 7% of women undergoing laparoscopy [1]. The most common sites are the serosa of the uterus and the fallopian tubes, and less frequent sites include the paraovarian area, omentum, urinary bladder, ureter, spleen, colon, appendix and lymph nodes [2]. Histologically, ES refers to the presence of ectopic tubal-type

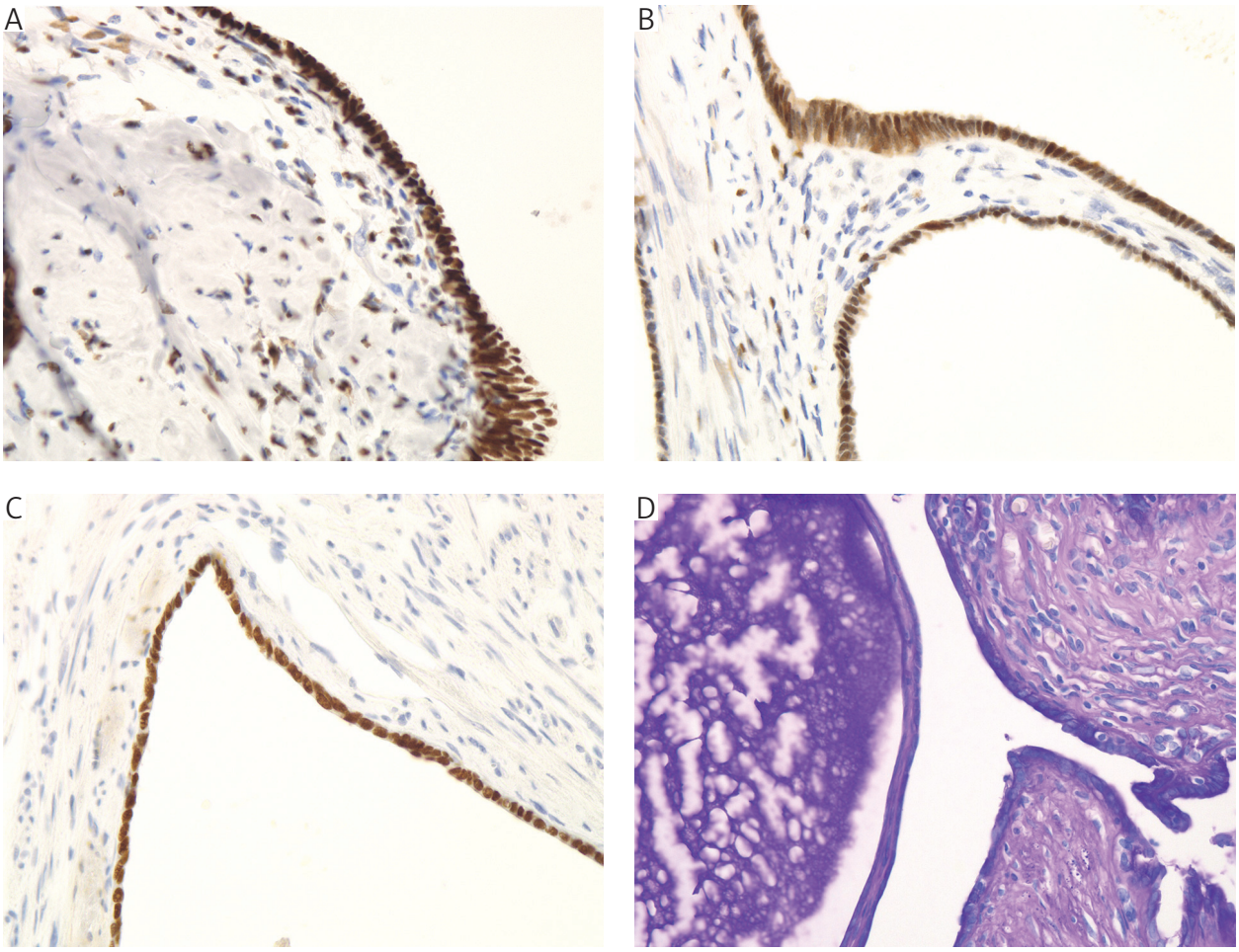

Fig. 2. A-C) Immunohistochemical analysis revealed that the epithelial cells were positive for ER (A), PgR (B) and PAX8 (C). D) Mucin was present in the apical cytoplasm and was retained within the glandular lumens

ciliated glandular epithelium that resembles the normal endosalpinx. The epithelial cells are typically positive for ER and PgR. Mucin is often present in the apical cytoplasm and within the glandular lumens. Florid cystic endosalpingiosis as designated by Clement and Young, can be differentiated from the typical ES by the fact that tumor-like masses are present with clear intrusion of the process into the subperitoneal connective tissue and the muscular walls of the uterus and other pelvic viscera [5]. Florid cystic endosalpingiosis is rare: only 18 cases have been reported in the literature to date.

The intramural type of FCE is mostly considered a leiomyoma with cystic degeneration or cystic adenomyosis [4]. Cystic ES is part of müllerianosis, disorders consisting of the heterotopic presence of müllerian-derived tissue [9]. Müllerianosis is a rare entity consisting of an organoid admixture of two or three types of glands of müllerian derivation (cervical, endometrial or tubal) in heterotopic locations.

Lucy *et al*. compared women with ES to those with endometriosis, particularly regarding chronic pelvic pain and infertility [10]. Endosalpingiosis differs from endometriosis in that it has ciliated glandular epithelium, no endometrial stroma and does not exhibit the same inflammatory response. Endosalpingiosis and endometriosis occur concurrently in 34% of ES cases [10], as in the present case. They are also both speculated to be involved the secondary müllerian system.

The pathogenesis of FCE is largely unknown. The distinction between an extraovarian serous cystadenoma and cystic endosalpingiosis is arbitrary [5]. Endosalpingiosis biomarker expression closely resembles that of serous neoplasms, thus strengthening the growing body of evidence that all Müllerian serous carcinomas arise from tubal-like epithelium. Traditionally, ES may develop via metaplasia of the peritoneum. In contrast, serous cystadenoma/adenofibroma is thought to develop from invaginations of ovarian surface epithelium and then undergo tubal metaplasia. However, it is now thought that ovarian low-grade serous tumors and their non-invasive implants, ovarian epithelial inclusion glands and ES, might arise from the fallopian tubes [11] it was thought that ovarian high-grade serous carcinoma arises from the ovarian

surface epithelium and epithelial inclusion glands and that the pathogenesis is *de novo*; nonetheless, a convincing precursor in the ovary or peritoneum has not been identified to date. During the last few years, however, there has been a dramatic shift in thinking, and a candidate precursor is now recognized in the fallopian tube, especially within the fimbriated end – serous tubal intra-epithelial carcinoma (STIC). Florid cystic endosalpingiosis shows that ES can involve the muscularis layers of the hollow pelvic viscera, resulting in clinically and macroscopically detectable masses and an infiltrative pattern on microscopic examination [5].

As mentioned previously, premenopausal women with ES are more likely to have gynecologic malignancy [10], and a case of adenocarcinoma arising in ES was reported [12]. One study showed that ES can be found in association with serous borderline tumors, and other investigators have observed chronic salpingitis in association with ES or serous borderline tumors [2]. Awareness of this lesion will facilitate the correct diagnosis by the pathologist. We reported our experience with a rare case of FCE.

The authors declare no conflict of interest.

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