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Short Clinical Report

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A case of Aphallia associated with neonatal intestinal obstruction

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CASE PRESENTATION

A preterm newborn weighing 2 kgs, received 7 hours after birth with penile agenesis associated with greenish vomiting. The antenatal history of the mother was uncomplicated. On examination, the baby had a sick general state with respiratory distress and abdominal distension; Absence of the penis and normal scrotum with normally descended testes were also evident (Fig. 1A). The anal opening was normal with a squirt of urine passed through the anus (the urethra opened on the anterior anal canal near the anal verge). Full blood count and renal state were abnormal. The imaging studies were done which includes an echocardiography assessment that revealed atrial septal defects, ventricular septal defects, and abdominal ultrasound which revealed bilateral hydroureteronephrosis with thick free fluid and debris. In addition, an abdominal x-ray showed multiple air-fluid levels and peritoneal calcifications (Fig. 1B).

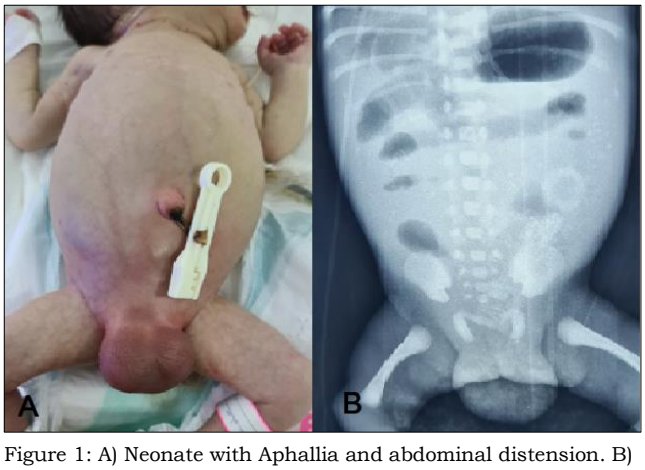

Figure 1: A) Neonate with Aphallia and abdominal distension. B) X-ray showing multiple air-fluid levels.

After 8 hours of aggressive resuscitation and operative preparation, a laparotomy was done. The intraoperative finding showed intra-peritoneal meconium with distal ileal atresia that led to ileal perforation; the urinary bladder was mildly distended. After peritoneal lavage, segmental resection of about 5 cm of distal ileum (a segment that involved atresia and perforation) was done and double barrel ileostomy was formed. No vesicostomy was done because the neonate was clinically unwell and in distress. In addition, the urinary bladder was mildly distended, and a squirt of urine passed through the anus.

Postoperatively the baby was admitted to the respiratory care unit with endotracheal intubation and monitoring, but the neonate succumbed to sepsis on 3rd postoperative day.

DISCUSSION

The embryology of this rare disorder has been described because of non-formation of the genital tubercle or failure in development into a phallus. [1, 2] Up to 100 cases of aphallia have been reported to date. [2-4]

In 1984, Skoog et al. classified this anomaly into three types: pre-sphincteric (28%)- there is recto-urethral (prostatorectal) fistula; post-sphincteric (60%)- the urethra opens any region on the perineum or near the anterior anal verge, over the pubis or anterior scrotum; and urethral atresia with recto-vesical fistula. [5, 6] Our case had a post-sphincteric type lesion.

The classical aphallia includes penile agenesis, 46 XY male karyotype, a normal scrotum, and descended testicles. [5] Talebpour Amiri et al. [5] during the review of three cases of aphallia found that the urethra was short leading to the distal rectum. In this instance, we report penile agenesis with the urethral opening on the anterior anal canal. In addition, the physical examination revealed abdominal distension, normal scrotum, testicular descent, and normal anal opening. Penile agenesis should be differentiated from intrauterine penile amputation, micropenis, concealed penis, rudimentary penis, severe hypospadias, and epispadias. [7]

Penile agenesis may be accompanied by other common malformations including genitourinary, cardiac, and gastrointestinal tract defects. [5] Chakraborty et al. [2] found solitary penile agenesis. The present case is unique in that the patient presented with complex malformations including atrial septal defects, ventricular septal defects, bilateral hydroureteronephrosis, and distal ileal atresia with perforation.

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REFERENCES

1. Demirer Z, Aip BF, Uguz S, Guragac A, Irkilata HC. A rare case of penis agenesis (Aphallia) with associated multiple urogenital anomalies. Int J Surg Case Rep. 2015; 15:10-2.
2. Chakraborty M, Chakraborty S. Penile agenesis-an extremely rare urogenital anomaly. Int J Cur Res Rev. 2018; 10:31.
3. Kane AD, Ngom G, Ndour O, Alumeti DM. Aphallia: A case report and literature review. Afr J Paediatr Surg. 2011; 8:324.
4. Moghtaderi M, Boroomand M, Kajbafzadeh A, Arshadi H, Ghohestani M, Mehdizadeh M. A rare case of Aphallia. J Ped Nephrol. 2016; 4:74-7.
5. Amiri FT, Ghabaee DN, Naeimi RA, Seyedi SJ, Mousavi SA. Aphallia: Report of three cases and literature review. Int J Reprod BioMed. 2016; 14:279.
6. Berhane M, Abera G, Alemu S, Eshetu B. Aphalia: An extremely rare congenital genitourinary malformation-A case report. Ethiopian J Health Sci. 2019; 29:409-12.
7. Qiang S, Li FY, Zhou Y, Yuan Y, Li Q. Congenital absence of the penis (aphallia): A rare case report. Medicine. 2019; 98: e15129.