Case Report

Mayer Rokitansky Kuster Hauser (MRKH) syndrome

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Abstract

MRKH syndrome is condition that belongs to class 1 Mullerian duct anomalies characterized by congenital absence of uterus and upper vagina. We feel it is important to understand this fairly uncommon condition due to the social and medicolegal aspects associated with it.

Keyword: MRKH syndrome , Mullerian anomalies

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Received Date: 04/10/2015 Revised Date: 20/11/2015 Accepted Date: 01/12/2015

Access this article online

Quick Response Code:

Website: www.statperson.com

DOI: 16 December 2015

INTRODUCTION

MRKH has a reported evidence of 1:4000-5000 live births. There are two different forms of this syndrome, Typical form (TYPE A): congenital absence of the uterus and upper vagina with normal ovaries and fallopian tubes. Atypical form (TYPE B): associated abnormalities of ovaries, fallopian tubes and renal system.[1]

CASE REPORT

A 19 years old healthy unmarried female was referred to our department for MRI PELVIS study with history of primary amneorrhoea. There was a family history of delayed menarche due to which she did not consult any doctor until recently. On examination, she had normal secondary sexual characteristics and external genitalia were well developed. Primary investigation in the form of pelvic ultrasound was done outside which was suggestive of? hypoplastic,? Absent uterus with normal ovaries. Further MRI evaluation was suggested. MRI findings revealed absent uterine tissue with agenesis of upper 2/3rd of vagina with normal sized bilateral ovaries-findings suggestive of MRKH syndrome.

DISCUSSION

Mullerian ducts are paired embryological structures that normally fuse between 6th and 11th weeks of gestation forming the uterus, fallopian tubes, cervix and most of the upper vagina.[2] Failure of development of Mullerian ducts results into MRKH syndrome. However in 0.1-10% of the general population, fusion fails to occur or faulty fusion occurs resulting in various mullerian duct abnormalities like uterine hypoplasia/agenesis, unicornuate, septate, arcuate, bicornuate and uterus didelphys.[3,4] Clinically patients presents with normal hormonal levels with fully functional gonads. Patients may present with primary amenorrohoea, infertility, obstetric complications and endometriosis.[2,5] It is estimated that about 15% of patients with recurrent miscarriages have mullerian duct anomalies[6].In addition 30-50% of these women have concurrent renal anomalies which include agenesis, ectopia, fusion, malrotation and duplication.[2,3,5] Patients with this syndrome may present with an atrophic uterus, however vaginal abnormalities may range from hypoplasia to agenesis. Although ultrasound is generally the initial investigation performed MRI remains the definitive diagnostic modality. T2 sequences aid in the zonal anatomic differentiation. Additional oblique, coronal and axial planes must be added to ensure adequate visualization of the uterine fundus.

How to site this article: Aruna Deodhar, Valmik Kadpe, Pranav Mahadevkar, Vedprakash Biradar. Mayer Rokitansky Kuster Hauser (MRKH) syndrome. *International Journal of Recent Trends in Science and Technology.* December 2015; 17(2): 116-117. http://www.statperson.com (accessed 18 December 2015).

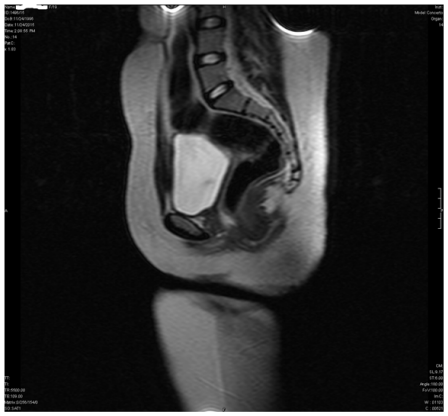

Figure 1

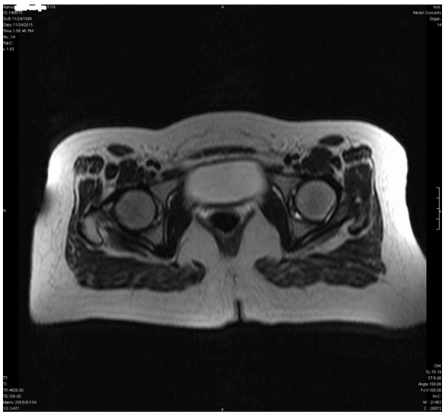

Figure 2

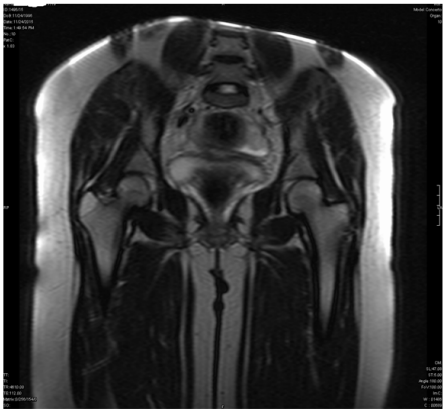

Figure 3

Legend

Figure 1: T2W Sagital MRI image
Figure 2: T1W Axial MRI image
Figure 3: T1W Coronal MRI image

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Source of Support: None Declared
Conflict of Interest: None Declared