

“Mayer Rokitansky-Kuster-Hauser” syndrome with adenomyosis in rudimentary horn: a case report

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Background

Mayer-Rokitansky-Kuster-Hauser syndrome is a rare congenital malformation that affects about 1:4500 female newborns. This syndrome is characterized by complete or partial vaginal agenesis, tubal and uterine-cervical abnormalities. The rudimentary uterus present in MKRH may still have functioning endometrium tissue.

Rarely, MRKH has a rudimentary horn and adenomyosis in a

Case report:

A 31-year-old female went to Vietnam National Hospital of Obstetrics and Gynecology with primary amenorrhoea. She complained about cyclic low abdominal pain for 2 years, for which she had previously experienced multiple emergency room visits but was misdiagnosed. Transabdominal and transvaginal ultrasonography showed aplasia of the uterus and cervix. Ultrasound findings have shown the presence of two distinct rudimentary uterine horns with functional endometrium and adenomyosis. Ovaries are malpositioned, and there is an endometrioma in each ovary. The kidneys were normal in size and position. Further evaluation was advised. Magnetic resonance imaging findings (MRI) confirmed the diagnosis of MRKH and adenomyosis in the rudimentary horn and endometriosis in both ovaries

Discussion:

MRKH syndrome is a rare disorder described as aplasia of the uterus, cervix, and upper two-thirds of the vagina due to early arrest in the development of the Mullerian duct. Adenomyosis in the rudimentary horn of MRKH syndrome has been reported in the literature; however, in most instances, the proper diagnosis is missed. Patients are either misdiagnosed with endometriosis, ovarian masses, or ovarian fibroma.

Conclusion:

This was a rare case of MRKH with functioning endometrium in the remnant horn. Non-invasive imaging modalities, such as ultrasound, CT, and MRI, are the first choices for the diagnosis of pelvic tumors. Experienced and skilled imaging physicians are critical in diagnosing MRKH syndrome.

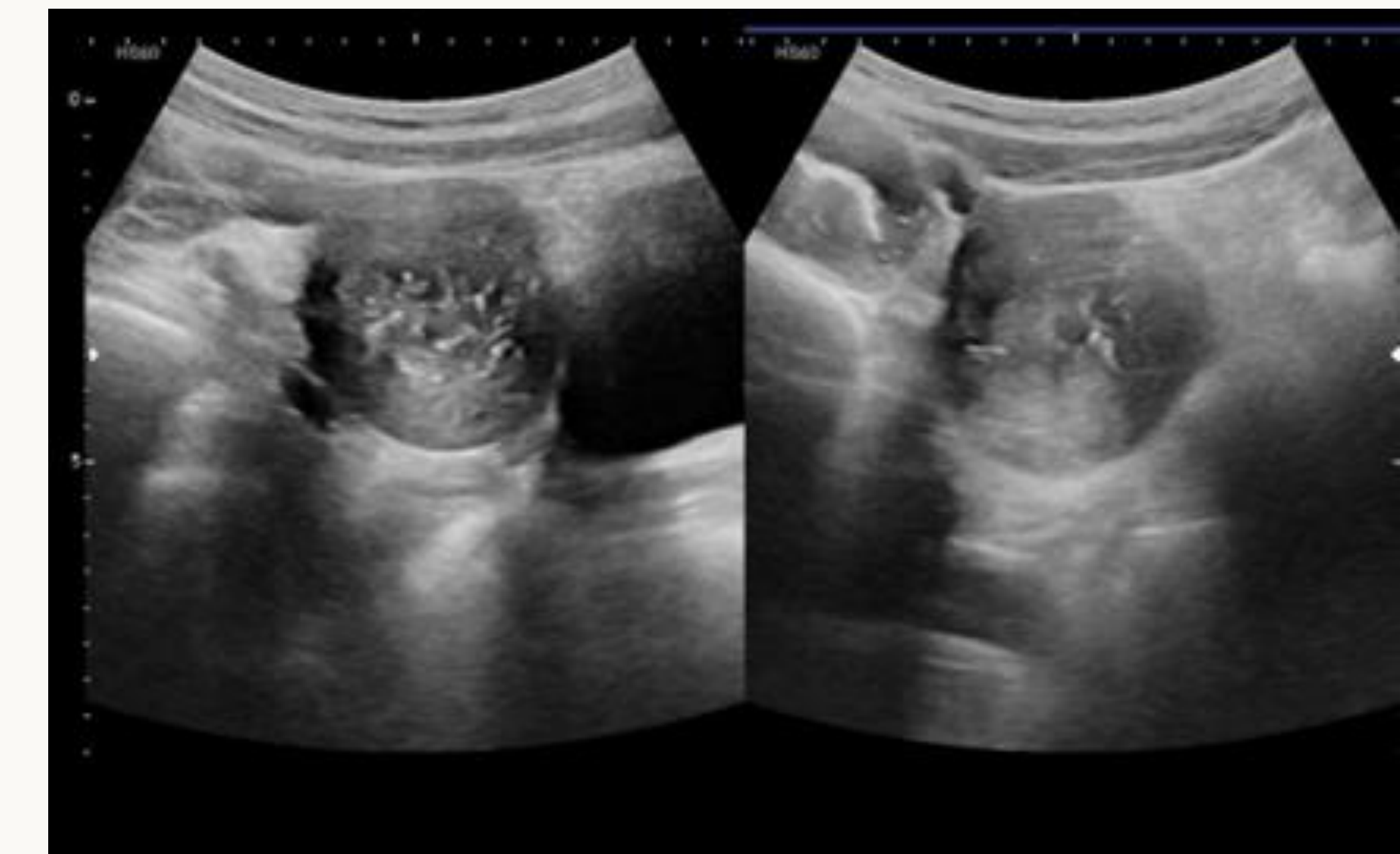


Figure 1. Hyperechogenic islands and echogenic subendometrial lines and buds in rudimentary uterine horns



Figure 2. Functional endometrium and adenomyosis