

Syndrome of Rokytansky A Case Report and Review of the Literature

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Case Report

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Abstract: Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) is a rare entity which includes a set of deformations of the feminine genital tract and the internal reproductive organs. The surgical treatment aims at restoring at the most the vagina to make possible sexuality but the function of reproduction remains limited. We describe a case of 24 year old patient who consults the day after its wedding night for imperforation of the hymen. And to whom we discover a syndrome of rokytansky type1 with vaginal total agenesis and hypoplastic uterus and ovary. The creation of a neovagina was impossible

Keywords: Rokytansky; amenorrhea; néovagina; sexuality; psychology.

INTRODUCTION

Rokitansky Kuster Hauser's syndrome (RKH) is an anomaly rare malformative (1/5 000 women); associating an uterine and vaginal aplasia with normal ovaries. To woman with a normal karyotype 46, XX [1].

MClassified as either MRKH syndrome type 1 (corresponding to isolated utero-vaginal aplasia) or MRKH syndrome type 2 (utero-vaginal aplasia associated with other malformations) [2].

CASE REPORT

It is a 24-year-old girl admitted in emergencies the day after its wedding night for an imperforation of the hymen; which presented a primary amenorrhea for which she has never consulted.

She has no medical or surgical history and no family history of malformation or of congenital defects. The beginning of the juvenile development was towards the age of 12 years but patient did not have her menarche.

The clinical examination finds sexual characters secondary developed: developed breasts; presence of a pubic pilosity and under armpits; small lips and big lips of normal aspect. A normal size: 1m70

/ 75 kg, slim waist; hips widened. No sign of scrawny deformation. The gynecological examination finds the absence of clitoris; of the hymen and the vagina. We realized a MRI which showed: (fig 1; 2; 3) a very small uterus which is 1.8 cm (hypoplastic). Cervix of uterus not identified with absence of visualization of the vagina. The both ovary are very small. 2 loins are normal.

Karyotype was normal. Hormonal balance sheet was not realized.



Fig-1: Sagittals cutting of a magnetic resonance imaging which shows the absence of vagina and a womb hypoplastic

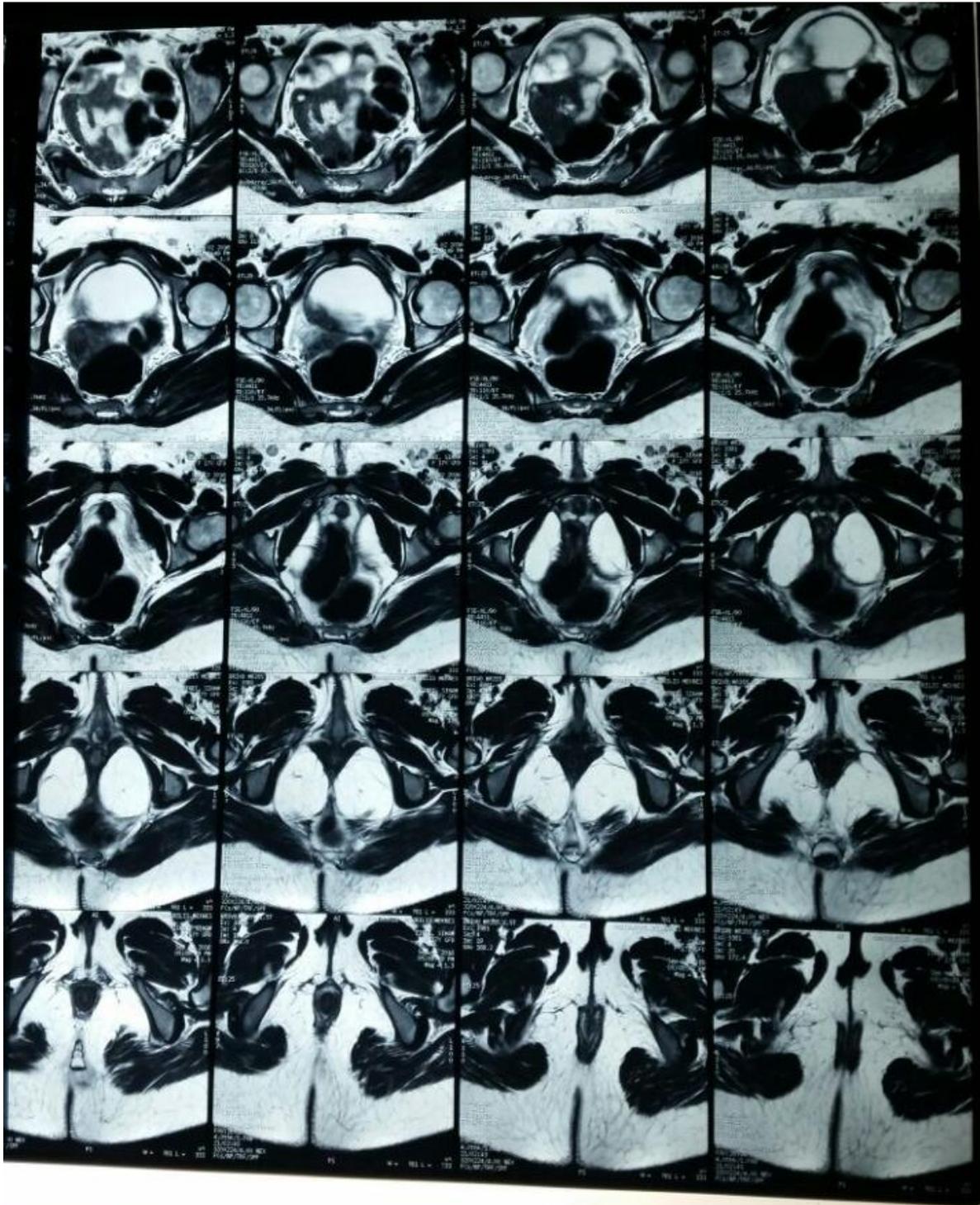


Fig-2: Cross sections of an image by magnetic resonance which shows a hypoplastic womb



Fig-3: Cross section of an image by magnetic resonance with injection which shows Small-sized ovaries

DISCUSSION

The syndrome of Mayer-Rokitansky-kuster-Hauser is a rare disorder which affects the women, characterized by the incapacity of the uterus and the vagina to develop correctly at the women's who have a normal ovarian function and normal secondary sexual characteristics during the puberty, but have no menstrual cycle (primary amenorrhea). The severity of

the syndrome MRKH can vary considerably and the disorder is decomposed into type I, who occurs as an isolated discovery, and of type II, which occurs with anomalies of systems of additional organs, in particular loins and skeleton[3].

A radiological balance sheet is necessary to establish an exact lesional balance sheet and detect the

presence of the other associated malformations one often resorts to the magnetic resonance imaging for a precise diagnosis [4].

On the embryological plan, the utéro-vaginal aplasia is understandable by a cessation of the progress of both channels of Müller after surcroisement of the channels of Wolff; trunks deriving from the proximale part of the channels of Müller are normally formed [5].

Besides the limited sexuality function of the extent of the vaginal atresia the reproductive function is limited and the recourse to fertilization in vitro can solve the problem in case of the presence of a normal womb.

A vaginal reconstruction could satisfy a normal sexual function if it is made a success and we have at present plusieurs technique for the creation of an anatomical and functional neovagina [6]. The application of vaginal dilators on the vaginal dimple for at least 20 minutes/day for several months.

Psychological support and counseling is strongly recommended for affected women [7]. In our case it is about a syndrome of typical MRKH I with total absence of the vagina in what made the surgical coverage delicate; we were not able to have the consent of the patient which benefited from a psychological accompaniment.

CONCLUSION

This observation highlights the possibility of diagnostic delay of the syndrome of Mayer rokytansky kuster Hauser in spite of the existence of a primary amenorrhea seen the normal development of secondary sexual characters what implied an enormous psychological suffering s In front of the difficulty of a normal sexual intercourse and the impossibility of the conception of a pregnancy. The surgical treatment can

be useful in limited cases by restoring the vagina to allow a normal sexuality.

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