CASE REPORT

FIBROID ARISING FROM THE RUDIMENTARY UTERINE HORN IN MRKH SYNDROME

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INTRODUCTION: Uterovaginal agenesis, also known as Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a rare congenital anomaly affecting 1 in 4000 -5000 female births. It is the second most common cause of primary amenorrhoea characterized by hypoplastic uterus, absent vagina and normal female external genitalia and normal development of secondary sexual characters. Leiomyomas are the commonest uterine tumours, but the occurrence of fibroid in a rudimentary uterus is very rare.
KEYWORDS: Mullerian agenesis, MRKH syndrome, fibroid, leiomyoma, rudimentary horn.

CASE REPORT: A 42years old unmarried lady presented to our casualty in December 2013 with history of sudden onset of lower abdominal pain. She had primary amenorrhea for which she was evaluated at 18years of age and was diagnosed to have Mayer Rokitansky Kuster Hauser syndrome. She was a known hypertensive and diabetic on treatment. On examination she had normal secondary sexual characters. Abdominal examination revealed a firm swelling of 10x8cms in the hypogastrium with restricted mobility. External genitalia appeared normal. Vaginal orifice was absent. Ultrasonogram showed a large right adnexal mass of 10.5x7.9cms (Fig. 1) probably rudimentary horn fibroid and a small rudimentary horn of 2 x 1.7cms just medial to left ovary. (Fig. 2) Both ovaries were atrophic. CT scan findings were a soft tissue lesion of 8.5 x 8.8 x 10cm in the abdominopelvic location with calcification and non enhancing hypodense areas. (Fig. 3) Uterus was seen separately as hypoplastic.

She was taken for laparotomy after controlling hypertension and diabetes. Laparotomy findings were a large mass of 10x10cms with right tube, ovary and right round ligament attached to it which was the right rudimentary horn with fibroid. (Fig. 4) There was the left rudimentary horn with tube and ovary and multiple small seedling fibroids. Excision of the right rudimentary horn with fibroid and left rudimentary horn and bilateral salpingo oophorectomy was done. Histopathology report came as right rudimentary horn with leiomyomata of 10cms diameter and left rudimentary horn with leiomyomata uterus measuring 1cm in diameter. Both tubes and ovaries were histologically normal.

DISCUSSION: Congenital absence of both the uterus and vagina is termed Mullerian agenesis or Mayer Rokitansky- Kuster- Hauser syndrome. It is a class I Mullerian duct anomaly and it has been subdivided into typical or type A and atypical or type B.(1) Patients with type A syndrome have symmetric muscular buds and normal fallopian tubes. Patients with type B have asymmetric muscular buds (aplasia of one or both buds or, when both buds were found, one bud was smaller than the other one), abnormal fallopian tubes and may be associated with other congenital

anomalies like renal agenesis, syndactyly and cardiac defects.[2] Approximately 15-36% of women with uterine agenesis also have defects in the urinary system and 12% have scoliosis. A syndrome known as MURCS (mullerian duct aplasia, renal aplasia, cervicothoracic somite dysplasia) has been described recently.[4]

In mullerian agenesis, as ovarian function is normal, estrogen dependent pathological conditions can develop including myomas and endometriosis. Reports have described patients with functioning endometrial tissue or even a hematometra in one or both of the rudimentary uterine horns.[3] Leiomyomas of uterus are estrogen dependent tumours which originate from smooth muscle cells of normal uterus. Their growth has been associated with genetic predisposition, hormones and few growth factors. Mullerian ducts have smooth muscle cells at their proximal ends, which probably may give rise to the growth of leiomyoma from the rudimentary uterus in MRKH syndrome.[6] However, the exact etiopathogenesis of leiomyoma from the rudimentary uterus in MRKH syndrome is not known. Non communicating uterine horns of unicornuate uterus has also been reported to have myomas.[5]

CT and MRI are very useful in the diagnosis of MRKH syndrome with leiomyoma and in providing road-map for surgery, latter being very sensitive and specific.[2] On computed tomography (CT), leiomyomas are well circumscribed masses iso- to hypodense to myometrium and show variable enhancement patterns.

Laparotomy/laparoscopy is indicated when a pelvic mass is detected in a case of Mullerian agenesis. The removal of the symptomatic tumor with the adjacent uterine remnant is indicated[7] which can be done laparoscopically.[8]

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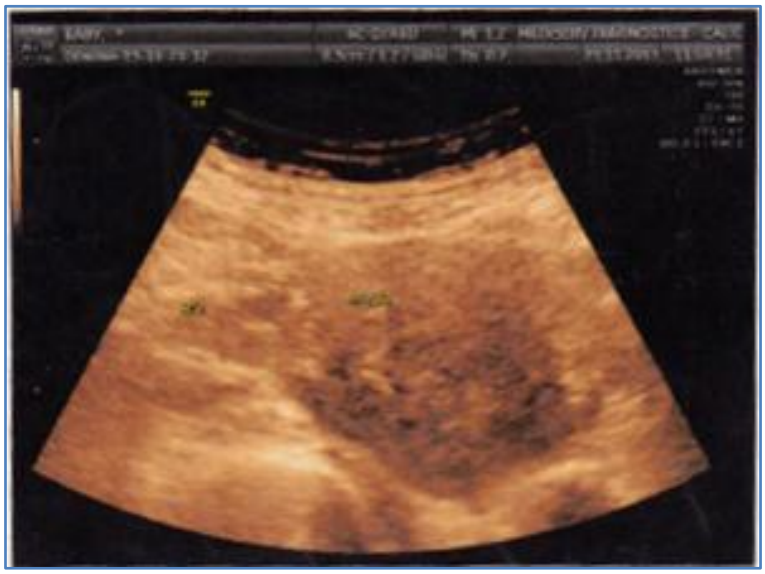

Fig. 1: Ultrasonogram showing the mass and the right ovary

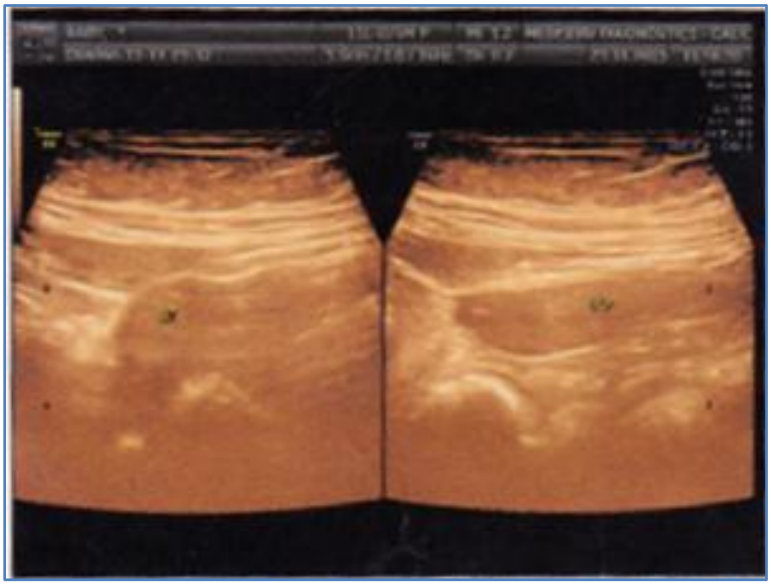

Fig. 2: Ultrasonogram showing the hypoplastic uterus and left ovary

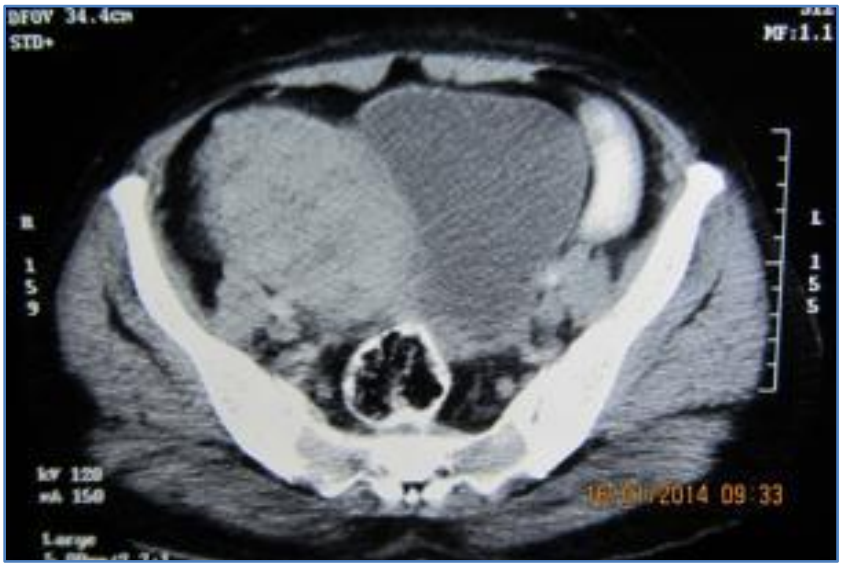

Fig. 3: CT scan showing the soft tissue mass in the pelvis

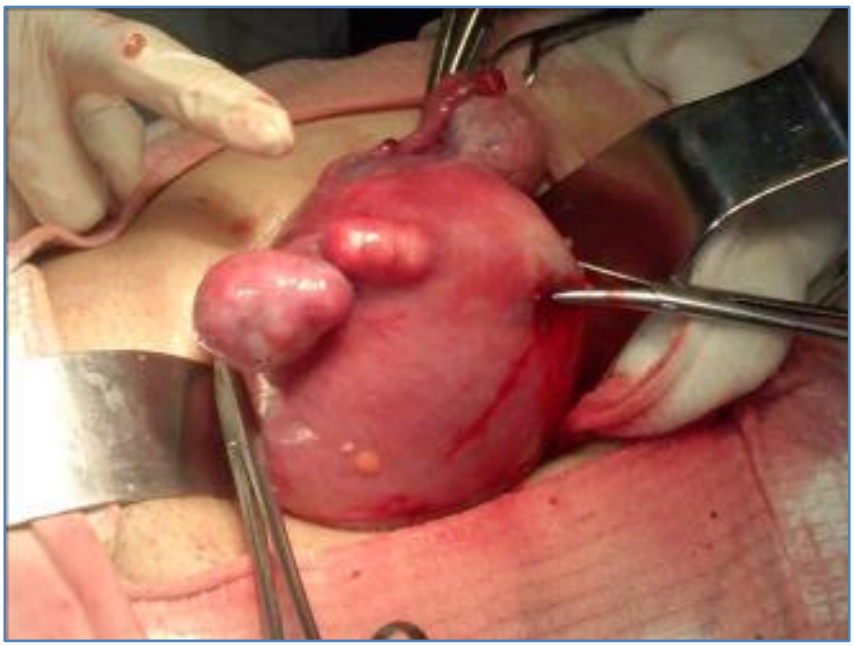

Fig. 4: Fibroid with normal right tube and ovary attached to it

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