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Ohvira Syndrome[Uterine Didelphys with Obstructed Hemivagina and Ipsilateral Renal Agenesis] : A Case Report

KEYWORDS: Uterine didelphys, Obstructed hemivagina, Mullerian agenesis , Renal agnesis

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ABSTRACT *OHVIRA syndrome is a rare mullerian developmental defect of female urogenital system. We are reporting a case of 23 yr female married nulliparous with lower abdominal pain and abdominal lump. Patient was giving a confusing clinical picture having different sonographical reports. Finally MRI diagnosed it a rare case of OHVIRA syndrome.*

INTRODUICTION:

OHVIRA syndrome is also known as Herlun-Werner-wunderlich syndrome, its classical presentation is a triad of uterine didelphys , unilateral obstructed hemivagina , and ipsilateral renal agenesis (figure 1).Patient with this anomaly usually presents after postmenarche. These patients presents with andominal pain ,abdominal lump, dysmenorhoea and infertility. Early detection and treatment prevents complication.

CASE REPORT:

A 23 years married, nulliparous female presented with lower abdominal pain ,dysmenorrhea ,dyspareunia and primary infertility.Her menstrual cycle was regular with moderate flow, but associated with dysmenorrhoea since menarche .She was giving history of pain which was initially only during menstrual cycle but since last few years pain was continuous ,dull aching type and irrespective of menstrual cycle .She came to us for this pain, lump and infertility.

On General Examination , patient average built ,weight and height. Her secondary sexual characters were normal. Clinical abdominal examination revealed a14 weeks size lump towards left side. On Local examination, external genitalia was normal. Per speculum examination showed , cervix deviated to right side. During per vaginal examination ,firm mass felt in left part of vagina probably associated with uterus .High resolution tran-svaginal sonography shows, Bicornuate uterus with bulky ovaries. Her MRI Showed didelphys uterus, Left vagina is thinned out with mild haemorrhagic collection in cervix , mild haematometra and left haematosalpinx, multiple small endometriomas in bilateral ovaries, Absent left kidney. Patient planned for laparotomy to relieve her symptoms. Hematometra drained, resection of the left blind uterine horn along with left salpingectomy done .Bilateral chocolate cyst removed.

Discussion

The true incidence of mullerian defects is about 1.1-3.5%[1]. While OHVIRA constitutes 0.16-10% of these anomalies. A strong associate of renal agenesis with uterus didelphys (81%) has also been suggested[2].

There are different clinical variations of OHVIRA syndrome . Most of the females present after menarche due to more evident symptoms. Few patients presents with regular cyclical menses but severe dysmenorrheal and increasing lump abdomen due to patency of single side outflow tract .Usually these cases are misdiagnosed and detected later. Some may present with increasing abdominal pain , initially cyclical and then persistent , having amenorrhea and abdominal lump. These patients mostly detected at early ages and on examination hematocolpos , hematometra and hematosalpinx.

A didelphic uterus is suggestive of an embryologic arrest occurring during the 8th week of gestation which ultimately affects the Mullerian and Metanephric ducts[3]. The exact cause of this developmental defect is not known. However, if one of the Wolffian ducts is absent, the kidney and ureter on the ipsilateral side will fail to fuse at midline. This process may occur completely or incompletely. If the failure to fuse is complete, then a uterus didelphys is formed. The Mullerian duct, on the side lacking the Wolffian duct, displaces itself laterally and cannot come into contact with the urogenital sinus in the center resulting in a blind sac, imperforate or obstructed vagina. However, the distal part of the vagina, originating from the urogenital sinus is not affected[3]. Each ureteric bud develops from the Wolffian duct which is ultimately responsible for the development of the kidney. Occasionally an ectopically obstructed ureter inserts into the obstructed hemivagina on the side of the congenitally absent ipsilateral kidney . The ureteric bud grows dorsocranially into the metanephric blastema inducing differentiation of the metanephric nephrons. If the ureteric bud fails to either form or make contact with the metanephric blastema, the kidney on that side will fail to develop[3].

The diagnosis of OHVIRA , should be suspected in cases of women presenting with pelvic pain and a pelvic mass with ipsilateral renal agenesis. Vaginoplasty with resection of septum , in order to prevent complications or to do hemi-hystrectomy demonstrated successful pregnancy rates similar to females with didelphic uteri [4].

Conclusion:

OHVIRA syndrome is an uncommon congenital anomaly with clinical significance . MRI plays an important role in detection as majority of cases are misdiagnosed. Appropriate surgery should be done . An early correct diagnosis is the goal to relieve the symptoms and prevent complications , caused by retrograde menstruration which may result in endometriosis and also preserve sexual and conception abilities.

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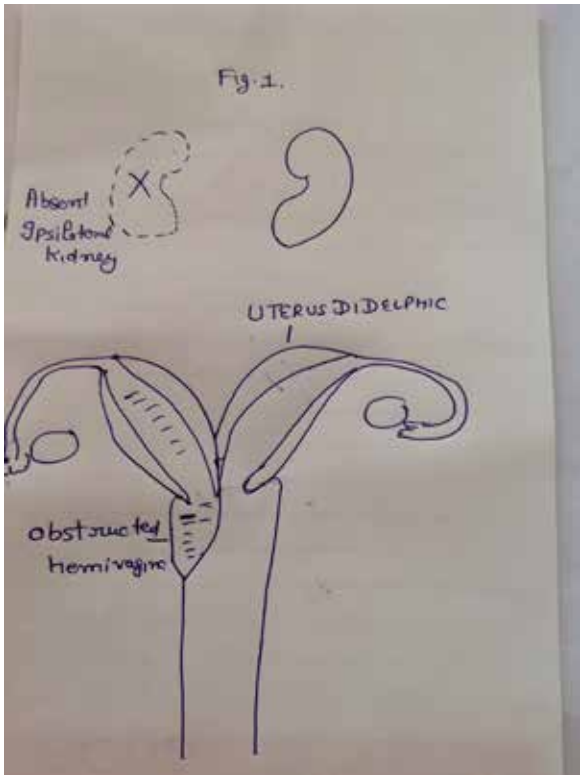

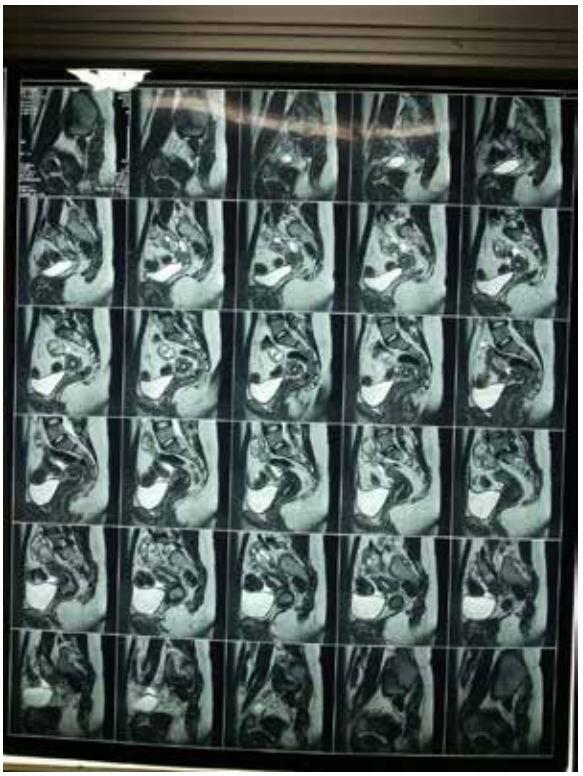

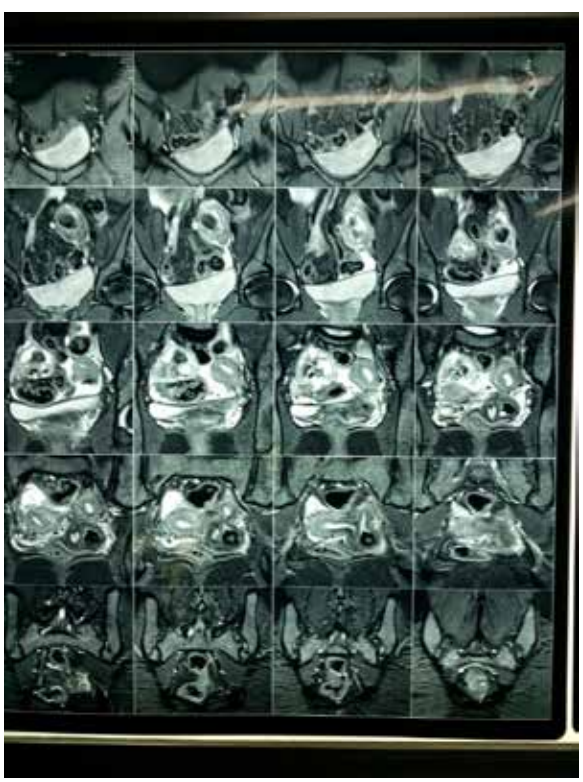

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