CASE REPORT

Malignant transformation of superficial peritoneal endometriosis lesion

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SUMMARY

A 63-year-old woman with no medical history underwent an abdominal surgery with hysterectomy and bilateral salpingo-oophorectomy for a 10 cm peritoneal cyst with increased cancer antigene-125. A large suspicious tumour of the Douglas space, with contact to the uterus and the rectal wall was described. The rest of the exploration was normal, specially the rest of the peritoneum. Histopathology revealed a malignant transformation of a superficial peritoneal endometriosis. Secondary surgery was thus completed by laparoscopy with bilateral pelvic and para-aortic lymph node dissections, omentectomy and multiple peritoneal biopsies. All staging samples were free of cancer; therefore no complementary therapy was administered. After 18 months of follow-up, consisting of clinical examination and pelvis magnetic resonance imaging every 6 months, we did not observe any recurrence. Malignant transformation of superficial peritoneal endometriosis is a rare disease and surgical management seems to be the main treatment.

BACKGROUND

Endometriosis is a common and benign gynaecological disease. It is defined by the presence of endometrial glands and stroma outside the uterine cavity. Estimated prevalence is 5–15% in women of childbearing age and only 3–5% in postmenopausal women. The first case of malignant transformation was described in 1925. The author proposed three diagnostic criteria to establish the relationship between endometriosis and cancer: coexistence within the tumour of endometriosis and neoplastic tissue, histological type of cancer must be seen to arise in endometriosis (epithelial glands in an endometrial stroma) and, finally, absence of other possible origin of the carcinoma.[1] The malignant transformation of ovarian endometriosis is well known and the prevalence of endometriosis in patients with epithelial ovarian cancer, especially in endometrioid and clear cell types, has been confirmed to be higher than in the general population.[2–4] However, cases of extraovarian endometriosis associated to cancer are rare and therapeutic management is not standardised. One case of cancer developed on superficial peritoneal endometriosis lesion has recently been described in our department.

CASE PRESENTATION

The patient, 63 years, virgin, menopaused at the age of 56 years without hormonal therapy, with no personal and family medical history consulted for a pelvic discomfort and abdominal distension. The pelvic ultrasound found a cyst of 10×6 cm, poly-lobed, thin-walled, of undetermined origin. Pelvic magnetic resonance imaging (MRI) confirmed the presence of a 9 cm cystic mass on the right side of the uterus, with several vegetation tissues on T2 sequence and raising after gadolinium injection. The origin suspected was first the right ovary. An extension in Douglas space was described but no abdominal effusion or nodule of the carcinosis was seen in MRI. The thoracoabdominal CT and positron emission tomography scan were normal. Cancer antigene-125 (CA-125) (65 IU/mL) and CA 19–9 (61 IU/mL) were increased. The patient presented symptoms of ovarian cancer. Laparoscopic exploration was proposed. Abdominal exploration showed a normal uterus and normal ovaries. A large suspicious tumour of the Douglas space, with contact to the uterus and the rectal wall was described. The rest of the exploration was normal, specially the rest of the peritoneum. At the end of surgery (removal of the tumour, hysterectomy and bilateral salpingo-oophorectomy in a laparotomic way), there was no residual macroscopic tumour. Histopathological examination concluded to moderately differentiated endometrioid adenocarcinoma grade II in a formation corresponding to a cystic endometriosis lesion of the pouch of Douglas. Uterus, ovaries and peritoneal cytology were free of cancer.

TREATMENT

The multidisciplinary oncological meeting of our department and the 'Rare Tumors Paris Meeting' opted for a complementary surgery by laparoscopy consisting in omentectomy, peritoneal biopsies, bilateral pelvic and para-aortic lymph node dissections as in cytoreductive surgery for ovarian cancer. All of these samples were free of cancer, so we decided a simple follow-up without complementary treatment.

OUTCOME AND FOLLOW-UP

After 18 months of follow-up, consisting in a clinical examination and pelvis MRI every 6 month, we did not observe any recurrence.

DISCUSSION

The frequency of extraovarian endometriosis degeneration varies from 0.8% to 0.9%.[5 6] For Brunson, 90% of these transformations are endometrioid adenocarcinomas (supprimer: whereas ovarian forms are predominantly clear cells tumours).[7] Clinical signs are usually abdominal

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pain (51%), pelvic mass (25%), vaginal bleeding (18%), haematuria and rectal bleeding (2%). Carcinomatosis may also be revealing.[8] Diagnostis was performed only on histological analysis and radiological examination is not specific. There is no specific tumour marker. Therapeutic strategy is not consensual because of the rarity of the disease. Five recent cases of superficial degeneration of the Douglas reported in the literature related surgery with total hysterectomy, bilateral adnexectomy and resection of the mass with or without node staging and adjuvant chemotherapy.[9–11] In the case described by Chung, there was no indication for reoperation for staging because of a rapid and significant recurrence.[9] Modesitt[12], in a series of 21 cases of cancer developed on extraovarian endometriosis, showed that cancer extension is a survival predictive factor. He recommended staging of the disease consisted in cancer surgery without macroscopic residual tumour, pelvic and para-aortic lymph node dissections, omentectomy and multiple peritoneal biopsies.

Effectiveness of chemotherapy is unknown. On one side there are low benefits of chemotherapy in endometrial cancer, and on the other side peritoneal and ovarian cancers are chemosensitive. Regarding the role of radiotherapy, there is no recent data on this subject; only Heaps,[8] in 1990, proposed radiotherapy for pelvic confined forms. Prognosis of endometrioid tumours confined to the pelvis is good; Heaps reported a 5 years survival rate of almost 100% after surgery and radiotherapy, but in cases of peritoneal carcinomatosis, it would not exceed 10%, for a median survival of 3.5 years. Recently, a laparoscopic excision of a cystic retroperitoneal adenocarcinoma arising in a focus of extraovarian endometriosis was described without the need for any adjuvant chemotherapy or radiation therapy. The patient remains with no evidence of recurrent or metastatic disease after 3 years of follow-up.[13] In addition, a complex cyst in a postmenopausal woman with increased CA-125 must be removed with caution. In this case the cyst was removed without spillage. In the case of rupture, chemotherapy could have been indicated because of the peritoneal spread of the lesion, as in Federation of Gynecology and Obstetrics stage IIIc of ovarian cancer.

Learning points

- The malignant transformation of extragonadal endometriosis is extremely rare.
- The patient presents symptoms of having ovarian/primary peritoneal cancer.
- The surgical therapy is equal to ovarian cancer, an oncological surgery as complete as possible, without residual disease. It might consist in total hysterectomy, bilateral salpingo-oophorectomy, peritoneal cytology, omentectomy combined with a pelvic and para-aortic lymphadenectomy.
- It is not known if and when to give adjuvant chemotherapy, but should be discussed in case of lymph node incolvement.[14]
- The role of irradiation is probably limited.

Competing interests None.

Patient consent Obtained.

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REFERENCES

1 Sampson J. Endometrial carcinoma of the ovary, arising in endometrial tissue in that organ. *Arch Surg* 1925;10:1–72.
2 Nezhat F, Datta MS, Hanson V, *et al*. The relationship of endometriosis and ovarian malignancy: a review. *Fertil Steril* 2008;90:1559–70.
3 Pearce CL, Templeman C, Rossing MA, *et al*. Association between endometriosis and risk of histological subtypes of ovarian cancer: a pooled analysis of case-control studies. *Lancet Oncol* 2012;13:385–94.
4 Munksgaard PS, Blaakaer J. The association between endometriosis and ovarian cancer: a review of histological, genetic and molecular alterations. *Gynecol Oncol* 2012;124:164–9.
5 Mostoufizadeh M, Sculli R. Malignant tumors arising in endometriosis. *Clin Obstet Gynecol* 1980;23:951–63.
6 Stern RC, Dash R, Bentley RC, *et al*. Malignancy in endometriosis: frequency and comparison of ovarian and extraovarian types. *Int J Gynecol Pathol* 2001;20:133–9.
7 Brunson GL, Barclay DL, Sanders M, *et al*. Malignant extraovarian endometriosis: two case reports and review of the literature. *Gynecol Oncol* 1988;30:123–30.
8 Heaps JM, Nieberg RK, Berek JS. Malignant neoplasms arising in endometriosis. *Obstet Gynecol* 1990;75:1023–8.
9 Chung Fat B, Terzibachian JJ, Govyadovskiy A, *et al*. Postmenopausal malignant transformation of extragenital endometriosis. A case report. *Obstet Gynecol Fertil* 2008;36:51–5.
10 Benoit L, Arnoult L, Margarot A, *et al*. Malignant tumours arising in extraovarian endometriosis: three cases reports and review of the literature. *Ann Surg* 2004;129:376–80.
11 Paillocher N, Pessaux P, Catala L, *et al*. Malignant tumors arising in extra-ovarian endometriosis: a case report. *J Gynecol Obstet Biol Reprod* 2005;34:501–3.
12 Modesitt SC, Tortolero-Luna G, Robinson JB, *et al*. Ovarian and extraovarian endometriosis-associated cancer. *Obstet Gynecol* 2002;100:788–95.
13 Beiko D, Bryson SC, Bell CD, *et al*. Laparoscopic excision of a cystic retroperitoneal adenocarcinoma arising in a focus of extra-ovarian endometriosis. *Can Urol Assoc J* 2010;4:E71–3.
14 Benoit L, Arnould L, Cheynel N, *et al*. Malignant extraovarian endometriosis: a review. *Eur J Surg Oncol* 2006;32:6–11.