Case Reports

Adenosarcoma of the ovary. A case report

P. Litta, C. Pozzan, G. Sacco, F. Merlin, C. Saccardi, A. Ambrosini

Department of Gynecological Sciences and Human Reproduction, University of Padova (Italy)

Summary

Adenosarcoma of the ovary is a rare condition. We report a case of a 32-year-old patient that has been treated in our Department. The diagnosis of ovarian adenosarcoma was carried out after laparoscopy with removal of an ovarian endometriotic cyst. Laparoscopic homolateral ovariectomy was then performed and conservative treatment was decided on considering the young age, low stage and low grade of the disease. Second-look laparoscopy, clinical evaluation and ultrasound were performed for four years of follow-up. No recurrence has been detected. Conservative treatment should be proposed in fertile age with low-grade ovarian adenosarcoma, but a strict follow-up is always necessary.

Key words: Ovarian adenosarcoma; Laparoscopy.

Introduction

Adenosarcoma of the ovary is a rare extrauterine tumor histologically similar to adenosarcoma of the uterus. This kind of tumor was first described in the uterus in 1974 by Clement and Scully, who defined it as "a malignant, usually low-grade, stromal component and a generally benign, but occasionally atypical, glandular epithelial component" [1].

In the literature 57 cases of ovarian adenosarcoma have been reported since 1967. In 2002, 40 of these cases were reviewed exhaustively by Clement and Scully [2]. According to the authors [2, 3], microscopic features of ovarian mesodermal adenosarcoma are considered: conspicuous non-invasive mullerian-type glands within a predominant malignant stromal component, either homologous or heterologous; periglandular cuffs of cellular stroma, intraglandular protrusions of cellular stroma, or both; usually at least mild stromal atypia; variable stromal mitotic count but may be very low [2].

Case Report

A 32-year-old nulliparous woman presented at the end of 1999 to our Department of Gynecologic Sciences and Human Reproduction, University of Padova, Italy. She had a history of dysmenorrhea since menarche, which was particularly intense in the last months. She had been taking oral contraceptives since 1990 with annual suspensions of two months. In April 1998 she had a negative pelvic ultrasound. In October 1999 an abdominal ultrasound revealed a left-sided ovarian cyst, 63 x 50 mm in diameter, that was likely to be of endometriotic origin. In January 2000 the patient underwent operative laparoscopy, with removal of bilateral ovarian cysts: on the right side there was a 1 cm-deep endometrioma and on the left side (ovary 6 cm in diameter) multiple endometriotic cysts (Figure 1). Cysts were removed by stripping and then coagulation of the ovarian bed was performed. In particular, in the deepest part of the left cyst friable brown tissue was accurately removed. Cromosalpingoscopy was performed revealing bilateral tubal patency. Endometrial biopsy was performed.

Histological examination revealed a partially benign endometrial adenofibroma with wide areas of proliferative activity of endometrial stroma, with a polypoid intraglandular and periglandular growth both expansive and infiltrating. No atypia were detected in the stromal component. Indefinite areas of smooth muscle and fibrous tissue were detected. It was not possible to determine if this neoformation was intraovarian or if it extended outside the ovary. Definitive diagnosis was an endometrial mixed neoplasia without glandular atypia and with low-grade proliferation of the stromal component - adenosarcoma with low-grade malignancy, that arose from ovarian endometriosis. Subatrophic endometrium was detected. Immunohistochemical evaluation was performed. Proliferative activity was detected at MIB-1 and topoisomerase was confined to the stromal component. Estrogen and progesterone receptors were both positive in the stroma; p53 was negative.

In November 2000, second-look laparoscopy was performed with left ovariectomy and peritoneal washing. Abdominal inspection was completely negative. In the removed ovary a small endometriotic focus was detected without atypia or hyperplasia; peritoneal washing was negative.

The patient was followed-up with a clinical examination and transvaginal ultrasound every six months and in November 2001 she underwent a third laparoscopy: removal of a 1.5 cm endometriotic right ovarian cyst, peritoneal biopsies and washing were performed. Histologic examination was negative.

Every six months the patient was evaluated by abdominal-transvaginal ultrasound.

In January 2004 (4 years of follow-up) the patient was free of disease at clinical examination and transvaginal ultrasound. She still complains of mild chronic pelvic pain. She has never stopped taking oral contraceptives (etinilestradiol 30 μg, gestodene 75 μg).

Discussion

Considering the literature, ovarian adenosarcoma is a rare neoplasia that is still a matter of debate. What is eye-catching in previously reported cases is the heterogeneity of the clinical behaviour of this tumour, and that guide-

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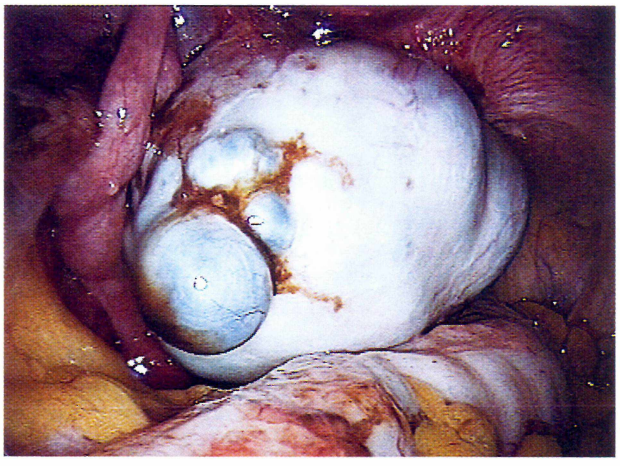

Figure 1. — Ovarian adenosarcoma associated with multiple endometriotic cysts in the left ovary (6 cm in diameter).

lines about its treatment are not yet definable, especially in fertile age.

Prognostic variables have been analysed with the aim of detecting those patients at risk for recurrence of disease after surgical treatment. It is clear that higher recurrence rates are associated with high grade tumours and those with extraovarian spread. However it is important to underline that also Stage I and low-grade tumours recur with an incidence of 63% and 73%, respectively.

Women are affected by this tumour especially in their fifth and sixth decade of age (57.5%) [2]. Age is an important prognostic factor: women younger than 53 years are more likely to have recurrence than postmenopausal women. Recently however a very aggressive form of adenosarcoma of the ovary has been reported in a 77-year-old woman [6]. This aspect makes this form of adenosarcoma different from its uterine counterpart, which has a peak age incidence in the eighth decade. Prognosis appears different, too: death from tumour is 37.5% in the ovarian form compared to 16% in uterine adenosarcoma [3-5].

Our case report could confirm the link between endometriosis and ovarian adenosarcoma [7, 8]. Our patient was affected by endometriosis in both ovaries at the time of the first operation. An endometriotic nodule was also found in the ovary that was removed with the second laparoscopy and another endometrioma was found in the third intervention.

Surgical treatment is certainly the first necessary step in this kind of neoplasia. Our case reports the use of laparoscopy to treat this kind of tumour. Considering the prevalent unilaterality, young patient age and the possibility of a future pregnancy, it was decided with the patient's consent to practise a conservative surgical approach. A strict follow-up has then been performed to detect every possible recurrence. Considering the literature, the reported interval to the first recurrence from the time of staging of tumour is 0.4-6.6 years (mean 2.6 years). In our case report the follow-up covers four years with no relapse of disease, but with persistence of endometriosis.

Conclusion

Our report describes a case of an ovarian adenosarcoma that has been treated conservatively by laparoscopy.

If ovarian adenosarcoma is diagnosed after an accurate and radical removal of an ovarian cyst, it is correct to program a second-look laparoscopy after six months to restage the tumour. With the aim of reducing the risk of a local recurrence, removal of the residual ovary could be recommended. This therapeutical strategy should be considered in selected women in fertile age with low-stage and low-grade tumours. A strict follow-up is always necessary. Clinical evaluation together with ultrasound must be performed every six months for at least five years.

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Address reprint requests to:
P. LITTA, PH.D.
Department of Gynecology Sciences
and Human Reproduction
University of Padova
Via Giustiniani, 3
35128 Padova (Italy)