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Experience with neonatal hydrometrocolpos in the Niger Delta area of Nigeria: Upsurge or increased recognition?

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ABSTRACT

Background: Hydrometrocolpos (HMC) is a clinical condition in which there is a cystic distension of the vagina (hydrocolpos), uterus and sometimes, fallopian tubes (hydrosalpinx) with fluid. This study described our experience with cases of HMC seen in our practice, and highlighted the flare in our practice. **Patients and Methods:** A retrospective study of cases of HMC managed at the University of Port Harcourt Teaching Hospital between September 2010 and August 2012. **Results:** There were seven cases; their ages ranged from 2 to 27 days (median 13 days). All the patients had abdominal distension but abdominal mass was obvious only in four. Other features varied depending on the presence of sepsis or other associated anomalies. The diagnosis of HMC was missed in all cases by the referring clinicians. One patient was referred with the diagnosis of tracheoesophageal fistula and esophageal atresia, 2 with bladder outlet obstruction, 1 with intestinal obstruction, 2 with anorectal malformation, and 1 with neonatal sepsis. Ultrasound identified bulky uterus in two cases and upper urinary tract dilatation in 3 patients. Six patients had laparotomy, 1 had hymenotomy only. Postoperative complications were basically wound sepsis and rectovaginal fistula resulted. **Conclusion:** Diagnosis of HMC should be considered as a differential in newborn girls presenting with lower abdominal mass. Attention to clinical detail is necessary to avoid a misdiagnosis.

Key words: Hydrometrocolpos, Girls, Diagnosis, Management

INTRODUCTION

Hydrometrocolpos (HMC) is a clinical condition in which there is a cystic distension of the vagina (hydrocolpos), uterus and sometimes, fallopian tubes (hydrosalpinx) with fluid. It is thought to arise from accumulation of secretions in the vagina and uterus due to excessive stimulation of the fetal cervical mucous glands by maternal estrogen in the presence of an atretic vagina or imperforate hymen.[1,2] Such congenital obstructions of the female genital tract have been long recognized.[3] Conversely, neonatal HMC is a rare condition despite the fact that imperforate hymen is the most common congenital anomaly of the female genital tract.[4] This suggests that imperforate hymen on its own does not explain the development of HMC. Imperforate hymen most usually passes unrecognized in neonatal life, being identified later due to associated problems or their complications.[5] The aetiological basis for development of HMC in the neonate is therefore yet unclear. There had not been reported cases of HMC in the Niger Delta region of Nigeria; but we have observed a sudden increase in hospital incidence in the past 2 years. Reports of HMC from other regions of Nigeria have not indicated such high incidence within a relatively short period.[6,7] We reviewed our experiences with HMC, highlighted the difficulties of diagnosing and managing unusual cases in an environment with limited facilities.

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PATIENTS AND METHODS

Data were retrospectively extracted from the notes of cases of HMC managed by the authors between September 2010 and August 2012. Data obtained were place of abode of mother, age at presentation, presenting features, associated anomalies, investigation findings, findings at surgery, and outcome. Data were subjected to a simple descriptive analysis with the Microsoft Office Excel 2007.

RESULTS

There were seven cases of HMC during the period under review. The ages of patients at presentation ranged from 2 to 27 days (median 13 days). Four of the mothers lived in oil producing communities of Rivers and Bayelsa states of Nigeria. At presentation, all the patients had abdominal distension, and abdominal mass in four. Other features included sepsis or other associated anomalies [Table 1]. The diagnosis of HMC was missed in all cases by the referring clinicians. One patient was referred with the diagnosis of tracheoesophageal fistula and esophageal atresia, 2 with bladder outlet obstruction, 1 with intestinal obstruction, 2 with anorectal malformation, and 1 with neonatal sepsis. Ultrasound identified the abdominal mass as the uterus in two cases and upper urinary tract dilatation in 3 patients. Intravenous urography was not done but plain abdominal X-ray identified distended bowel loops in 4 patients. Six patients had laparotomy, while one had hymenotomy only [Table 2]. The appearances of the pelvic mass were typically, hydrocolpos [Figures 1-3], and uterine didelphys [Figure 3]. The fluid was milky white in all cases and volume estimated 50–250 ml [Figure 4]. A biopsy of the vaginal wall in Case 1 was normal [Figure 5]. Biopsy in the subsequent cases was considered unnecessary. There was one partial wound dehiscence, which healed before discharge. A rectovaginal fistula resulted while we tried to do a vaginoplasty (Case 7), then necessitating a sigmoid colostomy. The fistula has since been repaired and patient is presently waiting for colostomy closure. There were two deaths due to respiratory failure (Case 3) and severe neonatal sepsis (Case 6).

DISCUSSION

Neonatal HMC is known to result from excessive fetal cervical secretion caused by maternal estradiol in the presence of a congenital obstruction of the genital

Table 1: The main features in the patients

Cases	Age (days)	Referral diagnosis	Main physical signs	Main investigation findings	Associated anomaly
1	27	Urinary retention? Cause	Visible peristalsis, abdominal mass, imperforate bulgy hymen, empty rectum and palpable mass in front of rectum	US – Bilateral hydronephrosis, pelvic mass? Ovarian teratoma Abdominal X-ray – Distended bowel loops Serum creatinine – Normal	Postaxial polydactyly
2	15	High ARM	Tachypnoea, tense abdominal distension, jaundice	Cross table lateral X-ray – High ARM US not done	Persistent cloaca
3	2	TEF and EA	Excessive salivation, choking on feeds, abdominal distension abdominal mass	Nonpassage of NG tube, blind upper esophageal pouch US – Abdominal mass? Bladder tumor	TEF and EA
4	3	Imperforate anus	Absent anus, abdominal distension, superficial veins, bulgy hymen	X-ray – Distended loops of bowel, bilateral US – Hydronephrosis	Low ARM
5	13	Intestinal obstruction? Volvulus neonatorum	Abdominal distension, abdominal mass, visible peristalsis, patent anus, empty rectum	Abdominal X-ray – Distended loops of bowel US – Fluid filled uterus	None
6	5	Neonatal sepsis? Intestinal obstruction	Petechial skin bleeds, distended superficial abdominal veins, shiny abdominal skin, abdominal distension, lobulated abdominal mass, oedematous labia majora, absent anus	US not done Plain X-ray – Distended bowel loops	Postaxial polydactyly
7	17	Bladder outlet obstruction	Suprapubic fullness, visible peristaltic waves, edematous vulva, bulgy hymen	US – Enlarged uterus attached to a cystic mass Bilateral hydronephrosis X-ray not done	None

ARM: Anorectal malformation, NG: Nasogastric, EA: Esophageal atresia, TEF: Tracheo-esophageal fistula, US: Ultrasound

Table 2: Operative findings, treatment, and outcome

Case	Operative findings	Final diagnosis	Treatment given	Outcome
1	Distended uterus and vagina; normal but stretched fallopian tubes; imperforate hymen	HMC; imperforate hymen; obstructive uropathy	Posterior colpotomy + drainage of 200 ml + hymenotomy	Recovered
2	Bilobed uterus, both hemiuteri and vagina distended, vaginal atresia, 250 ml of milk white fluid drained	HMC; uterine didelphys; persistent cloaca	Anterior colpotomy + colostomy	Recovered. awaiting PSARVUP
3	Blind upper esophageal pouch; fistula to lower esophageal pouch, globular uterus	TEF and EA; HMC; uterine didelphys vaginal atresia	Esophagostomy gastrostomy, anterior colpotomy	Died
4	Distended vagina, normal uterus and fallopian tubes	Hydrocolpos; distal vaginal atresia	Anterior colpotomy + vaginoplasty + colostomy + anoplasty	Recovered
5	Imperforate bulgy hymen; thick milky effluence	HMC; imperforate hymen	Hymenotomy	Recovered
6	Distended uterus and vagina; normal but stretched fallopian tubes; imperforate hymen	HMC; imperforate hymen	Posterior colpotomy; hymenotomy	Died (severe sepsis)
7	Distended uterus and vagina. No vaginal orifice	HMC; obstructive uropathy; vaginal septum	Vaginoplasty; drainage; colostomy	Recovered rectovaginal fistula

HMC: Hydrometrocolpos, EA: Esophageal atresia, TEF: Tracheoesophageal fistula, PSARVUP: Posterior sagittal anorectovaginourethroplasty

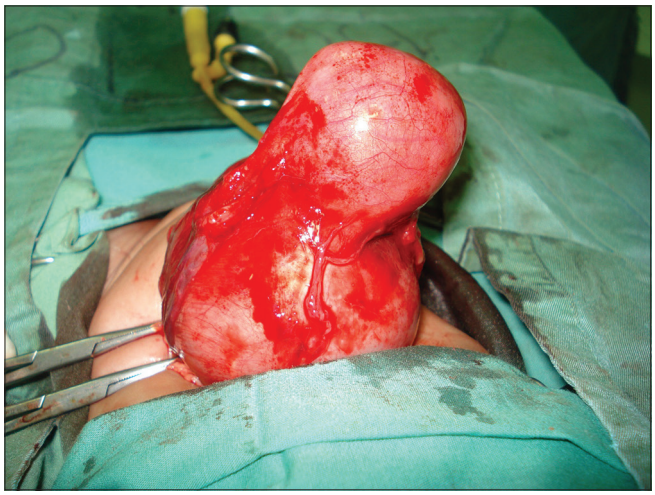

Figure 1: Hydrometrocolpos

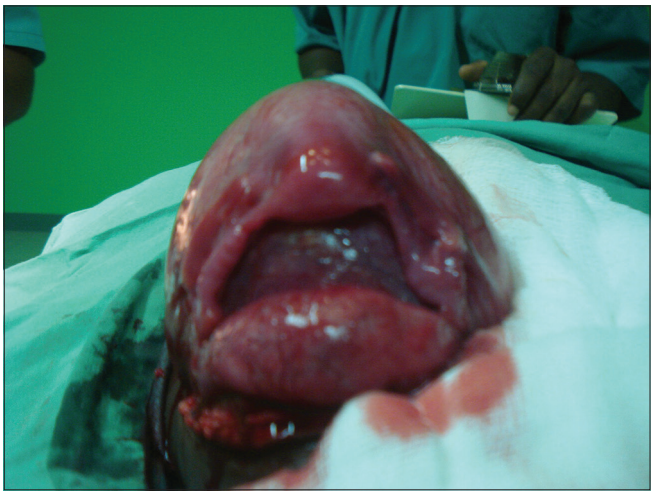

Figure 2: Hydrocolpos as seen in Case 4

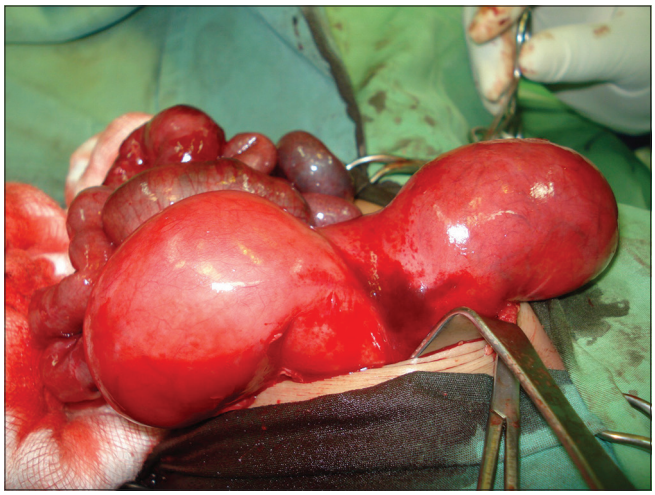

Figure 3: HMC as seen in Case 2

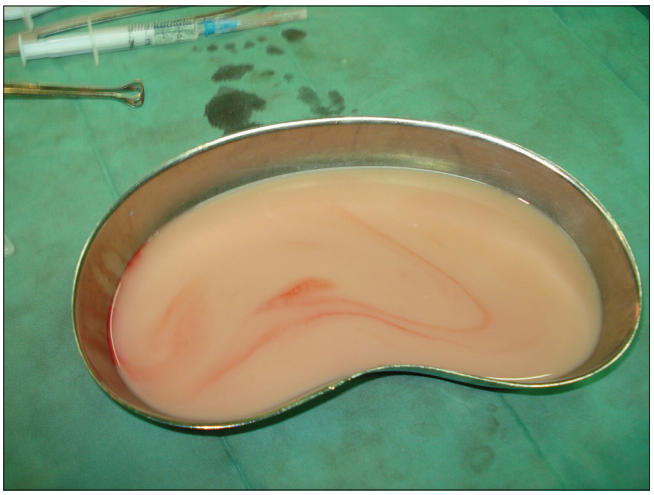

Figure 4: Milky fluid content of HMC

tract.[2,8] Imperforate hymen being the most common congenital anomaly of the female genital tract, it would be expected that HMC by extension, should be common as well. The rarity of HMC therefore suggests that there are some other factors that determine its manifestation and whether it will manifest in the neonatal period or

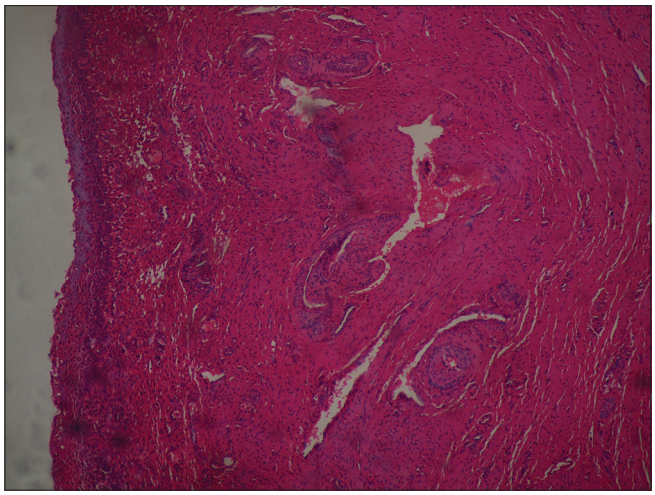

Figure 5: Normal vaginal wall

later at puberty. Seven cases of neonatal HMC in 2 years in an environment where it had been almost unknown even in older children and adults elicit significant interest. The fact that all the referring clinicians did not pick up the right diagnosis suggests that this upsurge may not be due to increased awareness of the condition. It also highlights the low index of suspicion and paucity of awareness of this uncommon condition even among specialists. Difficulty with diagnosis has been identified in many reports even in advanced countries.[9,10] This apparent low index of suspicion among caregivers in our environment indicates that there may be more cases than are seen in our service. In one report, three of the nine patients were only diagnosed intraoperatively.[5] In the only case reported from southeastern Nigeria, diagnosis was also made at surgery.[6] In our series, only three of the seven cases were diagnosed correctly preoperatively. Low index of suspicion for uncommon conditions and the variable presentations of patients are among the reasons adduced for the common misdiagnosis.[8] In our series, most of the patients presented with some other congenital anomalies or complications that tended to obscure the HMC. This indicates that irrespective of the problems any female neonate may present with, HMC must be specifically looked out for. Whereas an earlier report from Northern Nigeria included 9 patients seen over a period of 15 years, 7 patients seen in 2 years is a call for increased awareness. This is particularly in view of the serious urinary and gastrointestinal complications that arise from the obstructive effects of HMC.[4,11,12] Upper urinary tract damage as a complication makes prenatal diagnosis desirous. Whereas none of our cases were diagnosed prenatally, there are increasing reports from other parts of the world of the possibility and usefulness of prenatal diagnosis in ameliorating or forestalling the complications.[13-16] The typical features of neonatal HMC are a mass in the hypogastrum and a bulging of whitish tissue between the labia and behind the urethra. This hymenal bulge is absent when there is vaginal atresia or a urogenital sinus. Associated anomalies or complications often take the attention of the clinician away from the HMC. This was the situation in three of our patients who had tracheoesophageal fistula and esophageal atresia, persistent cloaca, and imperforate anus, respectively. Two of our patients who had postaxial polydactily could have been cases of the recognized associated syndromes such as Bardet Biedl syndrome, Mckusick-Kaufman syndrome but we have not confirmed this conclusively as one died 2 days postoperatively and the other is still being followed up.[17-19] A high index of suspicion is required in this environment where majority of newborns do not have the benefit of a scrupulous assessment by a neonatologist soon after birth. In our patients, the ultrasonographer mistook the pelvic mass for the urinary bladder. Imaging features of HMC have been described by researchers to aid recognition of HMC.[20,21] Two of the cases who had laparotomy and who had imperforate hymen could have benefitted from simple hymenotomy but for lack of clarity of the pathological anatomy before the surgery. In addition, a rectovaginal fistula, which resulted following vaginoplasty, is a mishap that could have been avoided if the pathology were properly delineated. A finger or catheter balloon inserted in the rectum in the course of the dissection could have guided the surgeon and possibly avoided the rectal injury. Proper delineation would certainly give a guide to a better quality of treatment. Some authors have made a case for needle aspiration under reliable ultrasound or magnetic resonance imaging guidance. We think that this mode of treatment may predispose to infection of the HMC cavity, incomplete drainage and recurrence, and bleeding into the cavity. It may only be useful in very ill patients and in circumstances where neonatal anesthesia is considerably risky.

In all the flare, we have experienced of HMC awakens the consciousness of the occurrence of this condition in our region and prepares us to recognize it early and treat appropriately. As we watch the trend in its incidence in our region and other regions with similar circumstances, effort should be made to increase the awareness of the occurrence of HMC among caregivers at the primary and secondary levels of the health-care system.

CONCLUSION

Neonatal HMC has been encountered in our practice with relative high incidence in the past 2 years. Most of

the babies are of mothers who live in the oil-producing communities of the Niger Delta region of Nigeria. The awareness and the experience to properly diagnose and manage it are still limited in our practice and some other regions of the world with similar circumstances. There is the need to create the awareness of this condition among all levels of health-care givers. There is a need for a closer look at the possibility that environmental factors may be contributory to the development of neonatal HMC.

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Conflicts of interest
There are no conflicts of interest.

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