Case Report

Congenital hydrometrocolpos: A diagnostic dilemma

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Abstract. Congenital hydrometrocolpos accounts for 15% of intrabdominal cystic masses in female neonates. The clinical features of hydrometrocolpos in newborn period are dominated by the lower abdominal mass with compression of adjacent structures. We present a case of congenital hydrometrocolpos of a 2 month old female child with an antenatal diagnosis of intra abdominal cyst, presented with lower abdominal distension since birth along with possible surgical management.

Key words: Hydrometrocolpos, Abdominal mass, Vaginostomy

Introduction

Hydrometrocolpos in neonatal period is a rare condition caused by congenital vaginal obstruction. The clinical features in newborn period are dominated by the lower abdominal mass with compression of adjacent structures. Preoperative diagnosis is difficult due to rarity of disease. This case report highlights the need for a thorough evaluation of a new born with abdominal mass to arrive at early diagnosis and appropriate treatment.

Case History

Our case was a 2 month female child with an antenatal diagnosis of intra abdominal cyst, presented with chief complaints of lower abdominal distension since birth along with decreased urine output for last 12 days. Child was admitted and examined under general anaesthesia and panendoscopy. On examination 16 x14 cm suprapubic mass arising from pelvis, firm in consistency, smooth surface and well defined margins was found (Fig 1a). Perineal examination showed single visible orifice (Fig.1b), deep vestibule and poorly developed labia. Ultrasound of abdomen showed heterogeneous cystic lesion with internal echoes behind the bladder (Fig.2a) and magnetic resonance imaging (MRI) of abdomen showed fluid filled elongated lesion behind the bladder (Fig.2b). Endoscopic findings revealed urogenital sinus with 2cm common channel, stenotic vaginal orifice at 5 o’clock position with evidence of urethritis and cystitis. Intraoperatively both vaginal and bladder openings were catheterised and dye (iohexol) study showed tubular distended vagina with rat tail tapering (Fig.2c) and small capacity bladder with bilateral vesicoureteric reflux (Fig.2d). Perineal vaginostomy was attempted but procedure abandoned due to high insertion of vagina into the sinus and thick inflamed tissue, hence mini laprotomy with tube vaginostomy was performed. Intraoperative findings were 12x8 cm pyometrocolpos with inflamed and thick walled vagina displacing bladder to right side (Fig.3a). About 150 ml pus aspirated (Fig.3b). Vagina mobilised completely and U shaped flap taken from anterior surface of vagina (Fig.3c) and tubularised into a tube and brought out as tube vaginostomy (Fig.3d). Postoperative period was uneventful. Child is on regular follow up and waiting for definitive surgical procedure.

Fig. 1 (A) Clinical examination showing suprapubic mass arising from pelvis with well defined margins (red dotted line) (B) Perineal examination showed single visible orifice (arrow), deep vestibule and poorly developed labia

Fig. 2 (A) Ultrasound abdomen showing heterogeneus cystic lesion with internal echoes behind the bladder (B) MRI abdomen showing fluid filled elongated lesion behind the bladder

Fig. 3 (A) Operative photograph showing pyometrocolpos with inflamed and thickened vagina displacing bladder to right side (B) Vagina mobilised and U shaped flap taken from anterior surface (C) Vagina tubularised into a tube and brought out as tube vaginostomy (D) Postoperative period was uneventful
DISCUSSION

Hydrometrocolpos can be defined as the dilatation of the female genital tract with fluid, which is usually caused by the combination of cervical and vaginal secretion and sometimes with accumulation of urine. Reported incidence is 1:16000 female births. Congenital hydrometrocolpos accounts for 15% of intrabdominal cystic masses in female neonates. The most common cause of hydrometrocolpos is imperforate hymen, other causes include septate or membranous obstructions to the distal vagina. The commonest type of fluid that accumulates is mucus followed by urine. Etiologically hydrometrocolpos can be classified as secretory or urinary. Anatomically hydrometrocolpos can be classified into five types based on types and level of obstruction (Table 1). Presentations in hydrometrocolpos can be early or delayed depending on the obstruction. Children with partial obstruction have a delayed presentation whereas children with total obstruction present early in neonatal period. Usually neonates present with a lower midline mass with or without hymenal protrusion. Sometimes effect of mass compressing adjacent structures can lead to obstructive uropathy, respiratory distress, and acute abdomen with paralytic ileus. Reported association with hydrometrocolpos includes McKusick-Kaufman syndrome, congenital heart disease, urinary and gastrointestinal abnormalities, Mullerian dysgenesis syndrome and severe hydrops. Work up in these children includes complete clinical examination of abdomen, perineum and rectum preferably under general anesthesia followed by radiological assessment by X-Ray abdomen, ultrasonography of abdomen, retrograde urethrogram. Endoscopic evaluation is to be done before proceeding for any surgical intervention. Historically various surgical procedures has been described for hydrometrocolpos. The principles of management are prompt drainage of collection followed by single stage or
multistage procedure depending upon the type of anomaly encountered. The definitive treatment varies depending upon anomaly and ranges from hymenectomy to total urogenital sinus mobilisation (TUM) and abdominoperineal vaginal and rectal pull through. \[9\] Definitive treatment for type I is hymenectomy while abdominoperineal repair is indicated in type II. In type III and IV abdominoperineal vaginal pull through is indicated. In type V temporary drainage by tube vaginostomy should be done followed by TUM if the length of the common channel is less than 3 cm or pull through via the posterior sagittal anorectoplasty route and vaginal substitution with bowel in the same sitting if the length of common channel is more than 3 cm. \[10\] Temporary diversion in form of tube vaginostomy has several advantages over external drainage tubes which include, better drainage of fluid and urine, easy nursing care, no indwelling catheter, lesser risk of infection. This catheterizable conduit can be used for contrast studies to delineated anatomy. In cases of urinary type of hydrometrocolpos, vesicostomy or external diversion by suprapubic cystostomy may be added for better drainage of bladder and to prevent urinary stasis and infection.

<table>
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<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>1</td>
<td>Low hymenal obstruction with protruding imperforate hymen</td>
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<tr>
<td>2</td>
<td>Mid-plane transverse membrane or septum in vagina</td>
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| 3    | a) High obstruction with distal vaginal atresia  
      b) High obstruction with distal vaginal atresia and gluteal swelling |
| 4    | Vaginal atresia with persistence of the urogenital sinus |
| 5    | Vaginal atresia with cloacal anomaly |

**Table 1. Types of hydrometrocolpos[4]**

## Conclusion

Hydrometrocolpos is a rare entity in neonatal age group. Any female child with a lower abdominal mass should have a thorough examination of perineum to look for the number of orifices and if it is less than three openings i.e. urethral, vaginal and anal then examination under anaesthesia and panendoscopy is mandatory to delineate the exact anatomy. Tube vaginostomy is a novel drainage technique to tide over the crisis till the definitive surgery is planned.

## References