A rare case of Leiomyoma in MRKH syndrome

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INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a congenital disorder in females characterized by a hypoplasia or an aplasia of the upper vagina, cervix, and uterus with normal external genitalia and ovaries.[1]

This is a rare syndrome with incidence of one in 4000–5000 female births.

Fibroids are common benign lesions in a normal uterus arising from the myometrium.

The occurrence of a myoma arising from the rudimentary uterus is a rare finding.[2]

CASE DESCRIPTION

A 52 year old nulliparous lady who was a known case of congenital absence of uterus and vagina (MRKH syndrome) presented to us with complaints of recurrent episodes of right lower abdominal pain.

She had history of a congenital cervical block vertebrae and a Patent Ductus Aarteriosus that was corrected at 2 years of age.

The patient was averagely built with well developed secondary sexual characteristics and a normal physical examination. External genitalia was normal. On per speculum examination, a blind vaginal pouch of 1 cm in length was seen. On per rectal examination, a mass of 4 cm was felt above the vault and anterior to the rectum.

MANAGEMENT

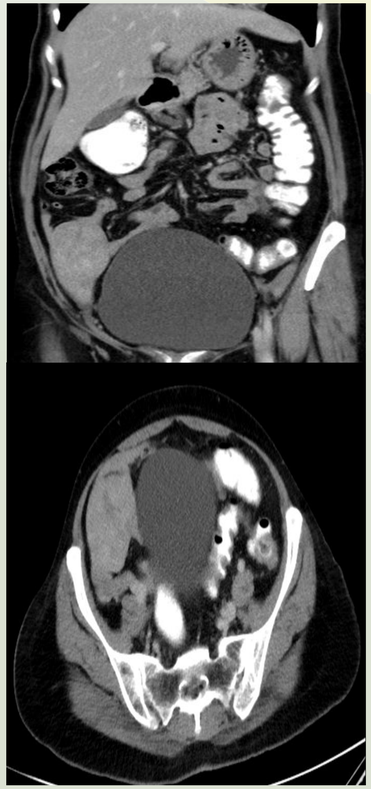
Patient was investigated with tumour markers, with MRI pelvis and CT abdomen with pelvis.

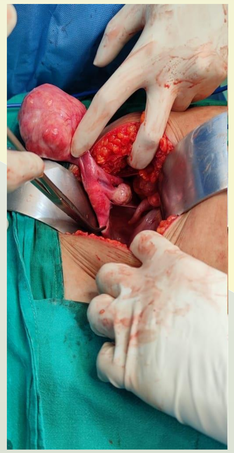
She underwent an exploratory laparotomy followed by excision of bilateral ovaries and remnant uterine horn with fibroid.

LABORATORY INVESTIGATIONS

Inhibin, (pg/ml)	**13**	Hb (g/dl)	10.5
CEA, (ng/ml)	**1.21**	Platelet(K/ul)	263
CA-125, (U/ml)	**9.08**	Wbc (k/ul)	10.12

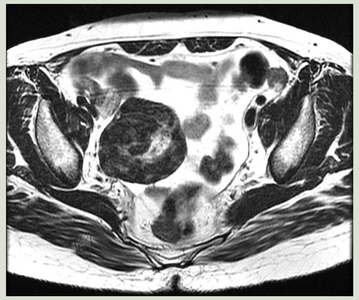
CT scan with solid right adnexal mass of 7.2 x 4.6 x 5.8 cm





Intra-op finding shows a right rudimentary horn with absent left ovarian and uterine pedicles and presence of a pedunculated fibroid arising from the horn

MRI showing a large lobulated right adnexal mass s/o right ovarian neoplasm probably of stromal origin



PATHOLOGY REPORT

Right and left ovaries were unremarkable. A fibroid of 7 x 6 x 4 cm with cut surface showing grey-white whorling was seen. On microscopic examination leiomyoma showing a hyaline degeneration with calcification was observed. The rudimentary horn of uterus (3 x 1.5 cm) showed fibrous tissue with areas of haemorrhage. Endometrium was not visualised.

DISCUSSION AND CONCLUSION

It is possible that a leiomyoma may arise even from a rudimentary uterine horn in case of MRKH. Differential diagnosis include endometriotic ovarian cyst, ovarian tumours and mesenteric cyst. These masses are easily mistaken for ovarian lesions due to lack of uterus as seen in this case. Complete removal of the masses with the uterine remnant is the recommended treatment.[3]

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