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Case Report

Diagnosis of Pseudo Hermaphrodite by MRI: A Case Report

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Disorders of sex development, a term referring to a group of congenital conditions in which the development of the chromosomal, gonadal or anatomical sex has been out of character. A 28 years old Sudanese male married for three years with no children, presented with supra pubic and abdominal pain, burning sensation and burning micturation. For exclusion any urinary tract abnormalities, ultrasound examinations for abdomen and pelvis were obtained, and semen analyses was required. MRI images for abdomen and pelvis were requested to confirm or exclude the presence of the uterus and fallopian tube. Regarding the presented case, Ultrasonography should be considered as the initial imaging modality for the initial evaluation of hermaphrodite and because MRI has better soft tissue resolution to characterize gonads, differentiate between clitoral hypertrophy and micropenis. Karyotyping and biochemical assays are also important in the clinical evaluation and differentiation between the true and false hermaphrodite

Keywords: Pseudo Hermaphrodite, MRI, Sex

INTRODUCTION

Disorders of sex development are defined as a condition in which chromosomal sex is inconsistent with phenotypic sex, or in which the phenotype is not classifiable as either male or female (Sax, 2002).

The ability to diagnose Developmental Sex Disorders has advanced rapidly in recent years. (Lambert et al., 2010). Radiology has a major conclusive role in the

diagnosis of these abnormalities before and after birth (Mehdi et al., 2008). Imaging modalities, Ultrasonography (US) and Magnetic Resonance Imaging (MRI), play an important role in demonstrating the genital anatomy for early and appropriate gender assignment and for planning the surgical corrective procedures (Hughes et al., 2006).

We present a case of a patient whose abdominal ultrasonography revealed Pseudo Hermaphrodite . This congenital disorder was depicted with the use of ultrasonography. We complete the imaging approach of the patient by providing MRI images.

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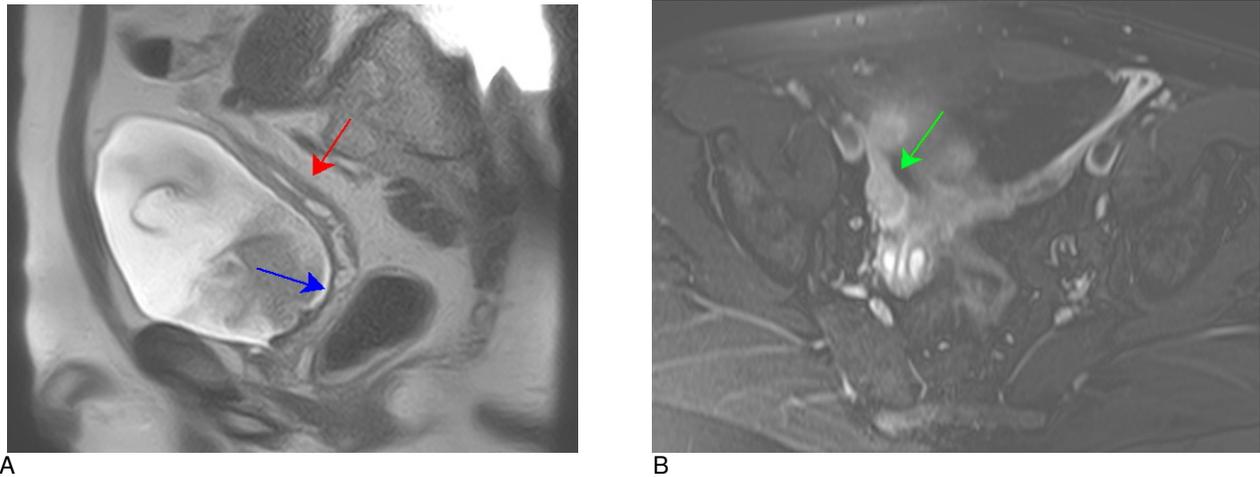


Figure 1. (A) – [The right hemi pelvic region is the seat of an oblique blunt tubular structure measuring about 51x28 mm elicits central cavitory line; the overall features mimic a uterine configuration (red arrow). Besides, this structure seen having intimate relation to the right sided cystic structure likely seminal vesicle versus an ovary (blue arrow) with non visualized left sided one.] (B) – [There is right sided ovoid shaped structure is seen resting upon right iliac vasculature measuring about 23x16 mm and elicits MR signal matching with a testicle (green arrows).]

CASE REPORT

A 28 years old Sudanese male married for three years with no children, was clinically presented with supra pubic and abdominal pain, burning sensation and burning micturation, laparoscopy was done and revealed presence of Uterus and Fallopian Tubes. For excluding any urinary tract abnormalities, ultrasound examinations for abdomen and pelvis were obtained, and Semen Analyses was required. Ultrasound examinations results showed normal both Kidneys with normal size, site and cortex echo-texture and preserved cortico-medullary differentiation. Normal liver, gall bladder and bile ducts. No focal lesions were seen in the pancrease and Spleen with normal echotexture, normal urinary bladder outline. No abdomen or pelvic mass or lymph nodes enlargement or ascites were also seen. Bilateral undescending testicles were seen with empty scrotum. Semen analyses revealed no detected sperms even after centrifugations of the entire provided sample, concluded as Azoospermia . Orchidopxy was done but vas deference was very short so complete repair could not be done and orchidopxy failed. MRI images for abdomen and pelvis were requested to confirm or exclude the presence of the uterus and fallopian tube. The patient was referred to the Magnetic Resonance Imaging (MRI) department.

Images were obtained at different planes (Axial, Coronal and Sagital) and different pulse sequences including (T_1 and T_2) weighted images with and without IV contrast medium and showed that the liver appearance was of average size with grossly intact parenchyma.

There was no focal hepatic lesion or intra-hepatic biliary radical dilatation. The gall bladder appeared normally distended with no obvious filling defect or signal voids. The pancreatic duct was not dilated with normal signal intensity of the pancreas and peri- pancreatic fat tissue planes. The spleen was not enlarged with grossly intact parenchyma. Both kidneys and suprarenal glands appeared intact, there were no obvious renal stone or back pressure changes.

There were no obvious pathological retro peritoneal lymph nodes or abdominal collections. The right hemi pelvic region was the seat of an oblique blunt tubular structure measuring about 51x28 mm elicits central cavitory line; the overall features mimic a uterine configuration (red arrow). Besides, this seen structure had intimate relation to the right sided cystic structure likely seminal vesicle versus an ovary (blue arrow) with non visualized left sided one. The urinary bladder shows uniform mural thickness with no stones. The prostate was of average size with normal signal. A right sided ovoid shaped structure was seen resting upon right iliac vasculature measuring about 23x16 mm and elicits MR signal matching with a testicle (green arrows). The left testicle was seen at left inguinal region measuring about 33x21 mm showing normal MR signal. No evidence of vagina like tissues. The final conclusion was: right hemi pelvic blunt tubular structure mimics a uterine configuration having an intimate relation to right seminal vesicle. The possibility of pseudo hermaphrodite should be considered for Karyotyping, left inguinal testicle and right hemi-pelvic undescended one.

DISCUSSION

Genital anomalies are rare conditions and estimated to occur in 1 in 4,500 births (Soheir et al., 2012).

Male pseudo hermaphrodites are 46, XY genetic males with normal or mildly defective testes. This is due to deficiency in testosterone and di-hydro-testosterone production caused by a deficiency of 5- α reductase (Hughes et al., 2012).

Clinical evaluation of patients with genital anomalies included: physical, biochemical, chromosomal and radiological examinations (Mehdi et al., 2008). In our case the clinical examination recommended the diagnosis, and the semen analyses reflected the absence of sperm. It was considered as pseudo hermaphrodites, because in the true hermaphroditism, fertility is possible (Kim et al., 2002) but our patient was infertile. In our case, ultrasound was firstly used to ensure the patient condition. This was because Ultrasonography is considered the primary modality for the demonstration of the internal reproductive organs, identification of the genital organs, and diagnosis the presence or absence Müllerian derivatives (Chavhan et al., 2008) also ultrasonography is easy and available, but it has some limitations, seeing poor demonstration of complex anomalies (Ravi and Bindushree, 2012) and is operator dependent (Stuart and Brain, 2004)

Magnetic resonance imaging (MRI), has multiplanar facility with excellent soft tissue characterization (Chavhan et al., 2008). Moreover, MRI is used as an addition modality to evaluate congenital anomalies, as it is helpful in clarifying the internal anatomy and searching for internal gonads (Ravi and Bindushree, 2012). This was done in our case. It is predominantly useful for assessment of the Uterus and Vagina, and may identify the presence of intra-abdominal gonads not seen on ultrasound (Rosemary, 2008). MRI in our study, give the final confirmation of the case. This what was stated in the literature that, MRI is useful in studying undescended testicles, as well as differentiating between the penis and clitoral hypertrophy. MRI images presented this condition in addition Karyotyping is recommended for confirmation.

The uterus and immature ovaries have intermediate signal intensity on T₁-weighted images and high signal intensity with an intermediate-signal intensity- outer dark rim on T₂-weighted images. In our study, the sagittal and axial T₂ weighted images were of great value in diagnosing the case (Figure1).

CONCLUSION

Regarding the presented case, Ultrasonography should be considered as the initial imaging modality for the initial evaluation of hermaphrodite, and because MRI has better soft tissue resolution to characterize gonads, differentiate between clitoral hypertrophy and micropenis is considered necessary for appropriate diagnosis. Karyotyping and biochemical assays are also important in the clinical evaluation and differentiation between the true and false hermaphrodite. Thus, the mentioned methods are all acknowledged.

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