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Case Report

Hernia Uterus Inguinale in a Young Woman with Vaginal Agenesis and Lateral Fusion Defects of the Mullerian System

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- Adult female

Abstract

Hernia uterus inguinale is an uncommon entity that is difficult to diagnose preoperatively, and for which the MR imaging features are poorly described. We report the MR imaging findings of bilateral hernia uterus inguinale in a 28 year-old woman with workup for infertility and review the published reports on this entity.

ABBREVIATIONS

HUI: Hernia Uterus Inguinale; MRI: Magnetic Resonance Imaging

INTRODUCTION

Hernia uterus inguinale is a rare entity, particularly in adult females, and is a challenging diagnosis that is often only made post-operatively. The MR imaging features of HUI are poorly described in the literature. Recognition and awareness of this entity and its imaging features could aid management planning,and help preserve uterine and ovarian function.

CASE PRESENTATION

A 28-year-old woman presented with chronic intermittent right groin discomfort. She had a history of scleroderma, primary amenorhoea and infertility. On physical examination, she had a female phenotype, normal stature, and normal secondary sexual characteristics. Her vagina was hypoplastic. No definite groin mass was palpated. Her hormonal profile showed a low oestradiol level of 189 pmol/L, with normal follicular stimulating hormone and luteinizing hormone levels.

Magnetic resonance imaging (MRI) of the pelvis showed lower vaginal agenesis with absence of a normal uterus in the pelvis (Figure 1). The kidneys were unremarkable. The ovaries appeared unremarkable with multiple small follicles (Figures 2-5). Two well-defined ovoid to tubular homogeneous soft tissue structures with intermediate T1 and intermediate to bright T2 signal were seen in the right inguinal canal (Figures 2-4, 8) and at the deep ring of the left inguinal canal (Figures 6-8). At that time, these were thought to represent bilateral ovotestes, suggesting a diagnosis of true hermaphroditism. However, subsequent genetic analysis showed a normal 46XX karyotype with negative SRY gene. In view of the indeterminate nature of the inguinal masses,

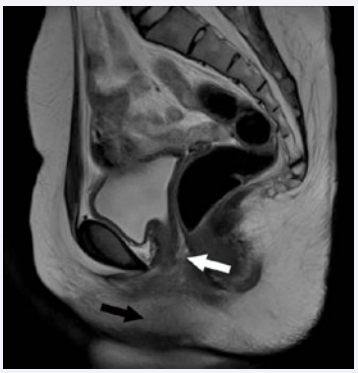

Figure 1 Sagittal T2-weighted MR image, showing absence of lower vagina (black arrow) and uterus (white arrow) in the pelvis.

the patient opted for surgical resection. Surgical exploration revealed bilateral indirect inguinal hernias, each containing a soft tissue structure which was resected. On pathologic review, the right inguinal mass was shown to be a herniated uterus with focal inactive endometrium, and the left inguinal mass was found to be a rudimentary uterine horn. Overall features were consistent with HUI.

Re-review of the MR images showed that neither the uterus nor the rudimentary uterine horn showed the characteristic tri-laminar appearance of a normal uterus, which usually shows bright signal endometrium, dark signal junctional zone, and intermediate signal outer myometrium on T2-weighted images. Instead, both uterine structures showed intermediate to bright signal on T2-weighted images with ill-defined whorl-like bands (Figure 3) without a defined endometrial cavity (Figure 2). A direct connection between the herniated uterine structures and the vagina could not be appreciated.

DISCUSSION

The MR appearance of HUI has not been described in detail previously. In our case of bilateral HUI, small discrete masses that lacked the characteristic tri-laminar appearance of a normal uterus were seen in the inguinal canals at MRI. Both masses showed intermediate to bright signal on T2-weighted images with ill-defined whorl-like bands, and without a defined endometrial cavity. The masses showed intermediate homogeneous T1-signal. The whorl-like appearance is speculated to be related to the myometrial fibres and possibly the primitive endometrial cavity in an underdeveloped uterus. Although lower vaginal agenesis was seen, no connection between the masses and the upper vagina was identifiable.

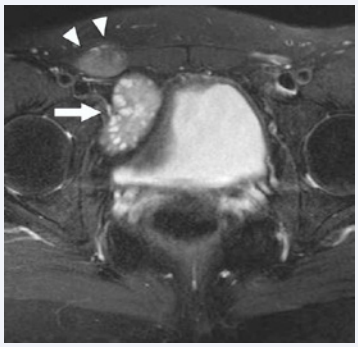

Figure 2 Axial fat-suppressed T2-weighted MR image, showing the right ovary (white arrow) with multiple T2-hyperintense follicles in the pelvis. In the right inguinal canal, there is a well-defined homogeneous soft tissue structure (white arrowheads), ovoid to tubular in configuration, showing intermediate to bright T2 signal. No defined endometrial cavity is seen.

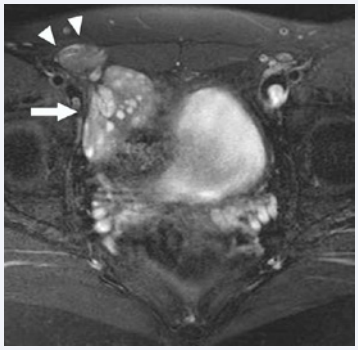

Figure 3 Axial fat-suppressed T2-weighted MR image, obtained at a slightly higher level than in Figure 2, showing the right ovary (white arrow) and the right inguinal soft tissue structure (white arrowheads). Note ill-defined whorl-like bands in the mass.

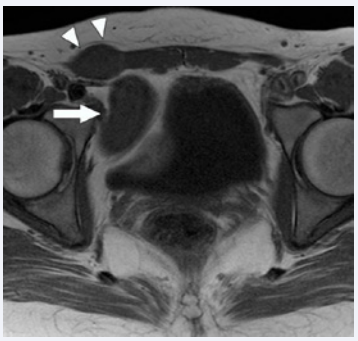

Figure 4 Axial T1-weighted MR image, showing the right ovary (white arrow) in the pelvis. The soft tissue structure in the right inguinal canal (white arrowheads) shows intermediate T1 signal.

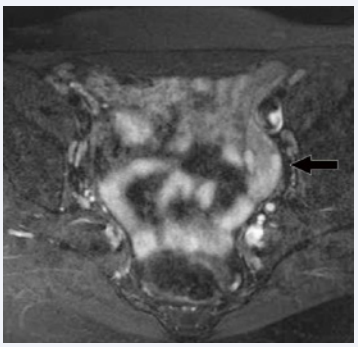

Figure 5 Axial fat-suppressed T2-weighted MR image, showing the left ovary (black arrow) with T2-hyperintense follicles in the pelvis.

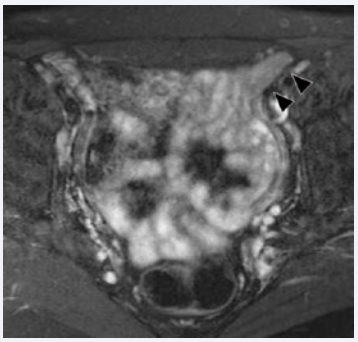

Figure 6 Axial fat-suppressed T2-weighted MR image, obtained at a level inferior to that in Fig. 3, showing a well-defined, tubular, homogeneous soft tissue structure (black arrowheads) at the left deep inguinal ring that shows intermediate to bright T2 signal.

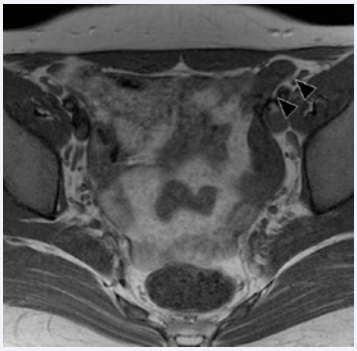

Figure 7 Axial T1-weighted MR image, obtained at the same level as in Figure 5, showing a well-defined, tubular, homogeneous soft tissue structure at the left deep inguinal ring (black arrowheads) that shows intermediate T1 signal.

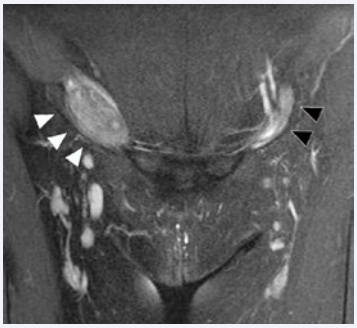

Figure 8 Coronal fat-suppressed T2-weighted MR image, demonstrating the soft tissue structures in the right inguinal canal (white arrowheads) and at the left deep inguinal ring (black arrowheads).

In two articles that described the imaging appearance of the non-herniated unicornuate uterus, Khati et al and Robbins et al described the unicornuate uterus as an asymmetric, laterally deviated structure with a preserved tri-laminar appearance of the normal uterus. The rudimentary uterine horn, if present, may also demonstrate the tri-laminar appearance if it is cavitatory [1,2].

Perhaps due to the atypical appearance of the uterus that can be seen with HUI, the diagnosis of HUI is usually not suggested preoperatively: all but one published cases were diagnosed at surgery and pathology. One prior case report included a single MR image of HUI, but the signal characteristics were not discussed [3]. In another report, Kamio et al described "a homogeneous appearance in the outer layer and had a cavity depicted with high intensity on T2-weighted image", with a single corresponding sagittal T2-weighted image [4]. Our MR findings may further assist with noninvasive imaging diagnosis for future cases.

HUI is a rare condition that is generally found in infants and individuals with disorders of sex development (previously termed as hermaphroditism and pseudohermaphroditism), such as androgen insensitivity syndrome and persistent Mullerian duct syndrome. In phenotypic and genotypic adult women, HUI is an exceedingly rare condition, with only a few case reports to date. In such cases, they are often associated with other congenital anomalies of the genital tract, as in our case.

On review of the medical literature, there are limited surgical case reports of HUI. Among these, there is almost invariably an associated uterine anomaly, most commonly one within the spectrum of lateral fusion defects. There is also a strong association with vaginal and renal anomalies (Table 1). These were also present in our case, except for the absence of renal anomalies.

Table 1: Literature Reports of Adult Female Hernia Uterus Inguinale.

Authors	Age	Hernia Contents	Uterus	Vagina	Kidney
Deutshman et al[4] (1923)	34	Uterus, ovary, oviduct	LFD	Agenesis	
Thomson et al[5] (1948)	20	Uterus, ovary, oviduct	LFD	Agenesis	
Mahmood et al[6] (1970)	35	Uterus	LFD	Normal	Agenesis
Riggall et al[7] (1980)	16	Uterus, ovary, oviduct	LFD	Agenesis	Normal
Elliott et all[8] (1989)	19	Uterus, ovary, oviduct	LFD	Normal	Agenesis
Keating et al[9] (1993)	26	Uterus, ovary, oviduct	LFD	Normal	Agenesis
Chen et al[10] (1994)	56	Uterus, ovary, oviduct		Normal	
Kriplani et al[11] (2000)	20	Uterus, ovary, oviduct	LFD	Agenesis	
Kamio et al[12] (2009)	25	Uterus	LFD	Normal	Agenesis
Kokcu et al[13] (2010)	23	Uterus, ovary, oviduct	LFD	Normal	
Mandel et al[14] (2010)	24	Uterus	LFD	Normal	Agenesis
Bar-Joseph[3] (2012)	25	Uterus, ovary, oviduct		Agenesis	Agenesis
Present case (2014)	28	Uterus	LFD	Agenesis	Normal

Abbreviations: LFD: Lateral Fusion Defect

An indirect inguinal hernia is the most common type of groin hernia, and results from the failure of embryonic closure of the deep inguinal ring and the persistence of a patent peritoneal evagination in the inguinal canal. In males, this processus vaginalis accompanies the testis as it descends into the scrotum. In females, the canal of Nuck accompanies the round ligament of the uterus; the ovaries also descend into the pelvis but do not enter the inguinal canals. If the peritoneal evagination remains patent, this is known as a patent processus vaginalis or canal of Nuck. Owing to this embryology and anatomy of the inguinal canal, HUI has often been reported in male infants with persistent Mullerian derivatives and disorders of sex development. It is rare in adults and women with normal secondary sexual characteristics. In adult women, the association of HUI with lateral fusion defects of the Mullerian system may be related to the excessive length and mobility of the suspensory ligaments.

Our patient had bilateral indirect inguinal hernias containing a unicornuate uterus and a rudimentary uterine horn, associated with lower vaginal agenesis and lateral fusion defects of the Mullerian system. In view of the absence of haematometra, the uterus was believed to be non-functional. Her groin discomfort resolved after the surgical resection. She also subsequently underwent vaginoplasty with the creation of a neovagina.

In summary, HUI should be considered in adult female patients presenting with a groin mass or pain associated with an absent intraperitoneal uterus and vaginal anomalies. At MR imaging, the uterine structures of HUI may lack the typical tri-laminar appearance of a normal uterus at T2-weighted imaging and may not show an obvious connection to the vagina. Rather, HUI may appear as an isolated intermediate to bright signal mass with ill-defined whorl-like bands, without a defined endometrial cavity, on T2-weighted images. If suspected, care should be taken to identify and preserve the ovaries, as well as the uterus which may be functional, in order to prevent premature ovarian failure and preserve fertility.

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