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Rokitansky-Mayer-Kuster-Hauser Syndrome

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1. Clinical Medical Image

The patient was 18 years old, followed for Hodgkin's lymphoma at puberty stage under treatment, admitted for absence of menarche and pelvic pain. Pelvic ultrasound revealed uterine agenesis, confirmed by pelvic MRI which showed uterine agenesis with hypoplasia of the upper 2/3 of the vagina continuing with elongated T2 hypointense structures in the shape of a corncob in connection with Muller's ducts. The ovaries were without abnormality and the kidneys were unremarkable.

Pelvic MRI in axial and sagittal T2 section, with axial section passing through the kidneys showed uterine agenesis within the framework of Mayer Rokitanski -kuster -Hauser type 1 syndrome.

2. Discussion

Müllerian congenital anomalies (MCAs) represent a large group of polymalformative syndromes associating a utero-vaginal malformation with malformations of the urinary tract and/or the spine and/or the ENT sphere [1].

The most severe of these is Rokitansky-Mayer-Kuster-Hauser syndrome (RMKHS). Although it is the second most common cause of infertility after Turner's syndrome, it is a rare disease that results from the lack of development, fusion defect or resorption defect of the Müllerian ducts.resorption of the Müller's ducts [2].

Polygenic and genetic patterns of inheritance have been described in the expression of these anomalies. As well as the role of factors related to the intrauterine environment (uterine infections) and extrauterine (ionizing radiation, drugs with teratogenic effects such as Diethylstilbesterol (DES)) which have also been incriminated in the developmental defect of the genital tract in the female fetus [3].

The first sign is primary amenorrhea, and 6-10% of patients complain of chronic pelvic pain due to the presence of functional endometrial tissue responsible for hematometrics, endometriosis and myomas. HRCS can be subdivided into two types: isolated (type I) or associated with other malformations (type II) [6].

Imaging, in particular suprapubic ultrasound, is used as a first line of defence.

First intention showing the absence of uterine structure between the

Bladder and rectum. However, it may give a false image of a hypoplastic uterus

hypoplastic by showing a quadrangular retrovesical structure, without a cavity, and therefore not containing a hyperechoic line, normally corresponding to the uterine mucosa: this is the vestigial blade located under the median part of the transverse peritoneal fold on the posterior surface of the bladder, where the utero-sacral ligaments are inserted [4].

Magnetic Resonance Imaging (MRI) is a more sensitive and specific examination allowing a precise diagnosis of uterine aplasia, vaginal aplasia or hypoplasia as well as the visualization of rudimentary horns and ovaries [5]. The examination thus allows the lesion assessment of a possible pelvic endometriosis and the search for other associated malformations, in particular renal and bone, by means of basic sequences (T2 Sagittal, T2 oblique coronal in the plane of the major axis of the uterus, oblique axial perpendicular to the major axis of the uterus, a plane chosen in T1 and T1 FAT SAT weighting).

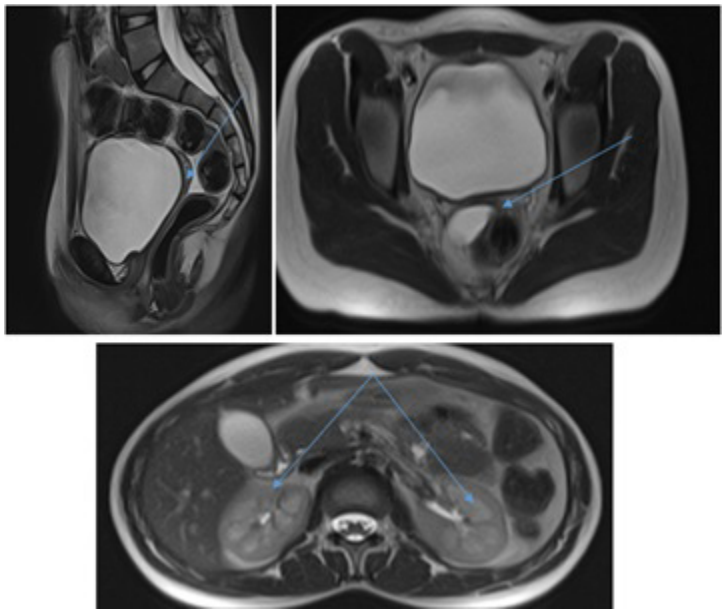

Figure 1:

Table 1: Different characteristics of Mullerian congenital anomalies

	Sd de MRKH	Aplasie vaginale isolee	Sd WNT4	SD d'insensibikite aux androgenes
Amenorrhee primaire	oui	non	oui	non
Uterus	Absent ou hypoplasique	present	absent	absent
Vagin	Absent ou hypoplasique	absent	absent	present
Gonades	ovaires	ovaires	Gonades masculinises	testicules
Caracteres sexuelles secondaires feminins	present	present	present	Pilosite pubienne peu develop
Hyperandrogenie	non	non	oui	non
Caryotype	46 XX	46 XX	46 XX	46 Xy

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