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International Journal of Current Research
Vol. 9, Issue, 04, pp.48740-48741, April, 2017

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

ISSN: 0975-833X

CASE STUDY

MAYER ROKITANSKY KUSTER HAUSER SYNDROME WITH HYPOTHYROIDISM WITH BENIGN SEROUS CYST ADENOMA OF OVARY

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ARTICLE INFO

Article History:
Received 26th January, 2017
Received in revised form
05th February, 2017
Accepted 15th March, 2017
Published online 20th April, 2017

Key words:
Mayer RokitanskyKuster Hauser Syndrome, Hypothyroidism, Benign Serous cyst adenoma, MRI

ABSTRACT

Mayer Rokitansky Kuster Hauser Syndrome is a congenital aplasia of uterus, bilateral fallopian tubes and upper 2/3 of vagina; with normal secondary sexual characters and a normal 46XX karyotype. It affects 1/4500 women and its first sign is primary amenorrhoea in young women. A 16 yr old girl presented with primary amenorrhoea with lower abdominal pain. There was blind vaginal pouch and secondary sexual characters normally present. On endocrinological profile patient was hypothyroid.MRI report showed MRKH Syndrome with 8X12cm left ovarian hemorrhagic dysgerminoma. Laparotomy was done where we found left ovarian cystic mass (around 8x12cm) filled with hemorrhagic fluid with intact capsule. Left sided salpingo oophorectomy done. Right side ovary and fallopian tube were normal. In pelvis there were two uterinovaginal bulbs connected by a band like uterus, with agenesis of upper vagina. On Karyotyping 46XX chromosomal pattern was found.

Citation: Dr. Priyanka Rahariya, Dr. Vinu Choudhary and Dr. KusumLataMeena, 2017. "Mayer rokitansky kuster hauser syndrome with hypothyroidism with benign serous cyst adenoma of ovary", *International Journal of Current Research*, 9, (04), 48740-48741.

INTRODUCTION

Mayer RokitanskyKusterHauser Syndromeis a congenital aplasia of uterus, bilateral fallopian tubes and upper 2/3 of vagina; with normal secondary sexual characters and a normal 46XX karyotype. It affects 1/4500 women and its first sign is primary amenorrhoea in young women. There are two subtypes of this syndrome**:**

MRKH Syndrome Type 1 (55-65%)or Isolated mullerianaplasia or Rokitansky sequence. This type of MRKH syndrome occurs as an isolated finding and is characterized by the failure of the uterus and vagina to develop. Sometimes fallopian tubes may be affected but ovaries are generally unaffected and functions normally.

MRKH Syndrome Type 2 (25%)- This characterizes type 1 in association with additional physical findings:-

- Renal anomalies (40%)
- Vertebral anomalies (7%)
- Cardiac anomalies
- Digital anomalies
- Hearing defect

MURCS association (10-12%): Mullerian duct aplasia Renal dysplasia Cervicothoracic Somite anomalies association.

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GRES: Genital Renal Ear Syndrome.

Both MRKH syndrome and hypothyroidism cause amenorrhoea but their occurrence in same patient is very rare. Ovarian tumours are rarely associated with MRKH syndrome.

Case report

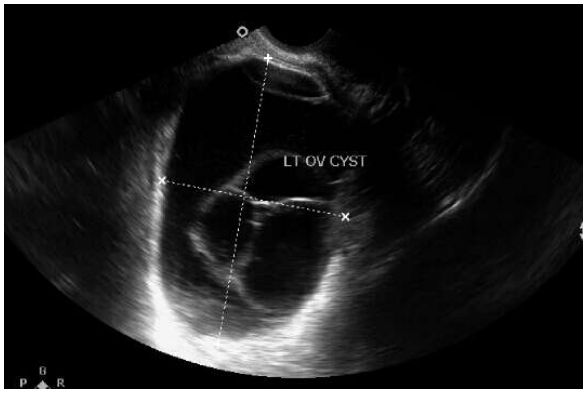
A 16 yr old girl presented with primary amenorrhoea with lower abdominal pain. On examination slightly tender mass present in hypogastrium extending in left iliac region. There was blind vaginal pouch and secondary sexual characters normally present.

Tumour markers: S. AFP, S. Ca125 and S.LDH were raised and S.CEA and S.beta HCG were in normal range.
Values are S.AFP = 12 IU/ml (.5-5.5)
S.Ca125 = 189.8IU/ml (upto 35)
S.CEA = .6ngm/ml (upto2.4)

On endocrinological profile patient was hypothyroid.
S. FTSH = 12.3 microU/ml(.4-4)
S. FT4 = .062 ngm/dl(.09-1.75)
S. FT3 = 1.95 pgm/dl(1.8-4.2)

Rest hormonal profile(S.LH, S.FSH, S.Prolactin) is within normal limits.Patient was having bradycardia which was improved after oral thyroxine given.

Ultrasound findings showed absent uterus with 10X12 cm left ovarian mass



MRI report showed MRKH Syndrome with 8X12cm left ovarian hemorrhagic dysgerminoma

Intra op findings

Laparotomy was done where we found left ovariancystic mass (around 8x12cm) filled with hemorrhagic fluid with intact capsule. Left sided salpingo oophorectomy done. Right side ovary and fallopian tube were normal. There was noextraperitonal involvement of disease. The omentum, lower surface of diaphragm and paraaortic lymph nodes were normal. In pelvis there were two uterovaginal bulbs connectedby a band like uterus, with agenesis of upper vagina.

Intra-operative images

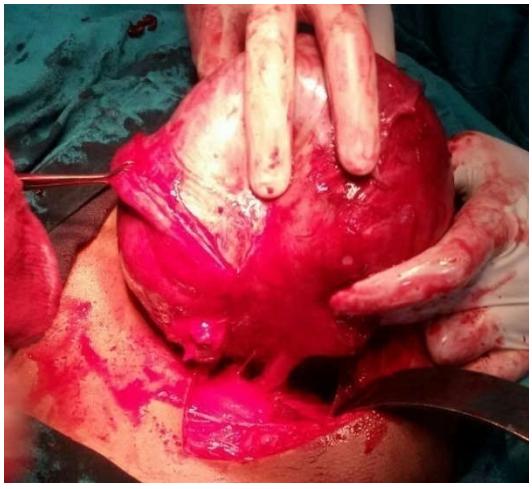


Fig.1. Left side ovarian tumor

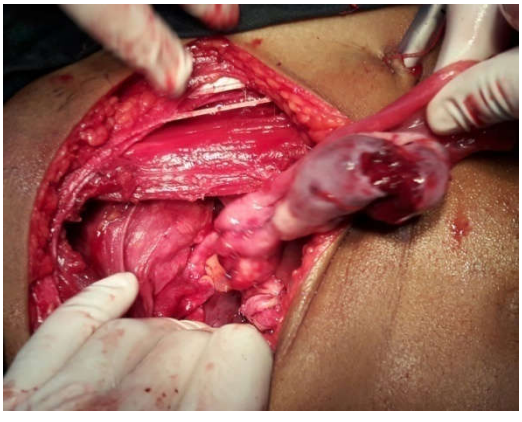


Fig.2. Picture after tumour removal

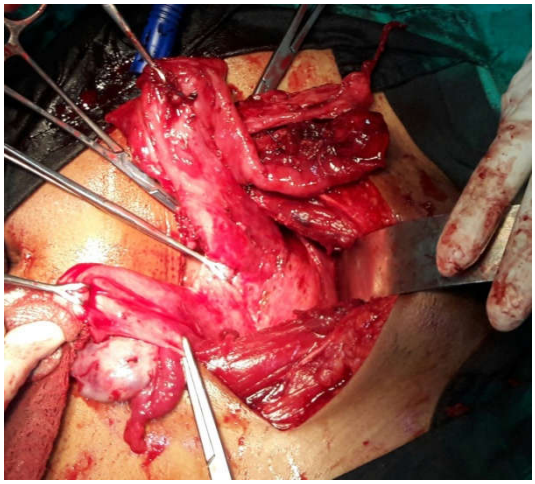
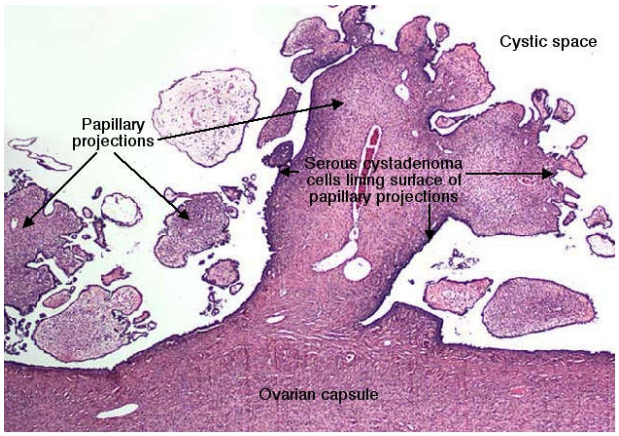


Fig.3. There are two uterovaginal bulbs, connected by fibrous band. Right sided fallopian tube and ovary are healthy

Microscopic examination (Histopathological report): of tumour revealed benign serous cyst adenoma of ovary. Cytological examination of peritoneal washing was negative for malignant cells.



On Karyotyping 46XX chromosomal pattern was found

Conclusion

We are presenting this rare case report of MRKH syndrome with hypothyroidism with ovarian tumour (Benign serous cyst adenoma), has not yet been reported in the literature.This patient was given all pertinent information regarding MRKH syndrome as well as treatment options. After this, it was decided to continue regular follow up until which time she starts having sexual activity.

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