RECTAL ATRESIA WITH COMPLETE ABSENCE OF POSTERIOR VAGINAL WALL (LARGE RECTOVAGINAL FISTULA): A CASE REPORT

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ABSTRACT

A case report of congenital anorectal malformation in 21 years old female patient is presented who had history of passing urine and stools per vaginum since birth. Examination under anesthesia revealed absence of anal orifice. Ultrasonography reported a big gas filled mass in the lower abdomen. Barium contrast study was inconclusive. The diagnosis made was high anorectal malformation with big recto vaginal fistula.

Key words: *Imperforate Anus, High Rectovaginal Fistula, Pull Through Procedure, Neo-anus.*

INTRODUCTION

Congenital anorectal malformations are not uncommon. These may be grouped together are called as " imperforate anus". One infant in 4500 is born with this anomaly. The abnormality is divided into two groups [1]. Low abnormalities like covered anus, ectopic anus, stenosed anus and membranous stenosis.[2] High abnormalities like anorectal agenesis, rectal atresis and common cloaca.

Congenital anorectal malformations are not uncommon. Most of these patients are managed by paediatric surgeons. These anomalies are more common in females with male to female ratio of 1:1.5.1. In our setup some of these children may be neglected till they reach adult life. A case of 21 years old female patient is reported here who seems to have been neglected with a high anorectal malformation.

CASE REPORT

A 21 years old female patient was seen in OPD on 27th March 2003 with history of passing urine and stools per vaginum since birth. She had been to many specialists and local hospitals in Peshawar several times but to no avail. According to her parents she was normally delivered at home and took normal feed. Her growth was some what retarded (exact measurement were not available at the time). She also gave history of constipation and use laxatives to get her bowels moved. No menarche. On examination soft distension was noted in the lower abdomen. However no masses were there on clinical check up. In view of her history and female gender she was admitted for examination under anaesthesia.

At examination under anesthesia (EUA) on 06.04.2003 she was found to have no anal orifice. Faeces were seen in the vagina and urethral orifice could be identified separately at its normal site. At this point the question was " what could be done about her to have normal anatomical lower end of the GIT, so that she could pass the stools via normal anal orifice.

A cascade of investigations was started. Routine blood test like Hb, TLC, DLC, Urea, Sugar, Electrolytes, LFT's were all normal. X-ray chest was normal. X-ray abdomen revealed scoliosis of moderate degree. IVP was normal. Ultrasonography reported a big gas filled mass in the lower abdomen. Barium contrast study was requested after discussion with radiologist. This was inconclusive as the contrast would be lost in a big cavity with out any anatomical landmarks. Barium follow through was also inconclusive. The most likely diagnosis made was high anorectal malformation with big recto vaginal fistula. Her parents and the patient herself were counseled for initial diverting transverse loop colostomy. She was admitted and after manually evacuating barium from the cavity a colostomy was done in right upper quadrant an 10.04.03 she went home alright and was readmitted an 17.05.03 for exploratory Laparotomy. She was catheterized pre

operatively. After thorough preparation she under went surgery on 12.06.2003. At operation a huge cystic mass of rectum and sigmoid was identified lying in the pelvis with a large communication with the posterior vaginal wall. This was a challenging situation, haw to separate vagina from rectum and how to give her continent anal orifice. The structures were dissected as down in the pelvis as possible. It was impossible to separate vaginal vault completely. So rectum was transected at the recto sigmoid junction. Distal end (cuff) was closed and so was incorporated into vagina. The proximal stump was pulled down behind the distal stump as far down as possible. A vertical incision about 2 3 inches was made into posterior wall of the distal stump and the proximal stump was pulled down through this hole down to the perineum. At the perineum a neo-anal orifice was fashioned. The proximal stump of the recto sigmoid was pulled inside the distal stump sleeve towards this neo-orifice and anastamosed with the transverse perineal muscles.

A pelvic drain was put in and abdominal wound was closed in layers. While perineal wound was dressed with a tulle gauze. She went home on 18.06.03. On 22.08.03 she was readmitted to assess the sphincter function. By now the wounds had healed. Her post up (check) Ultrasound done on 03.06.03, no more reported big cystic mass in pelvis (as was seen on 27.03.03 prior to surgery). Although this time the Ultrasonographer reported that she has a small rudimentary uterus with enlarged right ovary while left ovary was not seen. A retrograde barium enema was done to see the result of the operation. It showed almost normal lumen and length of the large gut. Perineum was healthy. A lubricated flatus tube was passed peranally and rectum filled with 200 ml of normal saline. She was asked to walk and do not allow the saline to flow out of her rectum. She could hold saline for two hours. The procedure was repeated three times. Her diverting transverse colostomy was closed on 28.8.03. Postoperatively she had uneventful recovery and went home on 01.09.03. Since she has been regularly followed up in OPD.

Last follow up was done on 09.03.05. She was symptom free and was moving her bowels normally.

DISCUSSION

In the developed world these congenital abnormalities are diagnosed in neonatal life. In our set up patient may present late in adult life when they themselves notice that some thing is wrong. Most of our babies are born at home by TBA's (Traditional Birth Attendants) who are not aware of such anomalies. A female child in the family also is not welcomed; therefore this case was neglected till she reached to the age of 21 years when she told her mother that she is passing stools and urine via a single passage.

This was a case of anorectal agenesis, as we saw a blind rectal pouch with a fistula in to the posterior wall of vagina.[1]

The problem in this case was “to achieve reconstruction with continence of flatus and faeces”. Recent work by Pena and de vries has shown that the muscles of continence are present in all cases of imperforate anus. Keeping this in mind the case was managed.[2]

Rectal atresis is a rare anomaly. It may be associated with other congenital anomalies like cardiac malformation.[3] Lumbosacral vertebral abnormalities are a common association of anorectal malformation and are one of the determinant of the eventual level of fecal continence that may be achieved.[4] Anorectal agensis with recto urethral fistula has been reported in a 48 years old man from Japan.[5] Uterovaginal malformations may occur with increased frequency in children with anorectal malformation. In this regard a case has been reported in the literature about a girl in whom there was total absence of vagina in association with anorectal malformation. This was diagnosed at the time of surgery for the condition.[6]

Authors from Northern India have recognized risk factors in the management of anorectal malformations. In babies with isolated high anorectal malformations, weighing more than 2.5 kg and brought to hospital with in first 48 hrs of life, the survival has been reported as 100%.[7] In this case the patient came to OPD at the age of 21 years.

It has been reported from Nigeria that anorectal malformations are more common in female than male, (a ratio of 1.5:1). It has also been observed over there that teenage mothers in rural communities has been effected and so like my patient the surgeon seems to be dealing with adult population in that country.[8] It has been reported that in adults after they had under gone surgery, psychological functioning had the most important effect on the quality of life while limitation in role functioning due to physical activity was not a serious problem.[9] In adults the results of surgery regarding fecal continence are not too good after conventional pull through operations. Therefore artificial bowel sphincter and gracilis neosphincter are used to achieve maximum benefit.[10]

CONCLUSION

In the developing world, patients may still

be seen in their adult life with congenital anorectal malformations. Surgeons may perform restorative procedures with the hope that continenence might be achieved.

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