

Case Report

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Bilateral Inguinal Hernia Containing a Rudimentary Uteri, Ovaries and Tubes in a Woman with Primary Amenorrhea

Francisco C Medeiros^{1*}, Francisco E Vasconcelos², Bruno HM Dias² and Renato MLV Leal³

¹Department of Maternal and Child Health, School Maternity Assis Chateaubriand, Brazil

²Federal University of Ceara (UFC), Fortaleza-CE, Brazil

³General surgeon and oncologic surgery, School Maternity Assis Chateaubriand, Brazil

Abstract

In this case report, we show a case of 21-year-old woman who presented amenorrhea, a blind vagina, a bilateral Nuck channel herniation. After initial examination, it was thought that she had the Testicular Feminization Syndrome. After follow-up, it was observed that she presented uterovaginal agenesis and ovary and uterine tubes hernia of the Nuck channel.

Keywords: Amenorrhea; Blind vagina; Nuck channel

Introduction

The processes vaginalis of the peritoneum accompanies the testis or round ligament through the inguinal canal into the scrotum or the labium major. In women, the homologous to this structure is called the Nuck channel. It usually undergoes obliteration after the seventh month of pregnancy, which explains the high incidence of hernias in premature kids [1]. It is possible, though, that the closure only happens after birth, during the first year of life. The failure of the closure of this canal can cause a hydrocele or a hernia [1]. There's an association between a hydrocele of the Nuck channel and a contralateral inguinal hernia [1], which should make us consider the existence of hydrocele in patients with a inguinal cystic mass and a history of inguinal hernia on the other side [2].

Considering the association between hydrocele and hernia of the Nuck channel, it is important to distinguish one from another. While a hernia can present bowel sound over an inguinal swelling and a bulging that is evident while standing, but disappears while lying on supine position [1], a hydrocele is a painless translucent swelling in the inguino-labial region, without any sound over it [1]. The diagnosis can also be established by ultrasonography or MRI [1].

Congenital vaginal agenesis is frequently present in patients with Mayer-Rokitansky-Kuster-Hauser Syndrome and Complete Androgen Insensitivity Syndrome (CAIS) [3,4]. Woman with Rokitansky syndrome have a 46, XX karyotype and normal functioning ovaries, while patients with CAIS present a 46, XY karyotype and testicular gonads [4]. Both conditions present primary amenorrhea [4]. The treatment for these cases usually consists of lengthening the vagina and, in CAIS, preceding a gonadectomy [4]. Since most of these cases present during adolescence, the patient should be included in the decision of treatment and psychological support must be required [4,5].

We present a case of a 21-year-old woman was presented to the Gynecology Department presenting primary amenorrhea and a blind vagina (Mayer-Rokitansky-Küster-Hauser Syndrome). Physical examination revealed Tanner V, thelarche and adrenarche, a 1-2 cm vaginal dimple (Figure 1), and a suspected bilateral inguinal hernia (During examination, when asked to stand up, it was observed a bulging of both labia majora (Figure 2). The patient's karyotype was 46, XX. Pelvic ultrasound showed that no ovaries, uterus, or vagina could be seen. Inguinal ultrasound showed a complex mass bilaterally at the inguinal areas (Figure 3). This could suggest the presence of testicles or of a hernia. Considering the amenorrhea, the blind vagina and the bulging, it was thought that the patient presented the Complete

Androgen Insensitivity Syndrome, which is usually associated with these finding, although this is not seen in females with 46, XX vaginal agenesis [3]. Therefore, a laparotomy surgery was indicated.

During the surgery, though, it was observed that the bulging was being caused by the presence of a ovary and uterine tubes hernia of the Nuck channel (Figure 4). In a woman presenting the Mayer-Rokitansky-Kuster-Hauser syndrome and herniation of the ovaries, the hernia sac must be opened, in order to release the ovaries into the abdomen and prevent infarction and maintain hormonal function [3]. After noticing the hernia, the correction procedure was executed and the lengthening of the vagina was realized.

Discussion

In this case, the patient presented an association of the Mayer-Rokitansky-Küster-Hauser (MRKH) Syndrome and a hernia of the Nuck channel. Considering the pathophysiology of the hernias and hydrocele of the aforementioned channel (failure of closure of the channel) [1] and the malformation represented by the syndrome [5], it is not illogic to question if there is a correlation between their genesis.

Inguinal hernias in phenotypical females with vaginal agenesis are usually seen with androgen insensitivity syndrome, but they have rarely been reported in patients with 46, XX vaginal agenesis [6-9]. Patients with Mayer-Rokitansky-Küster-Hauser syndrome and herniation of ovaries should have their ovaries reduced into the abdomen to prevent infarction and preserve hormonal function. Incarcerated Müllerian remnants are hormonally sensitive and may produce sporadic pain. When imaging patients who have vaginal agenesis and inguinal pain consideration should be given to viewing the inguinal canals for possible Müllerian remnants or ovaries. Etiology of Müllerian anomalies is not known; 92% of anomalies will have normal 46, XX karyotype, and most of the defects are likely to be related to polygenic and multifactorial causes [10]. Many anomalies are usually suspected based upon history and presenting symptoms. Ultrasound (US) examination is very helpful

***Corresponding author:** Francisco das Chagas Medeiros, Department of Maternal and Child Health, School Maternity Assis Chateaubriand, Brazil, Tel: (55-85)3366-8062; Fax: (55 85)3366-8065; E-mail: prof.fcmedeiros@ufc.br

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in diagnosis, but magnetic resonance image (MRI) has been recognized as the gold standard means. However, as it was the case in our patient, MRI may not be able to identify a rudimentary uterine horn, if it is



Figure 1: Vulvo-vaginal image of physical exam confirming vaginal agenesis.



Figure 2: Bulging of the labia majora and groin bilaterally after the Valsalva's maneuver.

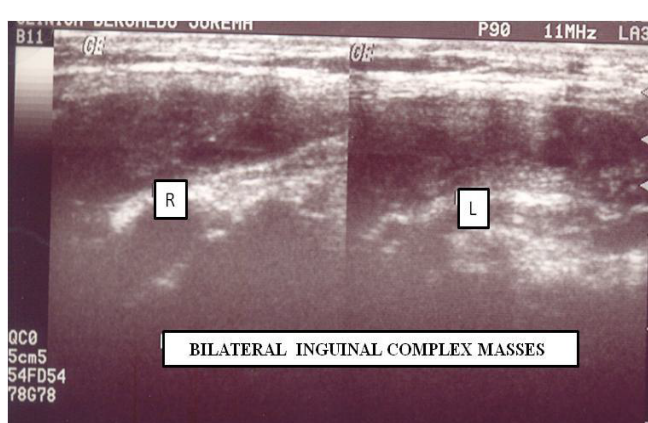


Figure 3. Inguinal Ultrasound showing both bilateral inguinal complex masses.

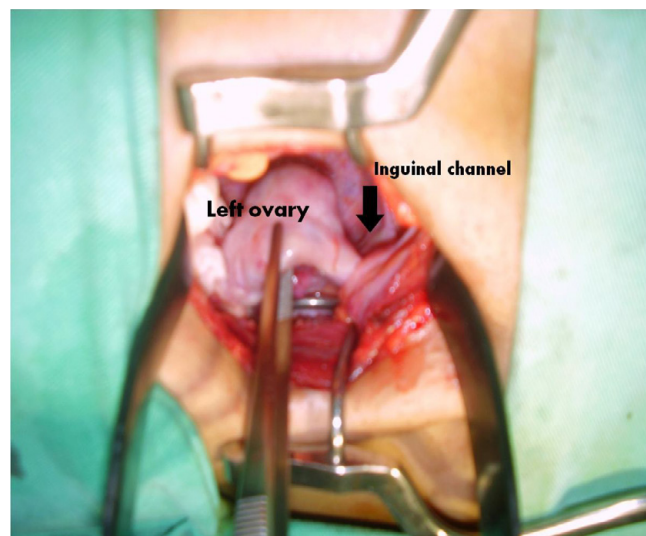


Figure 4: A view of the surgical procedure where it may be seen the left ovary and inguinal ring.

located laterally along the psoas muscle and pelvic sidewall [11]. Primary malformation here can be understood as Mayer-Rokitansky-Kuster-Hauser syndrome or its variants. Herniation of malformed uteri can be attributed to the imperfect closure of congenital openings and failure of fusion of Müllerian ducts, which remained attached in the original position of the inguinal fold. Ten cases have previously been reported until now [12,13].

Oppelt et al., in a study with 284 women showed an important correlation between MRKH syndrome and malformations of the renal system, for example [5]. Therefore, our goal, besides reporting a rare case is to interrogate a possible relationship between the two malformations. In order to explore this, more studies are needed.

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