e - ISSN - 2349 - 8005

INTERNATIONAL JOURNAL OF ADVANCES IN CASE REPORTS

Journal homepage: www.mcmed.us/journal/ijacr

INCIDENCE OF OBSTRUCTED HEMIVAGINA WITH IPSILATERAL RENAL AGENESIS AND PANCREATIC HEAD CYST IN AN INFERTILE WOMAN

Vadakkan Ouseph Thimothy* and Sunandakumari

Department of General Surgery, Amala Institute of Medical Sciences, Amala Nagar, Thrissur-680 555, Kerala, India.

Corresponding Author:- **V O Thimothy**
E-mail: vpthimothy@yahoo.com

Article Info
Received 22/04/2015
Revised 30/04/2015
Accepted 06/05/2015

Key words:
OHVIRA syndrome, Mullerian anomaly, Obstructed vagina, Pancreatic cyst.

ABSTRACT
Obstructed Hemivagina with Ipsilateral Renal Agenesis (OHVIRA) syndrome is an uncommon congenital anomaly rarely manifested. In this article, we present a case of OHVIRA along with a chocolate cyst and a cystic lesion in pancreas in a 31 year old female. The patient underwent laparoscopic pancreaticoduodenectomy, total abdominal hysterectomy plus right salpingoophorectomy. During the follow-up no recurrence of symptoms with good performance status were found.

INTRODUCTION

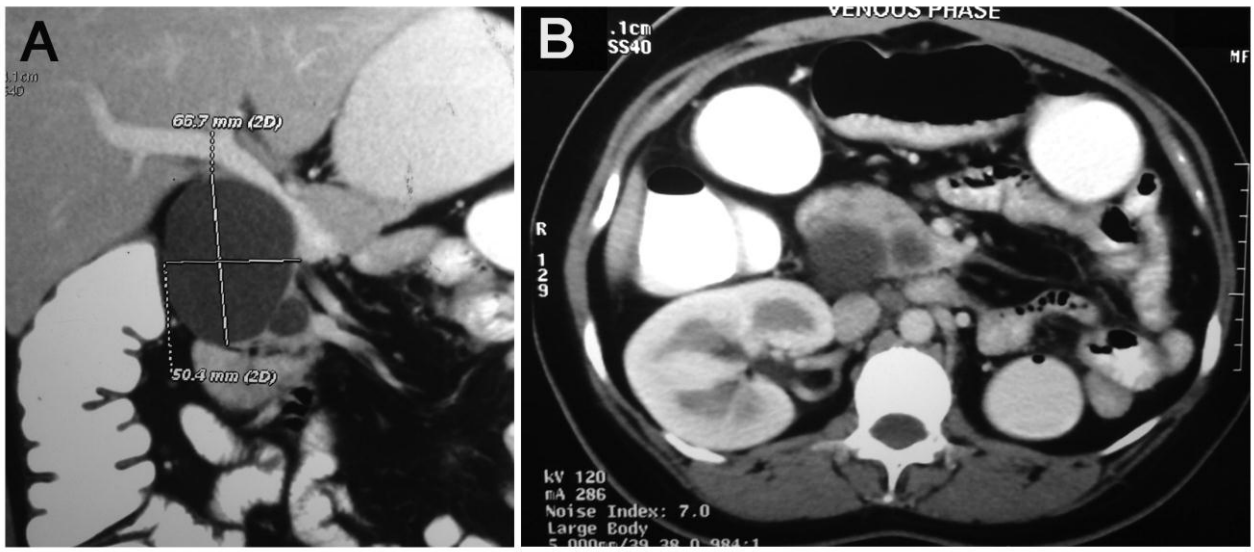
Obstructed Hemivagina with Ipsilateral Renal Agenesis (OHVIRA) syndrome is an uncommon congenital anomaly. The OHVIRA is also known as Herlyn-Werner-Wunderlich syndrome. The complex anolamy consists of uterus didelphys, unilateral low vaginal obstruction and ipsilateral renal agenesis, all 3 components being secondary to mesonephric duct-induced müllerian anomalies. Early correct diagnosis and surgical intervention are suggested to relieve the symptoms and for a good prognosis. In this case report, we present OHVIRA in a 31- year- old female who presented with abdominal pain with multiple episodes of vomiting.

CASE REPORT

A 31- year- old female presented with abdominal non radiating pain of 1 day duration in the epigastrium with multiple episodes of non bilious and non blood stained vomiting. History revealed that she has regular periods 5/32-33 days cycle dysmenorrhea, married 8 years back, nulliparous and IVF attempted. Patient was a known case of uterus diadelphys with haematocolpos, haematometra and haematosalpinx. She had excision of cervical septum and left salpingectomy for endmtriosis. On radiological investigation, the patient had agensis of left kidney. Trans-abdominal ultrasonogram (USG) showed a cystic lesion in head of the pancreas 5x4.1 cm, no features of pancreatitis, calcification and pancreatic duct dilatation. Inflammatory changes were found within mesentery near right ovary tortuous, tubular non compressible structure of 7.4 mm diameter in right iliac fossa close to the right ovary. Uterus was anteverted, bicornuate and septate. Left lateral wall showed an exophytic fibroid 3.1 x 3.1 cm and myometrium was normal. Endometrial thickness was 7.9 mm. Agenesis of left kidney and compensatory hypertrophy of right kidney 12.8x5.9 cm were found. Anterior margin of umbilicus showed two cystic lesions 8x8 and 7.9x5 mm in subcutaneous plane. Computed tomogram scan confirmed pancreatic cyst of 67.5 x 50.4 mm size (Figure 1A) and agenesis of left kidney (Figure 1B). A diagnosis of OHIVRA syndrome was made. In view of elevated tumour marker (CA 19-9: 3017 U/ml) and recurrent pain Pancreatico duodenectomy, total abdominal

hysterectomy plus right salpingoophorectomy were also done. Laparotomy findings were endometriosis, chocolate cyst involving umblicus (previous laparoscopic port site) thick walled cystic lesion involving head of pancreas posteriorly wedged between Inferior venacava (IVC) and portal vein. No infiltration to IVC/ portal vein was found. Right ovary enlarged with 8x10 cm chocolate cyst with flimsy bowel adhesions, uterus enlarged to 6-8 weeks with small left lateral wall fibroid uterine anatomy distorted due to previous unification procedure. Left fallopian tube and left ovary appeared normal as well as left cervix appeared redundant. Histopathological analysis of hysterectomy specimen showed leiomyoma and endometriotic cyst of right ovary, whereas that of pancreaticoduodenectomy specimen showed simple cyst of pancreas, portal vein nodes and peripancreatic nodes unremarkable. The surgical outcome was satisfactory. Patient has been on follow-up for last 2 years with no recurrence of symptoms.

Fig 1. Computed tomogram showing A) pancreatic cyst and B) Agenesis of left kidney



DISCUSSION

The syndrome of OHVIRA usually presented with uterus didelphys, unilateral low vaginal obstruction and ipsilateral renal agenesis. These 3 components were found to be secondary to mesonephric duct-induced müllerian anomalies. The true incidence of mullerian defects was found to be about 1.1 - 3.5% [1]. But in women with recurrent miscarriages and sub fertility, the incident is believed to around 25% [2]. A strong association of renal agenesis with uterus didelphys has also been suggested [3]. Women with diadelphic uterus may be asymptomatic and unaware of having a double uterus. They may present with complaints of dysmenorrhoea and dyspareunia. The patient in this case report has dysmenorrhoea and history of diadelphys with haematocolpos, haematometra and haematosalpinx.

According to embryological concepts proposed by Acien, the uterus and cervix were derived from fused paired paramesonephric ducts (2nd part) and divergent distal paramesonephric ducts (3rd part). Although lining of vagina revealed mullerian cells derived from mullerian tubercle, it was completely of mesonephric (wolffian) origin and hence the paramesonephric ducts do not contribute to formation of the vagina [4]. Duplication of the uterus results from the lack of fusion of the paramesonephric ducts in a local area or throughout their normal line of fusion. In uterus didelphys, individual horns are fully developed, normal in size with two cervices inevitably present. Each hemiuteri is associated with one fallopian tube. Ovarian malposition may also be present. A longitudinal or transverse vaginal septum may be noted. The vagina may be single or double. Duplicated vagina is more likely in a didelphic uterus. In this patient uterus didelphys was evident.

Renal agenesis most commonly occurs in association with uterine didelphys than with any other type of mullein anomaly. The reported incidence of renal anomalies in this group is 20% [5]. In our patient, agenesis of left kidney manifested that may probably cause hypertrophy of right kidney. Surgical intervention in the form of excision of vaginal septum to relieve obstruction and the associated pain has been suggested. In addition to this, surgery also reduces chances of pelvic endometriosis due to retrograde menstrual seeding. Most of the patients are able to have normal sexual life and if the obstructed uterus is preserved, they may even able to conceive and carry pregnancy to term. Altchek and Paciuc reported successful pregnancy in a young girl after surgery for the obstructed uterus in a uterus didelphys with unilateral distal vaginal agenesis and ipsilateral renal agenesis [6]

The complex anomalies can be diagnosed correctly with the help of imaging particularly with the

help of Magnetic resonance imaging [7]. This anomaly is most commonly manifested in adolescents with dysmenorrhea of progressive severity, abdominal pain and a pelvic mass [8]. Therefore, a woman who has ipsilateral renal agenesis with a pelvic mass, the diagnosis of OHVIRA should be suspected. Pelvic ultrasound, physical examination and laparoscopic findings established the diagnosis of hematometracolpos secondary to uterus didelphys with unilateral imperforate hemivagina [9]. Any association of pancreatic cyst with OHVIRA did not reveal by review of literature. In this case while grossly the pancreatic cyst appeared to be a chocolate cyst, endometrial tissue could not be demonstrated in the lining of the cyst by histopathological analysis. Therefore, we propose that the pancreatic cyst could be an association or outcome of this syndrome and further cases need to be analyzed in order to evaluate such a positive association.

This case report concluded that Obstructed Hemi-vagina with Ipsilateral Renal Agenesis syndrome may present with apparent gastrointestinal symptoms. Therefore, early correct diagnosis is inevitable for the surgical intervention to relieve obstruction and further prevention of complications.

ACKNOWLEDGEMENT

Authors gratefully acknowledge the valuable help of Dr. Ajith TA, professor, Department of Biochemistry and Dr. Joju Sebastian, Department of General Surgery, Amala Institute of Medical Sciences, Thrissur, Kerala, India during the preparation of the manuscript.

DECLARATION OF INTEREST STATEMENT

The authors declare that they have no significant competing financial, professional or personal interests that might have influenced the performance or presentation of the work described in this manuscript.

REFERENCES

1. Strassmann EO. (1996). Fertility and unification of double uterus. *Fertil Steril*, 17,165–76.
2. Acien P. (1997). Incidence of Müllerian defects in fertile and infertile women. *Hum Reprod*, 12, 1372–6.
3. Li S, Qayyum A, Coakley FV, Hricak H. (2000). Association of renal agenesis and mullerian duct anomalies. *J Comput Assist Tomogr*, 24, 829–34.
4. Acién P. (1992). Embryological observations on the female genital tract. *Hum Reprod*. 7, 437–45.
5. Golan A, Langer R, Bukovsky I, Caspi E. (1989). Congenital anomalies of the müllerian system. *Fertil Steril*, 51, 747-755.
6. Altchek A, Paciuc J. (2009). Successful pregnancy following surgery in the obstructed uterus in a uterus didelphys with unilateral distal vaginal agenesis and ipsilateral renal agenesis: Case report and literature review. *J Pediatr Adolesc Gynecol*, 22, 159–62.
7. Asha B, Manila K. (2008). An unusual presentation of uterus didelphys with obstructed hemivagina with ipsilateral renal agenesis. *Fertil Steril*, 90, 849, 9-10.
8. Morgan MA, Thurnau GR, Smith ML. (1987).Uterus didelphys with unilateral hematocolpos, ipsilateral renal agenesis and menses. A case report and literature review. *J Reprod Med*, 32, 47-58.
9. Pieroni C, Rosenfeld DL, Mokrzycki ML. (2001). Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. A case report. *J Reprod Med*, 46, 133-6.