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Case Report

An unusual case of septate uterus with double cervix and longitudinal vaginal septum–with pregnancy

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ABSTRACT

Various theories and classification systems have been proposed from time to time, to understand the embryology of the female genital system. Even then at times we come across something different, something new, which again questions our understanding of this subject matter and the theories which are most accepted worldwide. Mullerian anomalies result from the improper development and fusion of the embryological mullerian ducts. It is generally considered to occur in less than 5% of women. Here we present a rare and interesting case of a young female having septate uterus with double cervix and a longitudinal vaginal septum with pregnancy.

1. Introduction

The development of female genital tract is a complex process. The indifferent gonad differentiates to the ovary. The mesonephros, wolffian and mullerian ducts differentiate in an orderly manner to form the uterus, vagina and lower urinary tract. Abnormal differentiation of these structures can result in congenital abnormalities which can adversely affect the female reproductive tracts, renal tract and lower intestines. A number of rudimentary structures can persist and be encountered in clinical practice, most commonly are those derived from the wolffian ducts [1]. These anomalies, commonly known as the mullerian duct anomalies, can hamper the normal life of a female like infertility, hematometra, dyspareunia. So, there lies the need to understand the anomalies in detail and for that we must be aware of the embryological development of the female genital tract. There are various theories and classification systems given from time to time to explain the normal development and to classify these anomalies. The most accepted is the one given by American Fertility Society (AFS) [2]. But as we all know biology is a subject of exceptions, sometimes we come across such different instances, which are hard to be explained by the prevailing theories. We present to you one such rare and interesting case of a young female having septate uterus with double cervix and longitudinal vaginal septum with pregnancy.

2. Case report

A 20 years young female presented in the obstetrics and gynaecology outpatients department of our hospital with the history of bleeding per vagina after 3 months of amenorrhea. She attained menarche at 13 years of age. There was a positive history of dyspareunia with penetration difficulties in the past. There was no history of any drug intake or any miscarriages in the past as she conceived for the first time. Past medical and surgical histories were unremarkable. There was no family history of any congenital anomalies. Her clinical examination was normal. On per vaginal examination, a longitudinal vaginal septum was felt along with two cervices.

She was then further investigated with two and three dimensional sonography. On transabdominal and transvaginal sonography, ante-verted uterus showed two separate endometrial cavities with no dip in the myometrium in the fundal part of the uterus. The right side showed a single intra-uterine gestational sac of mean sac diameter 3.4 cm, with yolk sac and foetal pole of crown rump length 2.5 mm corresponding to a gestational age of 7 weeks 1 day. There was no cardiac activity in the foetus. The left side was normal. There were two cervical canals and a longitudinal

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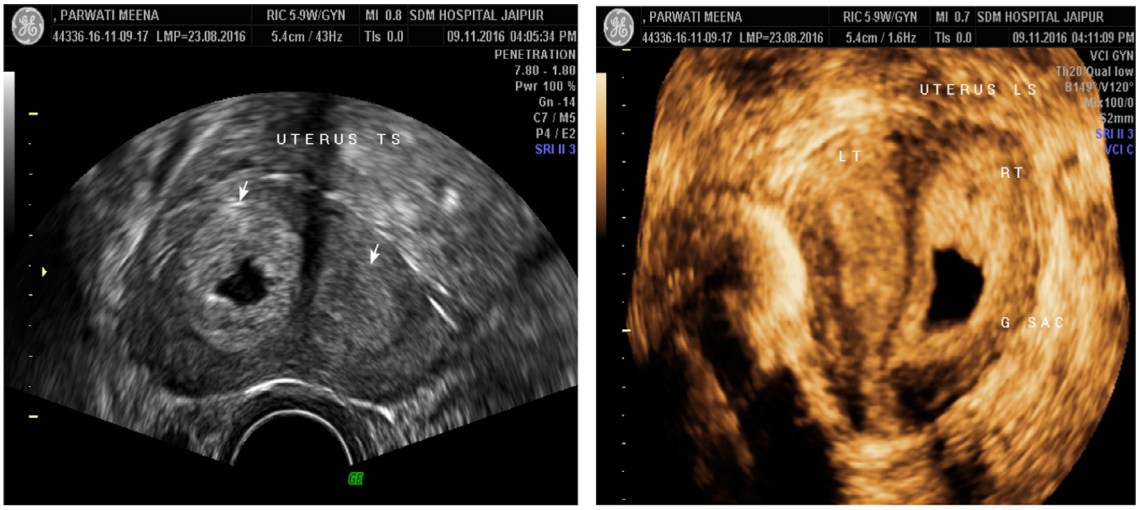

Fig. 1. (a and b) Two dimensional and three dimensional ultrasonography showing bifurcated endometrial canals. There is gestational sac in the right moiety. There is no myometrial dip in the fundal region.

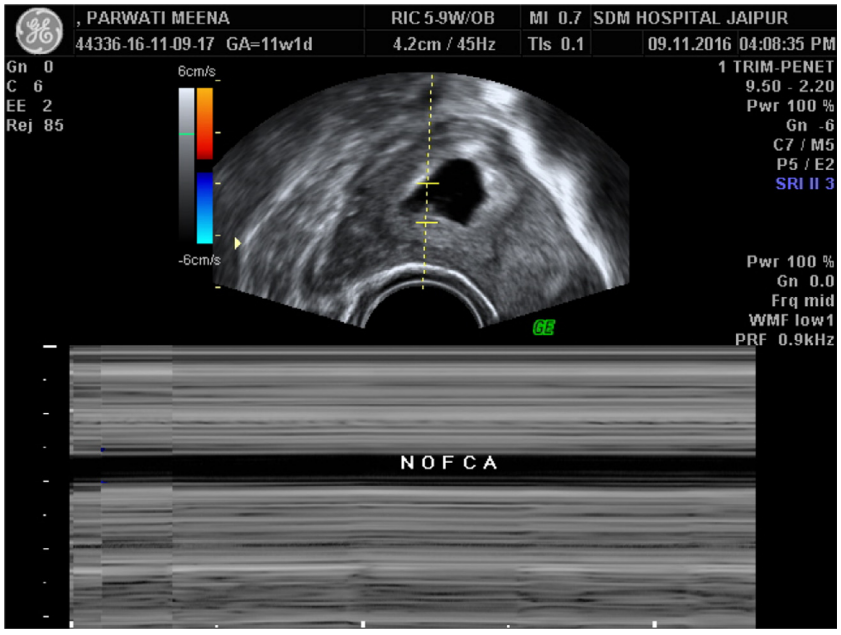

Fig. 2. Ultrasonography scan showing absence of foetal cardiac activity.

echogenic septum was seen separating the vagina into two cavities. Both ovaries and fallopian tubes were normal. Bilateral kidneys, ureters and urinary bladder were normal (Figs. 1–5). We advised MRI pelvis for better differentiation but patient was poor and could not afford it, so MRI scan was not done.

Patient was operated the very next day and the septum was removed on hysteroscopy. Thus our sonography findings were confirmed preoperatively.

3. Discussion

Septate uterus with double cervix and longitudinal vaginal septum is a rare anomaly. This anomaly is not explained by the present accepted theory of development of female genital tract which states that mullerian fusion is unidirectional caudal to cranially [3]. The theory has been questioned many times in the past by the reports of various rare anomalies [2,4]. In the case of septate uterus with cervical duplication and longitudinal vaginal septum, the anomaly suggested a failure in the fusion of the distal Mullerian ducts. In the presence of uterus and vaginal septum, the cervical duplication cannot be explained by the unidirectional theory.

In 1994, McBean and Brumsted, proposed a new embryological theory that the fusion and resorption of ducts proceed in both directions, cranial and caudal [5]. The embryological events are not clear yet and require further investigation.

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