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Research Letter

OHVIRA syndrome in post-cesarean period: An exclusive clinical scenario managed by two-staged operative procedure

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Dear Editor,

Obstructed hemi-vagina with ipsilateral renal anomaly (OHVIRA) syndrome is a rare and complex müllerian derivative anomaly (MDA) of pubertal age group, commonly presenting with amenorrhea. We present a case diagnosed with imaging conjugates of OHVIRA syndrome in post-cesarean period.

A 25 years $P_1A_1L_1$ patient presented with vaginal heaviness and abdominal pain for eight months. She underwent cesarean section nine months ago and had hypomenorrhea since then. Menarche was attained 13–14 years back and she gave an undocumented concurrent operative history for an abdominal lump where a large amount of thick altered collection was drained out. Clinical examination suspected a cystic lump in the pelvis and left fornix while the cervix could not be localized. Pelvic ultrasonography (USG) and magnetic resonance imaging (MRI) confirmed left-sided hematometra, hematocolpos and hematosalpinx (Fig. 1). MRI also showed left renal agenesis (Fig. 1A) and collapsed right hemivagina communicating with corresponding uterine horn suggestive of the didelphic uterus (Fig. 1E and F). The clinico-radiological profile lead to the diagnosis of OHVIRA syndrome probably complicated with vaginal restenosis.

Because of pelvic inflammation and anatomical distortion, the management of this patient was planned in two steps. The first stage of management involving drainage of old collected blood was done (Fig. 1G and H) by creating an opening in the most bulged-out part leading to resolution of patient's symptoms. Second-look MRI was done where the vertical vaginal septum, both uteri and their cervices were better identified (Fig. 2). Second-stage procedure involved septal resection and vaginal reconstruction by marsupialization of the vaginal cuff (vaginoplasty). The patient was discharged and the later course was uneventful.

The incidence of MDAs is about 3–4% [1] and 30-50% such cases are associated with renal anomalies [2]. Renal agenesis in combination with didelphic uterus is noted in approximately 81% cases [3] OHVIRA syndrome usually manifests during the puberty and the incidence ranges from 0.5 to 5.0% [4]. Conventional theory of separate origin of the upper and lower parts of the vagina is challenged by a new Acien theory [5] which proposes the origin of vagina completely from the wolffian system (mesonephric duct). Hence, it explains the co-existence of the müllerian and renal anomalies.

Recent history of cesarean section was a unique consideration in this case which was contradictory to the usual presentation of OHVIRA syndrome. Smith et al. [4] published the largest study on 27 cases of OHVIRA syndrome followed over a period of 12 years and highlighted their long term complications and management patterns. Two of their cases showed vaginal restenosis due to incomplete resection by the previous operative care providers. Thus, our case may be considered a chronic undiagnosed OHVIRA syndrome, where an un-planned partial septal resection without vaginoplasty lead to its delayed presentation. Definitive treatment of the syndrome is vaginoplasty with recent trend of single stage procedure, however; some neglected cases require interval procedures. Laparoscopy can be done for clarification of diagnosis and description of anatomy [4].

Anatomical delineation of urogenital system in patients with MDA is essential. The occurrence of renal anomaly with hematometra or any other uterine anomaly should be focused and evaluated keeping in mind the combination of rare possibilities.

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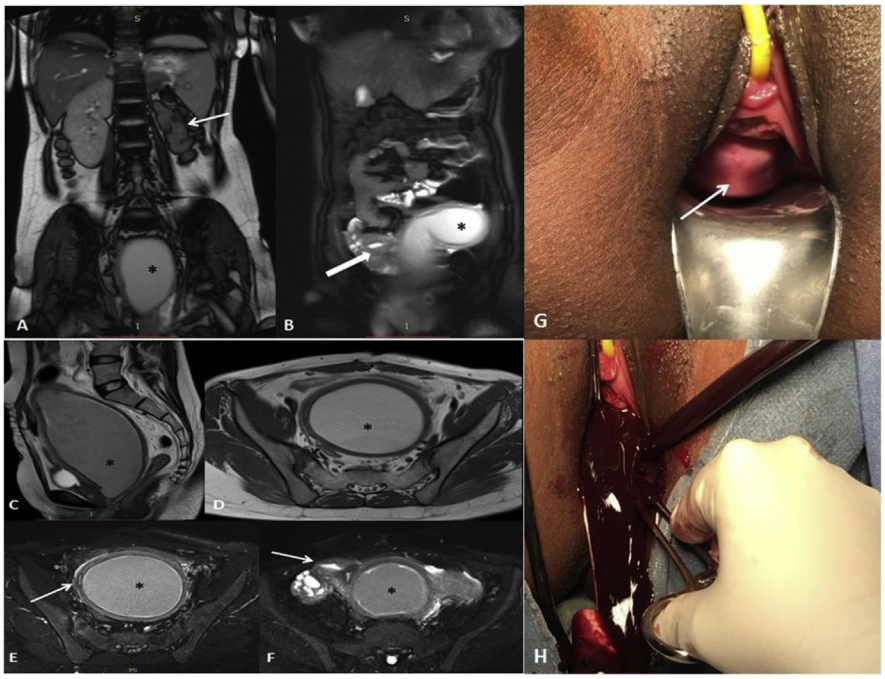

Fig. 1. (A) T2W-Coronal MRI (True fast imaging with steady-state free precession; TruFISP sequence) showing absent left kidney (white arrow) with hyperintense collection in the pelvis (asterisk). (B) T2W-Coronal MRI (Half-fourier-acquired single-shot turbo spin echo; HASTE sequence) showing hyperintense collection continuing in left adnexa (asterisk). Right-sided uterine horn with functional bright endometrium and adjacent ovarian follicles is visualized (thick white arrow). (C) T2W-Sagittal, (D) T1W-Axial MRI pelvis; showing distended uterus, cervix and vagina with T1 hyperintense/blood intensity collection showing abrupt caudal tapering (black asterisk). (E) and (F) T2W-Axial Fat-suppressed MRI in caudal to cranial sections showing linear hyperintense collapsed right hemi-vagina (white arrow) continuing into its corresponding uterus. (G) and (H) Clinical intraoperative images showing bulging and obstructed left hemivagina (white arrow) with opening and draining of the hematocolpos.

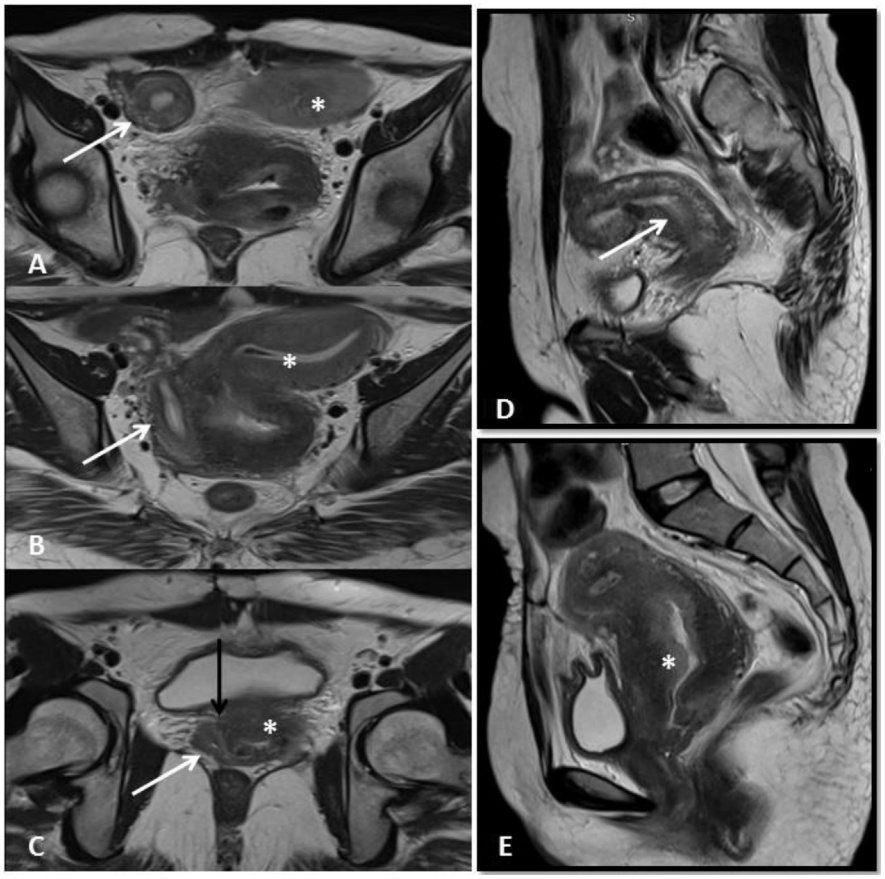

Fig. 2. (A, B and C) Post drainage T2W-Oblique-Coronal MRI images from cranial to caudal sections, showing right sided uterus, cervix and upper vagina (white arrows) with corresponding bulky left-sided utero-cervical and vaginal system (white asterisk). A linear vertical septum (black arrow in C) in between the two hemi-vagina. (D & E) T2W- Sagittal MRI showing right and left uteri and their corresponding cervices.

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None.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

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