Mayer-Rokitansky-Küster-Hauser syndrome and ovarian cancer

Report of a case

G. GHIRARDINI (*) - A. MAGNANI (**)

Summary: A case of ovarian cancer associated with the Mayer-Rokitansky-Küster-Hauser syndrome is reported. As far as we know, this is the second report of ovarian neoplasm associated with this syndrome. The need for gynecological control of ovarian and other possible pathologies of the genital tract in these women is discussed.

Key words: Genital anomalies; Vaginal agenesis; Mayer-Rokitansky-Küster-Hauser syndrome; Ovarian cancer.

INTRODUCTION

The Mayer-Rokitansky-Küster-Hauser syndrome (MRKHs) is a relatively rare developmental anomaly of the female genital system, occurring in about 1/5000 female births and characterized by absense of the vagina, absent uterus or rudimentary uterine horns with normal ovaries ([1]). Since the first description in 1559 ([2]) the attention of the Authors has been devoted to the different clinical pictures of the syndrome and to the reconstruction of a functional vagina, to allow normal sexual life to these women ([3]).

Different genital pathologies associated with this syndrome have only recently been described ([4, 5, 6, 7]).

Here we report a new case of ovarian cancer associated with the MRKHs. The need for normal gynecological controls in these women is discussed.

CASE REPORT

G. L., a 68 year old woman came to our observation in January 1993, with a history of increasing abdominal girth and pain for some months.

She was unmarried, had never menstruated in her life, nor had sexual intercourse. Past medical history revealed only hepatitis B in 1986 and transitory cerebral ischemia in 1989 without consequences. She was not taking any drugs.

The physical appearance was of a normal female, with normal secondary sex characteristics. Hymen and vagina were absent. There was only a dimple of 2-3 cm. The lower abdomen was fully occupied by a firm mass. Transabdominal sonography revealed a solid mass of more than 10 cm in diameter. The mass appeared to be ovarian, although it impossible to distinguish the site of origin. Neither uterus nor uterine rudiments were identified. Pielography was normal. CA 125 determination was slightly elevated: 63 U/ml (normal value: $\leq$ 35 U/ml).

Received 13-10-1994 from the
(*) Divisione di Ostetricia e Ginecologia
(**) Divisione di Medicina,
Ospedale Franchini
Montecchio Emilia (RE) (Italy)
Revised manuscript accepted for publication 13-12-1994.

An exploratory laparotomy was performed. The mass was irregular, firm, with wide necrotic areas. Because of firm adhesion to the pelvic wall and the bowel only biopsies of the neoplastic tissue were performed.

The histological examination revealed a poorly differentiated ovarian tumour, classified as FIGO III, G 3.

In February 1993 chemotherapy with cisplatin and cyclophosphamide was started and continued for 6 consecutive cycles until July 1993. At this time CT revealed only a residual mass of 4 cm in diameter in the right adnexal region.

In August 1993 a second surgical exploration was performed. The uterus was absent. The tubes were both of normal appearance, as was the left ovary. The right ovary was transformed into a mass of about 4-5 cm in diameter.

Secondary pelvic or abdominal lesions were not identified. Bilateral adnexectomy and omentectomy were performed.

The histological examination revealed a normal left ovary, a normal omentum, whereas the right ovarian mass was classified as poorly differentiated ovarian tumour.

The patient thereafter underwent 3 other cycles of adjuvant chemotherapy with cisplatin and epirubicin, the chemotherapy ended in January 1994.

In March and August a complete restaging was made. The patient is now well without relapse of the disease.

DISCUSSION

Since the first descriptions at autopsy, considering these patients as a matter of curiosity, the MRKHs have gained increasing attention during the last 150 years ([8]). This interest is continuing, both for minimal invasive therapy in the reconstruction of a functional vagina ([3]) and for the understanding of the pathogenesis of all Müllerian anomalies ([9]).

However this interest is almost always limited to the sexual problems of these patients; only a few recent reports have described common gynecological pathologies associated with this syndrome ([4, 5, 6, 7]). With the report of this new case we stress the need for checking these patients like other normal women for possible gynecological diseases. The descriptions of endometriosis, myomas and ovarian cancer in patients with MRKHs are a demonstration of possible development of different diseases of the female genital tract.

In conclusion, together with the interest in the resolution of vaginal agenesis from the first approach to the exploration of the possibilities of pathological development should be born in mind and the patients advised to undergo routine gynecological checking during their lifetime.

REFERENCES

1) Ghirardini G., Segre A.: "Vaginal agenesis (Mayer-Roktansky-Küster-Hauser syndrome): recent etiopathogenetical and anatomical views". *Clin. Exp. Obst. Gyn.*, 1982, *9*, 98.
2) Ghirardini G.: "First description of vaginal agenesis". *Am. J. Obst. Gyn.*, 1979, *133*, 132.
3) Ghirardini G., Popp L. W.: "New approach to the Mayer-von Rokitansky-Küster-Hauser syndrome". *Adolesc. Pediatr. Gyn.*, 1994, *7*, 41.
4) Beecham C. T., Skiendzielewski J.: "Myoma in association with Mayer-Rokitansky-Küster syndrome". *Am. J. Obst. Gyn.*, 1977, *129*, 346.
5) Rosenfeld D. L., Lecher B. D.: "Endometriosis in a patient with Rokitansky-Küster-Hauser syndrome". *Am. J. Obst. Gyn.*, 1981, *135*, 105.
6) Acien P., Loret M., Chehab H.: "Endometriosis in a patient with Rokitansky-Küster-Hauser syndrome". *Gyn. Obst. Invest.*, 1988, *25*, 70.
7) Koonings P. P., Marayati L., Schlaert J. B., Lobo R. A.: "Mayer-Rokitansky-Küster-Hauser syndrome associated with endodermal sinus tumor of the ovary". *Fertil. Steril.*, 1991, *56*, 577.
8) Ghirardini G., Popp L. W.: "The Mayer-von Rokitansky-Küster-Hauser syndrome (uterus bipartitus solidus cum vagina solida): the development of gynecology through the history of a name". *Clin. Exp. Obst. Gyn.*, 1995, XXII, *1*, 87.
9) Ghirardini G., Tridenti G., Vadora E., La Sala G. B.: "Human reproduction and a D.D.D. (Determination, Differentiation, Development) pathogenetical classification of genital anomalies in phenotypic females". *Acta Europ. Fertil.*, 1990, *21*, 257.

Address reprint requests to:
G. GHIRARDINI
Divisione di Ostetricia e Ginecologia
Ospedale Franchini
42027 Montecchio Emilia (RE) (Italy)