Communications in Brief

Multiple Fibroids in a Postmenopausal Woman with Mayer Rokitansky Kuster Hauser Syndrome

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Introduction

The Mayer Rokitansky Kuster Hauser (MRKH) syndrome is a congenital abnormality of the genital tract which occurs in approximately 1 in 5000 women. It is characterized by a maldevelopment of the Mullerian ducts at their lower portion, which results in congenital absence of the vagina and either the absence of uterine tissue or the presence of two uterine anlage, which are nonfunctioning myometrial tissue lying laterally on the pelvic side walls. Finally, a small redundant piece of uterine tissue may lie in the midline, but it is again nonfunctional. Leiomyomata are common occurrences in women with normal uteri but the occurrence of fibroids in MRKH syndrome is a rarely reported finding. There are only 15 reported cases in the existing medical literature.1–15 This case is unique in that it occurred in a postmenopausal woman and this has never been previously described.

Case Report

Mrs. E.C., a 70-yr-old caucasian woman, presented with a history of three years of increasing abdominal swelling and frequency of micturition, which occurred every 30 to 60 minutes. She suffered also with nocturia, which occurred twice nightly. She had been previously diagnosed with MRKH syndrome in 1948 and had a McIndoe Read split-thickness skin graft vaginoplasty performed in 1952. Sexual function had been entirely normal ever since, and she was happily married and had been for some 45 years. In 1991, in consultation with her general practitioner, she decided she would like to take hormone replacement therapy as she had been suffering with menopausal symptoms for some years and was concerned about osteoporosis. She was therefore started on Prempak-C (Wyeth, Berks, United Kingdom) 1.25 mg daily, which she had remained on for the preceding 10 yrs prior to presentation.

Clinical examination revealed a palpable mass suprapubically, and bimanual examination revealed a mobile, smooth mass arising primarily on the right side of the pelvis. An ultrasound scan of the pelvis revealed a 10-cm-diameter mass with appearances consistent with a fibroid (see Fig. 1). In view of the pressure symptoms on her bladder, it was agreed that the fibroids should be removed. At the time of laparotomy a 10-cm fibroid mass was found arising from the right uterine anlage but several smaller fibroids were seen in the left anlage. Both anlage and fibroids were removed uneventfully (see Fig. 2). The histology confirmed the diagnosis of multiple leiomyomata.

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Fig. 1. Vaginal ultrasound of fibroids.
Discussion

This is the first case ever described of a woman who has developed fibroids in MRKH syndrome in the postmenopausal period, presumably secondary to taking hormone replacement therapy. The etiology of the fibroids remains to be determined. There does, however, exist a cultural incidence, and therefore a genetic basis, for the etiology. It is clear that fibroids are dependent on estrogen in order to grow and, postmenopausally, it is a well-recognized phenomenon that they regress in size with the absence of estrogen. The exposure of postmenopausal women with existing fibroids to hormone replacement therapy may induce fibroids to be reactivated and increase in size, giving rise to further problems. This case of MRKH syndrome illustrates that the myometrium, even in this congenital absence state, may develop fibroids under the stimulation of postmenopausal estrogen.

References