CASE REPORT

Anomalous vasculature in Mayer-Rokitansky-Kuster-Hauser syndrome

Amelia P. Bailey, M.D.,[b] *Jennifer L. Young, M.D., M.P.H.,*[c] *Elisabeth Nigrini, M.D.,*[d] *and Susan C. Modesitt, M.D.*[a]

[a]Division of Gynecologic Oncology, [b]Department of Obstetrics & Gynecology, University of Virginia Health System, Charlottesville, Virginia; [c]Division of Gynecologic Oncology, Department of Obstetrics & Gynecology, Medical University of South Carolina, Charleston, South Carolina; and [d]Department of Obstetrics and Gynecology, Georgetown University, Washington, D.C.

Objective: To report a case of unique anomalous pelvic vasculature in a woman with Müllerian agenesis.
Design: Case report.
Setting: University hospital.
Patient(s): A 61-year-old woman with Müllerian agenesis and stage IIIc papillary serous ovarian cancer.
Intervention(s): Surgery was performed, including bilateral salpingo-oophorectomy, omentectomy, right terminal ileum and right ascending colon resection, ileoascending/transverse colon reanastomosis, and debulking of pelvic plaques along bilateral ureters. The patient had subsequent chemotherapy with taxane and platinum agents and at the time of writing was in remission.
Main Outcome Measure(s): None.
Result(s): Absence of the uterus was confirmed; rudimentary uterine horns and associated fallopian tubes and ovaries were noted bilaterally. The right ureter coursed below and behind the right common iliac artery, which did not bifurcate into external and internal arteries at the pelvic brim. Instead there was only an external iliac artery that gave off a pelvic branch just before the inguinal ligament. On the left, there was only an internal iliac artery that gave off an external branch after diving into the pelvis.
Conclusion(s): It is possible that the aberrant vasculature was partially responsible for the absence of the uterus. Preoperatively, it is important to recognize the possibility of abnormal pelvic vasculature in patients with Müllerian agenesis. (Fertil Steril® 2010;94:350.e1–e4. ©2010 by American Society for Reproductive Medicine.)

Key Words: Müllerian agenesis, Mayer-Rokitansky-Kuster-Hauser syndrome, anomalous vasculature, ovarian cancer, pelvis, aberrant vessel

Müllerian agenesis, also known as Mayer-Rokitansky-Kuster-Hauser syndrome (MRKHS), is a developmental defect characterized by the complete or partial absence of the uterus and vagina with normal female external genitalia and a 46,XX karyotype. Rudimentary uterine horns and fallopian tubes are usually present, and, because they are derived from a separate embryologic system, the mesonephros, the ovaries are unaffected and function properly. This disorder is seen in 1 per 4,000–10,000 newborn girls but is usually undiagnosed until the patient presents with primary amenorrhea (1). Müllerian agenesis can occur in conjunction with abnormalities in multiple organ systems, including the urinary tract and cardiovascular system. We herein present a case in which anomalous pelvic vasculature was found intraoperatively in a patient known to have Müllerian agenesis, although these abnormalities were not identified on a preoperative computerized tomography (CT) scan. The ramifications of this finding in our patient and in MRKHS women in general will be discussed.

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Reprint requests: Amelia P. Bailey, M.D., Department of Obstetrics & Gynecology, University of Virginia Health System, P.O. Box 800712, Charlottesville, VA 22903 (FAX: 434-924-9180; E-mail: ameliabailey@virginia.edu).

CASE REPORT

A 61-year-old nulligravid female had a colonoscopy which revealed a cecal sessile tubovillous adenoma. Biopsy of this lesion was returned as metastatic Müllerian serous carcinoma, for which she was referred to the gynecologic oncology department. The patient endorsed primary amenorrhea and reported being told in her teens that only parts of her uterus and ovaries were found during an exploratory laparotomy. However, a CT scan performed at an outside hospital after her cancer diagnosis showed absence of the uterus and an 8.1 × 7 cm left adnexal mass with metastatic disease on the colon. Physical examination revealed normal external female genitalia and a foreshortened vagina, which was pink with atrophic changes and approximately 4 cm in length; there was no cervix noted. A uterus could not be palpated during bimanual exam, although an adnexal mass was appreciated. The remainder of her exam was normal.

The patient was taken to the operating room for removal of the pelvic mass, staging, and debulking. The procedures performed included bilateral removal of ovaries and rudimentary bicornuate cords with associated fallopian tubes, omentectomy, right terminal

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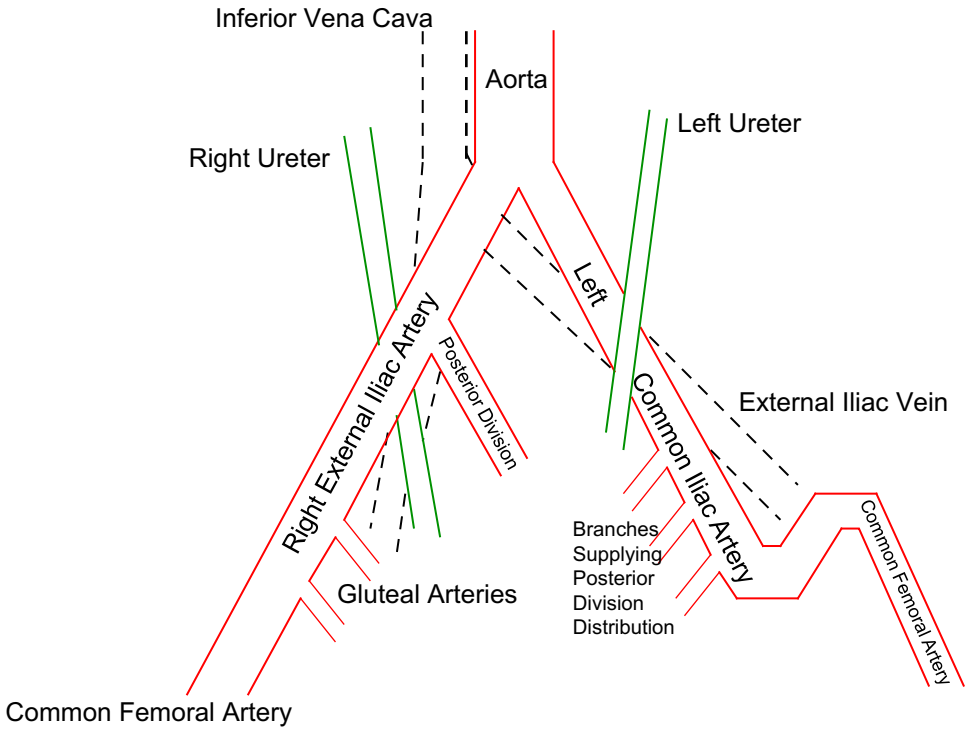
FIGURE 1

Line drawing of abnormal vasculature.



Bailey. Anomalous vasculature in MRKH syndrome. Fertil Steril 2010.

ileum and right ascending colon resection, ileoascending/transverse colon reanastomosis, and debulking of pelvic plaques along bilateral ureters. In this surgery, the absence of the uterus was confirmed and the following abnormal vasculature/anatomic relationships were noted (see Figs. 1–3).

Abnormally high aortic bifurcation, just inferior to the renal arteries.

Absence of bifurcation of the left common iliac artery; furthermore, the left common iliac vein traversed over the artery. An internal iliac artery was observed medial and inferior to the external iliac vein and was noted to dive deeply before giving off a femoral artery branch that crossed the external iliac vein and exited the pelvis under the inguinal ligament. The internal iliac artery then supplied the posterior division distribution.

The left ureter crossed appropriately over the ipsilateral internal iliac artery.

The right ureter coursed beside the ipsilateral external iliac artery and then crossed beneath it at the pelvic brim to enter the pelvis.

Absence of the bifurcation of the right common iliac artery at the pelvic brim, with only a right external iliac artery that coursed lateral and superior to the external iliac vein. This artery gave a pelvic branch just proximal to the inguinal ligament that crossed inferiorly over the external iliac vein to enter the pelvis.

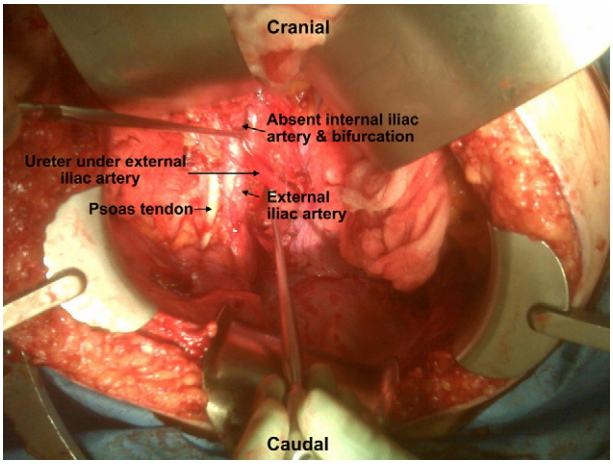
The patient had an uneventful postoperative course and was discharged home on postoperative day 3. The pathology revealed metastatic papillary serous ovarian carcinoma. The patient subsequently received six cycles of adjuvant chemotherapy with intravenous carboplatin and taxanes and at the time of writing was in remission.

DISCUSSION

The MRKH syndrome is a constellation of abnormalities that can include multiple organ systems, including the vascular tree. In the present case, regarding the vasculature, we saw absence of the internal iliac artery on the right and absence of the external iliac artery on the

FIGURE 2

Intraoperative picture of abnormal vasculature.



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