

CASE REPORT

Case Report on Endometriosis Associated Peritoneal Endometrioid Carcinoma

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Abstract

Endometriosis-associated neoplasms will develop in 0.7–2.5% of women with endometriosis. We present a rare case report of endometriosis associated endometrioid carcinoma of peritoneum. We report a case of 43 years female with chief complaints of menorrhagia, dysmenorrhoea and dyspareunia. Her imaging revealed an endometriotic cyst of in left ovary along with iso to hyperechoic nodular area seen in right iliac fossa. Laparoscopic exploration validated it to be a case of grade IV endometriosis along with presence of peritoneal nodule of 5.5 × 4.5 cm in right iliac fossa. The peritoneal nodular lesion was removed with en bloc wide local excision with clinically disease-free margin. Histopathology report of peritoneal nodule shows endometrioid adenocarcinoma. Complete surgical staging for peritoneal cancer was performed and was not advised any adjuvant therapy. Patient is disease free since last 3 years.

Keywords Endometriosis · Endometriosis associated malignancy · Peritoneal endometrioid carcinoma

Abbreviations

POD Pouch of douglas

EAM Endometriosis associated malignancy

Introduction

Endometriosis is a chronic inflammatory, benign gynaecological disease. Endometriosis is defined by the presence of endometrial-like tissue outside the uterus. Three subtypes of endometriosis have been described: superficial peritoneal, ovarian (endometrioma or ‘chocolate cysts’) and deep infiltrating type. Endometriosis-associated neoplasms will develop in 0.7–2.5% of women with endometriosis. There is

well-documented evidence of ovarian endometriosis being precursor to ovarian carcinoma, commonly of endometrioid and clear cell variety. Endometriosis at extra-ovarian sites such as colon, fallopian tube, parametrium, uterosacral ligament, vagina, previous caesarean scar, pelvic sidewall, chest wall can also cause malignancy [1]. Sigmoid colon and rectum are the most common site of extra-ovarian endometriosis associated malignancies. We present a rare case report of endometriosis associated endometrioid carcinoma of the peritoneum and discuss its etiopathogenesis and treatment protocol.

Case History

We report a case of 43 years female, para 2 live 2, with BMI of 33 kg/m², who presented at the clinic with chief complaints of menorrhagia, dysmenorrhoea and dyspareunia since last 4–5 months. Her imaging (ultrasonography abdomen-pelvis) revealed an endometriotic cyst of 7.5 × 6 cm in left ovary adherent to POD along with a recto-vaginal nodule of 3.5 × 2 cm. The right ovary was observed to be normal. There was well defined 5.5 × 4.5 cm, iso to hyperechoic nodular area seen in right iliac fossa. Her serum tumour marker CA-125 was 250 IU/ml. Laparoscopic exploration validate it to be a case of grade IV endometriosis. A fiery red, cauliflower like peritoneal nodule of approximately

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5.5 × 4.5 cm was seen in right iliac fossa (Fig.1), separate from right ovary and fallopian tube. The patient underwent total laparoscopic hysterectomy with bilateral salpingo-oophorectomy. The peritoneal nodular lesion was removed with en bloc wide local excision with clinically disease-free margin. Histopathology report of peritoneal nodule shows

a 5.5 × 3.5 × 3 cm size nodule with endometrioid adenocarcinoma, moderately differentiated with an extensive area of haemorrhage with scattered endometriotic spots (Fig.2). Remaining all the specimen were negative for malignancy. As a part of complete surgical staging for peritoneal cancer, bilateral pelvic lymphadenectomy, para-aortic

Fig. 1 Endo-view showing fiery red cauliflower growth like peritoneal nodule with adjoining normal adnexa

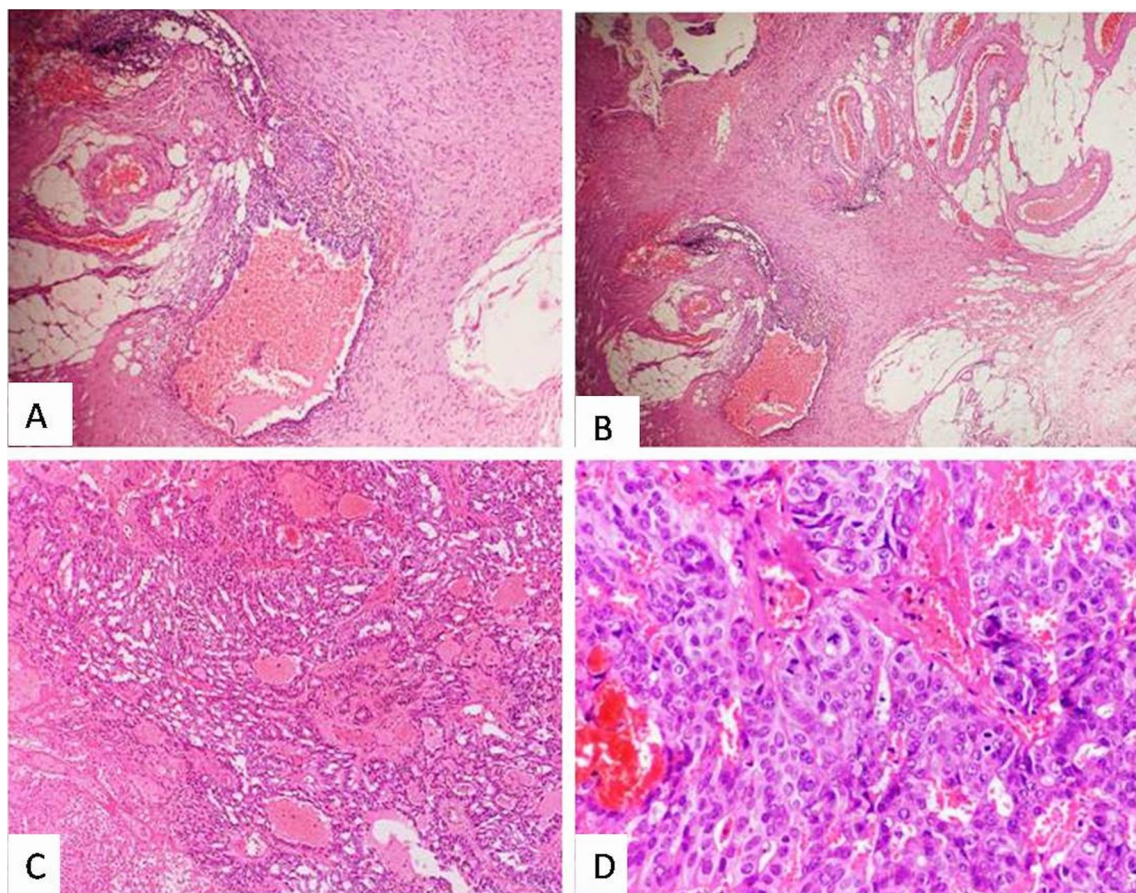
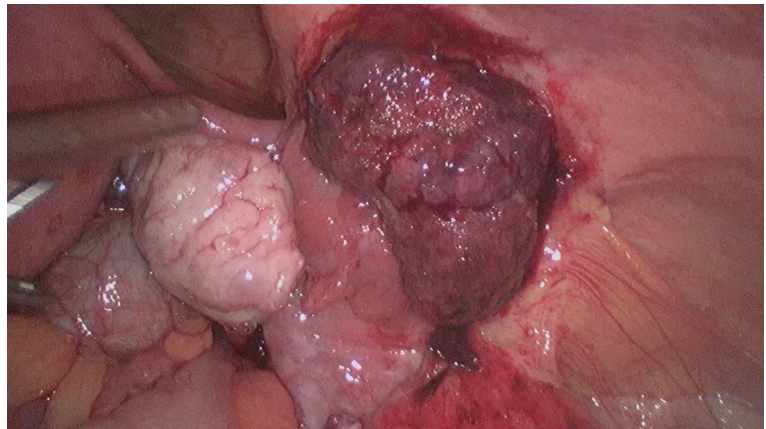


Fig. 2 Peritoneal endometrioid carcinoma associated with endometriosis. (A) and (B) Peritoneum showing focus of endometriosis (C) and (D) Peritoneal Endometrioid carcinoma

lymphadenectomy, infra-colic omentectomy, multiple peritoneal biopsies were performed. The final histopathology report was negative for malignancy in all the specimen. As per available literature on management of this rare case and after discussion in tumour board, the patient was not advised any adjuvant therapy and was kept on regular follow-up every 3 months with ultrasonography (abdomen-pelvis) and serum CA-125. The patient had been on regular follow-up since last 3 years and disease free to date.

Discussion

Primary peritoneal cancer is an idiopathic cancer arising from peritoneal layers of the abdominal cavity. The most common histological type of primary peritoneal cancer is serous carcinoma of the peritoneum which accounts for 10% of this malignancy occurring in the pelvis. Incidence of other histological subtypes such as endometrioid or clear cell variety of peritoneal cancer is rare and it is commonly associated with endometriosis. Sampson et al. in 1925, first established the specific criteria for defining cancers arising from endometriosis: According to the criteria there should be: (1) the presence of benign endometrial tissue and cancer in the same site, (2) the presence of endometrial stroma surrounding the glands, and (3) the exclusion of metastasis from another primary site. According to the criteria, the excised peritoneal nodule was diagnosed as primary peritoneal endometrioid carcinoma arising from peritoneal endometriosis. Two main hypotheses have been attempted to explain the origin of endometriosis-associated malignancies (EAM). EAM may arise by malignant transformation of endometriotic cells through intermediary lesions (atypical endometriosis) or presence of similar microenvironment of hyper-estrogenism, as provided through the activity of aromatase enzyme in endometriosis. Normally, aromatase enzyme is absent in eutopic endometrial tissue, but it is present at high levels in endometriotic tissue. This enzyme catalyses the conversion of androstenedione and testosterone into estrone and estradiol, respectively. In addition, estradiol stimulates cytokines as prostaglandin E2 production, which promotes tumour growth and triggers aromatase activity, resulting in a positive feedback loop in favour of continuous estrogen formation in endometriosis [2]. Zanetta et al. in 2000, in their study showed that in obese women, same mechanism of the peripheral conversion of steroid hormones into estrone by aromatase enzyme increases, leading to a hyper-estrogenic environment. Thus, we may hypothesise that the obesity-related hyper-estrogenism might have been a contributing factor for malignant transformation of endometriosis in our study (BMI 33 kg/m²).

The main management protocol for peritoneal carcinoma is surgical staging as in case of carcinoma ovary which

includes excision of localised lesion followed by multiple peritoneal biopsies, bilateral pelvic lymphadenectomy, para-aortic lymphadenectomy and omentectomy. As per final surgico-pathological stage of cancer, further adjuvant therapy as chemotherapy, radiotherapy or hormonal therapy will be planned.

Modesitt et al. in 2002, in a series of 21 cases of endometrioid peritoneal cancer, recommended same surgical staging of the disease. Same therapeutic approach was also followed in other studies by Marchand et al. (2013) and Poon et al. [3]. We proceeded with the same surgical treatment in our index case.

Role of chemotherapy or radiotherapy as adjuvant therapy in extra ovarian endometrioid cancer is not yet proven. In study by Poon et al. [3], it was concluded that if the primary disease was localised and excised without any residual tumour, adjuvant therapy can be omitted. Same treatment protocol was followed in a study by Marchand et al. (2013) and our study as well. In contrast to this, Oto et al. [4] in his study documented chemotherapy as adjuvant treatment. Benoit et al. in 2006 study, found that for patients with disease confined to the site of origin, 5-year survival rate has been 82–100% for endometrioid cell type. However, disseminated intraperitoneal disease, had a very guarded prognosis, with a 5-year survival rate of only 0–12%.

Owing to rarity of cases and absence of any standardised treatment protocol, treatment approach should be individualised. As per available literature, any suspicious lesion should be completely excised, followed by complete surgical staging. Role of adjuvant therapy needs to be validated. Further studies with a large sample size are required to substantiate this fact. The treatment protocol followed in our case corroborate with the management of these rare cases.

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Declarations

Conflict of interest The authors declare that they have no conflicts of interest and nothing to disclose.

Human and Animal Rights Research involving human participants and/or animals: It is a retrospective original case report.

Informed Consent Informed consent was obtained from individual participants included in the study.

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