Patent vitellointestinal duct with inverted ileal loop prolapse: A rare presentation

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The vitellointestinal duct (VID) connects the yolk sac to the gut in the developing embryo and provides nutrition until the placenta is established; the duct attenuates and separates from the intestine between the 5th and 7th weeks of gestation. Failure of obliteration of the embryonic VID leads to various congenital anomalies like – Meckel's diverticulum, vitelline cord, enteric cyst, umbilical sinus, patent VID with ileal intussusception prolapsing over the umbilicus. The most common reported anomaly of residual vitelline duct is Meckel's diverticulum with different presentations like bleeding, intestinal obstruction or diverticulitis. However, the patent VID with inverted ileal loop prolapse is a rarely reported. We have encountered three patients of this rare presentation who were managed successfully.

Key words: Ileal, intussusception, patent vitellointestinal duct, prolapse

INTRODUCTION

The vitellointestinal duct (VID) (omphalomesenteric duct) connects the yolk sac to the gut in the developing embryo and provides nutrition until the placenta is established; the duct attenuates and separates from the intestine between the 5th and 7th weeks of gestation. Failure of obliteration of the embryonic VID leads to various congenital anomalies like – Meckel's diverticulum, vitelline cord, enteric cyst, umbilical sinus, enteric fistula with ileal intussusception prolapsing over the umbilicus or hemorrhagic umbilical mass. The VID anomalies occur in approximately 2% of newborns and in 6% of these the duct remains patent, with 20% of patent omphalomesenteric duct cases being complicated by intussusception of the small bowel through the patent duct. This condition is 8 times more common in males, and 73% of these cases exhibit symptoms within the first 28 days of life. Another significant complication is progressive prolapsus of the omphalomesenteric duct, leading to a T-shaped bowel protrusion through the umbilicus (with a characteristic ram's horn appearance). Although Meckel's diverticulum is the most common vitelline duct anomaly seen in 2% of population, a patent vitelline duct with intussusception of small bowel is a very rare entity to be reported in world literature and only few cases have been reported. This anomaly needs to be managed urgently for the fear of gangrene of the intussucpted bowel. The principle of surgical treatment is to reduce the intussucpted gut and complete excision of the vitelline duct with restoring the ileal continuity as well as umbilical reconstruction. We have managed three cases of this rare anomaly and in one of the patients the presentation was typical ram's horn type appearance.

CASE REPORTS

Case 1

A 40-days old male child admitted with a history of a reddish mass coming out of the umbilicus for last 24 h. The baby was born by a full term normal vaginal delivery conducted by a trained medical personnel. There was a history of yellowish to green discharge from the umbilicus after which it was advised topical applications on the umbilical area by the attending pediatrician. On examination, there was a prolapse of a small intestine through the umbilicus giving a ram's horn appearance [Figure 1]. There was also cleft palate anomaly and baby was investigated for hematological as well as biochemical parameters. After resuscitation patient was operated via a transverse infra umbilical approach and the intussucpted small intestine was reduced manually [Figure 2]. At the enteric end of the patent VID
a wedge resection of the communication and primary anastomosis was done. Umbilical end of the fistula was approached from within and it was dissected from the umbilicus. Primary closure of the abdominal wound and umbilical reconstruction was done. Patient was discharged home after 1-week. Histopathological examination of the excised duct revealed the normal ileal mucosa.

**Case 2**
A 2-month-old female child was admitted with complains of prolapsing mass from the umbilicus. On examination, the small gut mucosa was visible, and it was coming out of the umbilicus [Figure 3]. After necessary investigations patient was operated via infraumbilical incision and the intussuscepted ileum was reduced, and surgical correction of the anomaly was done. Patient was discharged home in good condition, and histopathological examination of the excised duct revealed the normal ileal mucosa.

**Case 3**
A 2 weeks male baby was admitted with umbilical hernia and prolapsing gut through the center of the defect. On examination, the end of the prolapsed gut was gangrenous [Figure 4]. Patient was operated via transumbilical approach and surgical correction of the patent VID, which included the resection of the gangrenous portion of the ileum, as well as the repair of the umbilical hernia defect, was done. Patient recovered well.

**DISCUSSION**
Vitelline duct or omphalomesenteric duct anomalies are secondary to the persistence of the embryonic vitelline duct, which normally obliterates by weeks 5–9 of intrauterine life. These anomalies occur in approximately 2% of the population and may remain silent throughout life, or may present incidentally sometimes with an intraabdominal complication. In 6% of these duct remains patent and can either present as discharging umbilical sinus, umbilical nodule or polyp, bleeding from intestinal mucosa or even 1/5th (20%) of patent vitelline duct cases being complicated by intussusception of the small bowel through the patent duct. This condition is 8 times more common in males and in the present series two of three were males. Another significant complication is progressive
prolapsus of the omphalomesenteric duct, leading to a T-shaped bowel protrusion through the umbilicus and even ram’s horn type appearance which is also seen in one of case of present cases.[5,6]

It is difficult to explain the mechanism of ileal intussusception into the patent VID but two reasons have been hypothesized; wide mouth of patent VID and shorter distance between VID and ileocecal valve in infants leading to higher intraluminal pressure.[7,9] However, it is difficult to predict beforehand which one of the patent VID cases will develop this complication.

The diagnosis of a patent vitelline duct malformation is simply dependent on the history of type of discharge (fecal) from the umbilicus and physical examination. Investigations like fistulogram may only be done when there is need to differentiate patent VID and patent urachus,[10] but it is not necessary in presentations as was the present cases because it will not change the surgical decision.

The principle of surgical management is a reduction of the intussuscepted gut along with complete excision of the vitelline duct and restoring the ileal continuity as well as umbilical reconstruction.[2,4] There are three approaches described; infraumbilical, supraumbilical or through the umbilicus.[7] We have used the infraumbilical approach in the two cases and transumbilical approach in one patient for correction of this anomaly. However, the umbilical reconstruction could be done in only two cases because of local anatomy of the area.

To summarize, the authors conclude that patent VID with ileal intussusception and prolapse is a surgical emergency and the diagnosis is simply based on history and physical examination. Further diagnostic investigations like fistulogram are unnecessary as these do not add in making a surgical decision.

REFERENCES

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