University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2020, Vol. 6(8)

ERLYN WERNER WUNDERLICH SYNDROME - A RARE CASE REPORT

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Abstract : Herlyn Werner Wunderlich syndrome is a rare congenital mullerian anomaly characterized by uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. It is usually present after menarche with progressive pelvic pain during menstruation secondary to hematocolpos. Awareness is necessary in order to diagnose and treat this disorder properly before complication occurs. MRI is the preferred investigation of choice to delineate uterine malformation. When genital anomalies are encountered, a screening should also be made for congenital anomalies of renal tract and vice versa. We describe below the evaluation and surgical management of a 13 year old girl with the above condition who was diagnosed initially using ultrasound and later confirmed by using MRI. Resection of vaginal septum was done for obstructed right sided hemivagina.

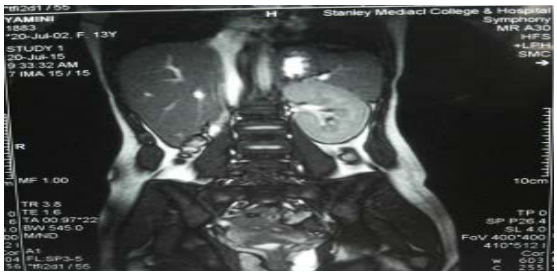
Keyword : Mullerian anomaly, Uterine didelphys, Obstructed hemivagina, Renal agenesis

Introduction

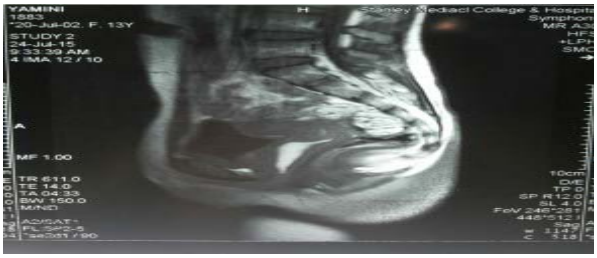
A spectrum of uterine fusion anomalies can occur during fetal development. Lateral fusion defects are the most common type of mullerian duct anomalies which can range from symmetric or asymmetric to obstructed or non obstructed fusion anomalies. 1Herlyn Werner Wunderlich syndrome is otherwise called OHVIRA syndrome (Obstructed Hemivagina & Ipsilateral Renal Anomaly). 5 Mullerian duct anomalies have an incidence of 2-3%, while OHVIRA syndrome has an incidence of 0.1 -3.5% of all such anomalies. Exact cause and pathogenesis were unknown. Symptoms usually present after menarche when hematocolpos develops during menstruation resulting in dysmennorhea and a pelvic mass. Early diagnosis & treatment can relieve the acute symptom and preserve the future fertility.

Case report

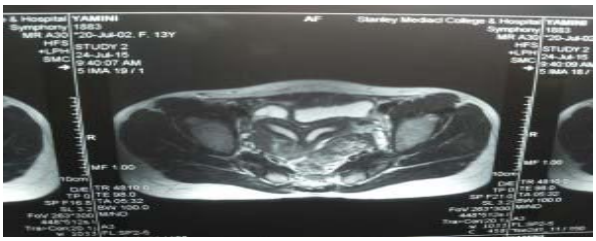
A thirteen year old girl arrived at our emergency department with history of lower abdominal pain and difficulty in passing urine for two days .She was on second day of her menstruation. she has attained menarche about eight months back from when she had regular menstrual cycles and the menstrual bleeding usually lasted for about five days with moderate flow and associated dysmenorrhea. On general examination, it was found that her height was 152 cm and weight as 48kg. She had well developed secondary sexual characters and her vitals were stable. During Abdominal examination, mild lower abdominal distension & tenderness were noted in hypogastric and right lumbar region. On Local examination, it was found that Hymen was intact and external genitalia appeared normal. During per Rectal examination, there was a bulge (firm swelling) of size 5x 3 cm felt in the right lateral aspect of vagina. Bladder was catheterised, clear urine, about 1000ml, was drained after which her pain subsided. Ultrasonogram of abdomen and pelvis showed Hematometrocolpos and right renal agenesis. 3MRI-abdomen and pelvis revealed Uterine Didelphys with Hematometrocolpos noted on right sided uterine cavity due to longitudinal vaginal septum and right Renal agenesis. Congenital Heart disease was ruled out with 2-D Echocardiogram. Under spinal anaesthesia, with aseptic precaution resection of vaginal septum was done for obstructed right hemivagina. There was 150ml of clotted blood was drained. Perioperative and postoperative period was uneventful. Patient was discharged on fifth postoperative day. On follow up patient did not have any complaints.



MRI Abdomen and Pelvis showing right Renal Agenesis and right Hematometrocolpos.



MRI Abdomen and Pelvis sagital view showing right side Hematometrocolpos.



MRI Pelvis showing Uterine Didelphys.

Discussion

Herlyn Werner Wunderlich syndrome is a rare congenital type III mullerian duct anomaly with good prognosis, if diagnosed and treated early. 2It was due to failed resorption or non-fusion of uterine septum at 6-9 weeks of intrauterine fetal life. Uterus , cervix, most of upper vagina developed from Mullerian duct and lower third of vagina developed from Urogenital sinus. Renal tract develops from Metanephric duct. Both mullerian and metanephric duct originates from Genital Ridge. Hence abnormal differentiation of mullerian anomaly is commonly associated with renal tract anomaly. These type of anomalies occur most commonly on right side rather than left side. Obstructed genital anomalies are rarely associated with cardiac and spine abnormalities. MRI is the best investigation modality to detect mullerian duct anomalies. 4Resection of vaginal septum is the treatment of choice for obstructed hemivagina. Early diagnosis and treatment of mullerian duct anomalies is required to prevent complications such as endometriosis, adhesion and subsequent infertility in future.

Conclusion

The prompt, searly and accurate diagnosis of female reproductive tract disorders including herlyn werner wunderlich syndrome is neccesary to prevent complications and preserve future fertility.

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