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Title: A RARE CASE REPORT OF HERLYN-WERNER-WUNDERLICH SYNDROME

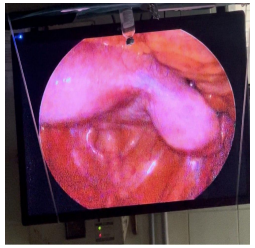
INTRODUCTION

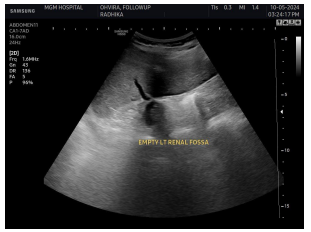
HERLYN WERNER WUNDERLICH SYNDROME or Obstructed Hemivagina and Ipsilateral Renal anomaly **(OHVIRA SYNDROME),**is a rare congenital anomaly of defective fusion of paramesonephric (mullerian) ducts during embryonic development. It includes a **TRIAD of UTERINE DIDELPHYS ,OBSTRUCTED HEMIVAGINA and IPSILATERAL RENAL ANOMALY.** It frequently presents with abdominal pain ,dysmenorrhea and abdominal mass secondary to hematocolpos.

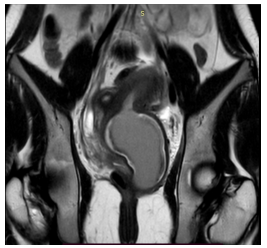
CASE REPORT

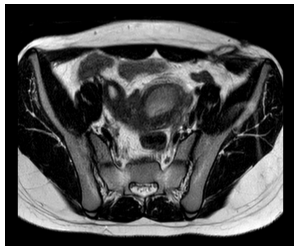
A 14 yr old girl presented with lower abdominal pain since 1month.she attained menarche at 13 years of age and had regular menstrual cycles associated with dysmenorrhea which progresses in severity. No history of urinary symptoms. Her secondary sexual characteristics are normal. On per abdominal examination , a palpable mass is noted . On inspection no bluge is seen at hymen. On further workup with ultrasound , CT and MRI abdomen and pelvis showed presence of 2 uterine cavities , 2 vagina and 2 cervix S/o uterine didelphys and left hematocolpos and obstructed left hemivagina, empty left renal fossa S/o left renal agenesis confirmed by IVP

MANAGEMENT









On laparoscopic exploration, Right horn and Left horn of uterus noted measuring 2x2cm and 8x6cm respectively. A single stage vaginoplasty was done .On re laproscopic examination, bluge at cervicovaginal or vaginal region below the left horn is reduced .foleys kept intravaginally for patency .

DISCUSSION

OHVIRA Syndrome is a rare anomaly with potential short and long term complication. The diagnosis is likely to be missed because of the normal menstruation and nonspecific abdominal pain. Reporting such cases increases awareness of the syndrome and helps to achieve early diagnosis and avoid potential complication.

REFERENCES

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