UTERUS DIDELPHYS WITH OBSTRUCTED HEMIVAGINA AND IPSILATERAL RENAL AGENESIS (A Case Report)

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KEYWORDS :

Introduction

Congenital malformations of the uterus and vagina are interesting clinical entities. The mal-developments occur in a variety of forms and each anomaly is distinctive. Abnormalities of the reproductive tract are often associated with anomalies of the urinary tract, highlighting that the development of the Mullerian and Wolffian systems is intricately intertwined [1]. The unique clinical syndrome of uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis is very rare but well recognized. A patient of this syndrome is presented here.

Case Report

A 18-year-old girl presented with pain lower abdomen and foul smelling vaginal discharge of 3 years duration. To begin with, the pain was associated with periods. Gradually, it assumed a chronic dull character with exacerbation during periods. She was particularly distressed with foul smelling discharge which had made her a social outcast. She attained menarche at the age of 15 years. Her menstrual cycles were regular and the flow was scanty.

On examination, she was poorly nourished. On gynaecological examination, there was foul smelling purulent discharge in the vagina. A small cervix was seen to the right of the vagina vault. There was no discharge from the cervix. Per vaginum examination revealed two tender lumps, one above the other, located to the left of midline. Since the source of vaginal discharge could not be localised, the cervix was probed with a fine karman cannula. This manoeuvre did not bring out any discharge. Ultrasonography confirmed two cystic structures on left side of the pelvis. Intravenous urography showed the absence of left kidney and outlined the course of right ureter. From the foregoing, a clinical diagnosis of uterus didelphys with obstructed cervix and pyometra was made.

Exploratory laparotomy showed double uterus with a healthy hemiuterus, tube and ovary on the right side. The left hemiuterus, however, was bulky with a shaggy surface. The left tube was edematous and dilated and was exuding pus. It became obvious that the patient's symptoms were caused by the chronically infected left sided genital organs. In view of these findings, a left hemihysterectomy with left salpingectomy was performed leaving behind a healthy right hemiuterus with its tube. A thick fibrous

union existed between the two hemiuteri which was divided. The walls of the obstructed vagina were extremely thick, posing difficulty in excision. The excised uterus and vagina were full of foul smelling pus. Post-operative period was uneventful. She was asymptomatic during a follow up visit at 2 months and had established normal periods. Keeping in view that she had only one kidney, she was counselled to seek prompt treatment for any urinary infections.

Discussion

The clinical syndrome consisting of a double uterus, obstructed hemivagina and ipsilateral renal agenesis is rare. The exact cause of this anomaly is not known. In this syndrome, renal agenesis on the side of obstructed vagina associated with double uterus is suggestive of an embryologic arrest at 8 weeks of pregnancy [2].

There are three diagnostic groups of this syndrome depending upon the anatomical findings [3]. In the first group, there is complete vaginal obstruction with hematocolpos. These patients present with progressive severe dysmenorrhoea and paravaginal mass. In the second group, there is incomplete vaginal obstruction. These patient present with intermittent foul smelling discharge and lower abdominal pain. Paravaginal mass is inconsistent. In the third group, there is a lateral communication between the two uteri [4]. Here, the patient presents with foul smelling discharge. Our patient belonged to the second group.

Treatment of such cases is aimed at relief of symptoms and preservation of fertility. If detected early, excision of the vaginal septum is curative. When the diagnosis is delayed, there is extensive damage to the obstructed side due to infective process. In such situations, a hemihysterectomy is performed.

Interestingly, Rook and Jones reported that renal agenesis is the rule when Mullerian obstruction ac-

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companies failure of lateral fusion. It is seldom seen in cases of double uterus without vaginal obstruction [3].

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