

Endometriosis and adenomyosis in a Mullerian agenesis patient: A delayed presentation



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ABSTRACT

Mullerian duct anomalies are developmental anomalies resulting from failure in organogenesis, fusion, or the reabsorption of the paired Mullerian ducts. Endometriosis and Mullerian anomalies have a high coincidence rate. Obstructive uterine malformations with functioning endometrium have higher chances of co-incident endometriosis. Continuous bleeding into functional uterine remnants can lead to the formation of hematometra, endometriosis, and adenomyosis. We report a case of primary amenorrhea who reported 18 years after vaginoplasty with pain abdomen due to the presence of endometriosis in bilateral ovaries and adenomyosis in the rudimentary uterine horn.

Key words: Endometriosis; Adenomyosis; Mullerian agenesis; Vaginoplasty

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INTRODUCTION

The reported incidence of Mullerian agenesis ranges from 1 in 4500 to 1 in 5000 women.¹⁻³ Magnetic resonance imaging (MRI) can diagnose rudimentary Mullerian structures in 90% of the patients with Mullerian anomalies.^{2,3} Endometriosis can develop in these patients due to the retrograde flow of menstrual blood from obstructed uterine horns. An approximate delay of 8–12 years can be present between the onset of pain symptoms and surgically confirmed endometriosis.³ The surgical removal is recommended in women with obstructed uterine horns and active endometrial tissue, with agenesis of the cervix and upper vagina. Surgical removal of uterine horn in these women leads to alleviation of endometriotic symptoms.

CASE PRESENTATION

A 37-year-old nulliparous, married female reported a history of pain lower abdomen for 2 months. The pain was of insidious onset, dull aching, and situated in the lower abdomen, not relieved on oral medications, and not associated with any aggravating factor. The patient had a known case of Mullerian agenesis and had undergone McIndoe vaginoplasty 18 years back (2006). Mullerian agenesis was diagnosed by ultrasonography and clinical examination. A blind vaginal pouch of 4–5 cm was found and a cystic swelling of size 5 × 6 cm was found on per vaginal and per rectal examination. The tumor markers (cancer antigen [CA]-125, carcinoembryonic antigen [CEA] 19–9, S. Lactate dehydrogenase) was of normal range except slightly raised serum CA-125 (149 U/mL). Radiological imaging including ultrasonography and MRI

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was done. On T2 weighted MRI (Figure 1), two widely separated, non-communicating hypoplastic horns with an intercornual gap of 70 mm in the transverse axis were discovered. The cervix was found to be absent. The right horn measured 28 × 37 × 32 mm and the left horn measured 36 × 37 × 35 mm. A 7 cm endometriotic cyst was found in between the uterine horns and mild right hydrosalpinx was also detected. The patient was planned for laparoscopy and proceeds and MRI findings were confirmed (Figure 2). On the left side, a small intestine was found to adhere to the posterior wall of uterine horn and adnexal structures. Both-sided adhesiolysis was done but the dissection plane could not be made out easily. The procedure was converted to open laparotomy due to dense adhesions (Figure 3). Bilateral uterine horn excision along with bilateral salpingo-oophorectomy was done. The post-operative period was uneventful. The histopathological report showed adenomyosis in bilateral uterine horns and endometriosis in bilateral ovaries (Figures 4 and 5).

DISCUSSION

In cases of Müllerian agenesis, MRI can reveal rudimentary Müllerian structures in approximately 90% of patients.³

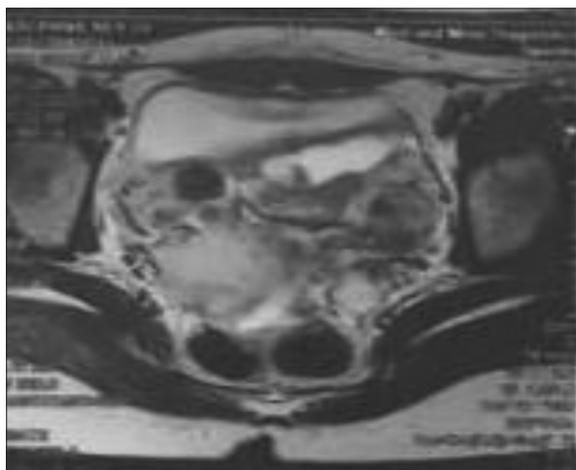


Figure 1: Magnetic resonance imaging showing bilateral rudimentary uterine horns



Figure 3: Image showing rudimentary uterine horn with cavity

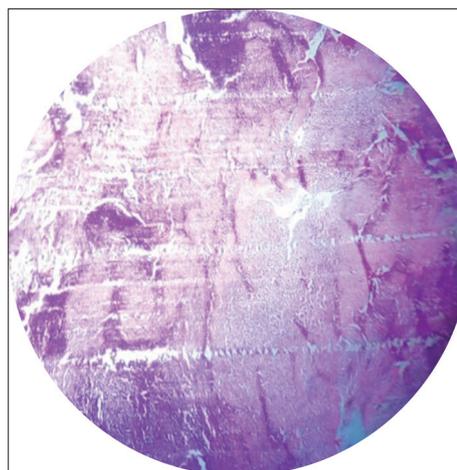


Figure 4: Histopathological slide showing adenomyosis in uterine horns

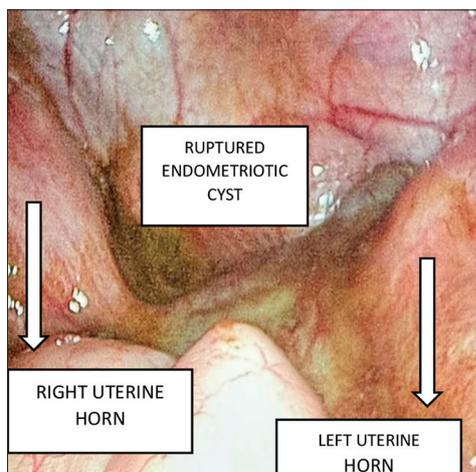


Figure 2: Laparoscopic image of bilateral uterine horns with ruptured endometriotic cyst

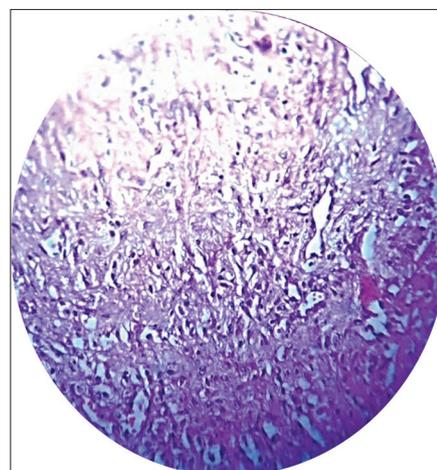


Figure 5: Histopathological slide showing endometriosis in ovary

These are non-functional remnants of the Müllerian ducts, which during normal embryological development, form the fallopian tubes, uterus, cervix, and the upper part of the vagina. Ultrasonography alone cannot diagnose rudimentary Mullerian abnormalities in all cases and MRI was not easily assessable in 2006 for this patient. Hence, the rudimentary horn was a missed diagnosis at the time of primary surgery.

In a case reported by Bindra *et al.*, cervical agenesis, partial vaginal agenesis, and complete bicorporeal uterus with functioning endometrium were present in association with adenomyosis and pelvic endometriosis. Excision of pelvic endometriosis along with hysterectomy of bilateral uterine horns was performed to alleviate the symptoms of pain. Vaginoplasty before this surgery was not done.⁴

In a case reported by Troncon *et al.*, neovaginoplasty was done in an 11 years old for Mullerian anomalies and she reported with endometriotic symptoms 8 years after primary surgery.⁵

CONCLUSION

A comprehensive evaluation with detailed history, physical examination, and advanced imaging techniques

(MRI and 3D ultrasonography) is advisable for patients with Müllerian agenesis to ensure a correct diagnosis of the anatomical anomalies before considering surgery.

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MG- Preparation of manuscript, **MC, VM, SC, SG,** and **MV**- Review of manuscript and treating gynecologists.

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