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ORIGINAL ARTICLE

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Large endometrioma in an adolescent girl with Mayer-Rokitansky-Küster-Hauser syndrome

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Abstract A 14-year-old 46 XX female presented with primary amenorrhea. A normal vagina ending in a blind pouch was found at gynecological examination. Diagnostic laparoscopy revealed the absence of a uterus with rudimentary fallopian tubes, round ligaments and uterosacral ligaments. The left ovary contained a 7-cm chocolate cyst, which was shown to be an endometrioma by pathological examination. This rare occurrence of ovarian endometrioma coexisting with Mayer-Rokitansky-Küster-Hauser syndrome in an adolescent patient might be secondary to Müllerian-directed metaplasia in the ovaries.

Keywords Endometrioma · Endometriosis · Mayer-Rokitansky-Küster-Hauser syndrome · Rokitansky syndrome · Müllerian agenesis

Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome probably develops secondary to a defect or retardation of the Müllerian ducts. In this condition, a superior part of the vagina, uterus and fallopian tubes are either hypoplastic or aplastic. However, ovaries are usually normal [1].

Retrograde menstruation and coelemic metaplasia are two proposed mechanisms in the etiology of endometriosis. We report a case of MRKH syndrome associated with severe ovarian endometriosis in the adolescent period.

Case report

A 14-year-old phenotypic female was referred because of primary amenorrhea. She did not have significant health problems in childhood and was normal intellectually. There was no family history of consanguinity, miscarriage, neonatal deaths or other members with primary amenorrhea. On physical examination, no abnormalities were detected. Her height was 155 cm (30p), weight 53 kg (50p) and blood pressure 100/80 mmHg. Thyroid, lung, heart and abdominal examinations were normal. No evidence of facial dysmorphism, webbing of the neck or any skeletal abnormality was present. The secondary sexual characteristics assessment showed axillary hair stage 3, breast stage 3 and pubic hair stage 4 according to Tanner. A normal vagina ending in a blind pouch was found at the gynecologic examination. The blood count, standard blood chemistry parameters and urinalysis were within normal limits. Other laboratory findings including the hormone profile are given in Table 1. All of the tests were normal except a moderate elevation of CA 125. Chromosome analysis revealed a 46,XX karyotype. Transabdominal sonogram showed a 68-mm complex ovarian mass on the left side, and no uterus was found.

An exploratory laparoscopy revealed the absence of a uterus with only remnants of rudimentary fallopian tubes, round ligaments and uterosacral ligaments. The right ovary was normal. The left ovary contained a chocolate cyst that was about 7 cm in diameter. Laparoscopic ovarian cystectomy by stripping the cyst wall off the ovary was performed. The pathology report described endometrial stroma and glands along with hemosiderin laden macrophages.

Discussion

A process of retrograde menstruation may have an important role in the establishment of endometriosis. Obstructive müllerian anomalies are probably associated

Table 1 Laboratory data

Parameter	Value (Range)
Estradiol (pg/ml)	55.3 (28-172)
Follicle-stimulating hormone (mIU/ml)	3.8 (2.5-10.2)
Luteinizing hormone (mIU/ml)	4.6 (1.9-12.5)
Prolactin (ng/ml)	11.0 (2.1-17.7)
Total Testosterone (ng/dl)	52.7 (14-76)
Free testosterone (pg/ml)	0.91 (0.45-3.17)
Dehydroepiandrosterone sulfate (μg/dl)	137 (10-333)
Thyroid stimulating hormone (mIU/ml)	0.9 (0.35-5.5)
Free triiodoyhronine (pg/ml)	3.7 (2.3-4.2)
Free thyroxine (ng/ml)	1.2 (0.9-1.8)
Cortisol (μg/dl)	11.2 (4.3-22.4)
CA 125 (U/ml)	63 (<35)

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more with endometriosis as compared with nonobstructive anomalies. A functioning endometrium, patent tubes and outflow obstruction are significantly associated with endometriosis [2]. However, the retrograde flow theory cannot explain the finding of endometriosis in which the metastasis of normal endometrium could not occur or is highly unlikely [3]. In our patient, menstruation was probably not possible due to the hypoplastic uterus and tubes. The rudimentary tubes were not hydropic, and no evidence of accumulated menstrual blood was present.

Many reports of endometriosis in adolescents are present [4]. However, ovarian endometrioma in association with Rokitansky syndrome has not been widely reported. We believe that this rare occurrence may be secondary to müllerian-directed metaplasia in the ovaries.

The coelemic epithelium is derived from the underlying mesenchyme. Müllerian (paramesonephric) ducts are formed from the coelemic epithelium that invaginates at several points. The superficial stroma of the ovary is also composed of the mesenchyme and the coelemic epithelium [1]. This embryonic kinship may explain the coexistence of an ovarian endometrioma and müllerian agenesis in our patient. Additional factors such as sensitivity to estrogens and the occurrence of ovulation may also be considered in the pathogenesis of endometriomas. Induction of müllerian-directed metaplasia by an increase in estrogens following puberty (as in our patient) could be possible. The beginning of ovulation might also trigger the metaplasia to end with endometrioma formation.

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