OHVIRA and OSVIRA syndrome

Sir,

I read with great interest the article titled, “Herlyn–Werner–Wunderlich syndrome presenting with infertility: Role of MRI in diagnosis” by Ahmad et al. in Indian Journal of Radiology and Imaging.[1] The manuscript is informative. However, I would like to make the following contributions.

OHVIRA, also known as Herlyn–Werner–Wunderlich syndrome, abbreviates a complex urogenital anomaly whose embryopathogenesis is still putative. It reads out as Obstructed HemiVagina and Ipsilateral Renal Agenesis/Anomaly.[1] Besides, it has a didelphic uterus.[1]

Central to understanding of the pathology is embryogenesis of vagina, a disputed topic as yet. While classically the upper vagina is believed to have Mullerian (paramesonephric) roots akin to fallopian tubes, uterus and cervix with sinovaginal bulbs forming the remainder of the lower vagina, recent studies debunk this age-old concept.[2] Acien[2] proposed Wolffian (mesonephric) origin of vagina in entirety – a notion which has been proved in experiments on female rats by Sanchez.[3]

Using Acien’s hypothesis, all three components of OHVIRA can be fully explained.[2] A faulty development of mesonephric duct fails to induce the metanephric blastema, the future kidney. Also because vagina is Wolffian in origin, it too does not develop. Further, lack of growth factors from the mesonephros disturbs the proper positioning and placement of the paired paramesonephric ducts, resulting in nonfusion (uterus didelphys). Hence, the result is OHVIRA syndrome.[2]

On a parallel track is a constellation of urogenital anomalies in males grouped under the so-called Zinner syndrome (ZS). It comprises atresia of unilateral ejaculatory duct that leads to obstruction and dilation of seminal vesicle (seminal vesicle cyst) with ipsilateral renal agenesis.[4] Because all the components of this syndrome are mesonephric in origin, Aswani et al. postulated similar embryopathogenesis of ZS in males as that of OHVIRA in females (as per new hypothesis of Wolffian origin of vagina).[5] This concept thus places ZS as a male equivalent of OHVIRA, unlike previously where ZS was thought to be a male counterpart of Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome in females.[6] The caveat here is that MRKH is a Mullerian anomaly, while ZS is Wolffian in origin.[5] Finally, Aswani et al. proposed OSVIRA as an acronym for ZS, similar to its female equivalent OHVIRA, which expands as Obstructed Seminal Vesicle and Ipsilateral Renal Agenesis.[5]

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Raj Bharatkumar Koticha
Department of Radiology, HBT Medical College and RN Cooper Hospital, Vile Parle West, Mumbai, Maharashtra, India
E-mail: rkoticha@gmail.com

References

Sir,

We allude to the interesting article titled, 'Intracranial translucency as a sonographic marker for detecting open spina bifida at 11–13 +6 weeks scan: Our experience by Teegala et al. [1] The authors have clearly illustrated the significance of the obliteration of intracranial translucency (IT) as a marker for open spinal dysraphism (OSD) in the first trimester. Evaluation of the mid-sagittal plane of the fetal face is an integral part of every first trimester scan between 11 and 13 +6 weeks. As the time spent on each scan is an important consideration in clinical practice, evaluation of IT in the same mid-sagittal plane gives a rich dividend of early detection of an OSD at no extra cost of time. However, there are instances when IT is difficult to assess, as is the case in this study as well.

In a recent study, Ramkrishna et al. proposed a novel marker for detection of OSD in the first trimester, namely the maxillo-occipital line in the mid-sagittal plane. [2] The maxillo-occipital line is a straight line drawn along the superior border of the maxilla that touches the occipital bone posteriorly. The authors observed that the junction of the thalamus with the midbrain was above this maxillo-occipital line in normal fetuses and below this line in the fetuses with open spina bifida. This can be attributed to the descent of the brainstem in open spina bifida due to the egress of the cerebrospinal fluid through the spinal defect. The observation is illustrated in a normal fetus [Figure 1] and a fetus with lumbosacral myeloschisis [Figure 2]. These findings are also evident in the cases shown in the article by Teegala ML et al.

Assessment of the case in this study in which the IT was not clear using the maxillo-occipital line may be enlightening.

Mid-sagittal section of the fetal face:
Still a showstopper

Figure 1: Mid-sagittal section of the face in a normal fetus at 12 +4 weeks. The maxillo-occipital line (red line) is drawn along the superior border of the maxilla (MAX) touching the occipital bone posteriorly. The junction (arrow) of the thalamus (T) with the midbrain (M) is above this line. Intracranial translucency (IT); Cisterna magna (CM)