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Congenital Duodenal Web in 1 Month Old Girl

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Abstract: ***Background: Congenital web of the gastrointestinal tract is a rare anomaly causing gastrointestinal obstruction in infants and children. It is a rare condition with approximately 100 cases reported in the literature with a reported incidence ranging from 1 in 10,000 to 1 in 40,000. The severity of symptoms and the age of onset vary according to the size and location of the web, and if the obstruction is total or partial. When the obstruction is incomplete, as it was in our case, the clinical symptoms are variable and the diagnosis can be difficult. Case Presentation: A one month old girl was hospitalized with history of vomiting since she was born. Frequency of vomiting about 2 to 3 episodes per day with volume was 3-5 spoons without bilious color. There were no history of fever, jaundice, bloating. Laboratory investigation showed hypokalemia and cholestasis. Babygram radiography revealed "single bubble appearance" in cavum abdomen that suggested to Hypertrophic Pylorus Stenosis. Upper gastrointestinal study showed partial stenosis in pylorus and distal bulbus duodenum, duodenal web can not be excluded and not supported to hypertrophic pylorus stenosis. Patient underwent an exploratory laparotomy, which revealed a partial obstruction caused by duodenal web hole type at duodenal pars II pre ampula vater then web excision and duodenoplasty with primary hecting was conducted. On the 21th days post operation, she was discharged from hospital in good condition.***

Keywords: duodenal web, congenital

1. Introduction

Congenital web of the gastrointestinal tract is a rare anomaly causing gastrointestinal obstruction in infants and children. It may develop at any site of the gastrointestinal tract, but is most commonly found in the stomach and small intestine. A congenital web in the intestine is a type of intestinal atresia includes duodenal, jejunal and ileal webs. The most common site of intestinal webs is the second portion of the duodenum. A duodenal web, which is one of the causes of duodenal obstruction, is a thin, round, partially permeable, web like structure.[1]

The duodenal web or duodenal diaphragm is a complete or incomplete obstruction of the duodenum resulting from the membranous web or intraluminal diverticula. In the duodenal web there is usually a small gap in the middle that distinguishes it from the duodenal atresia. It is a rare condition with approximately 100 cases reported in the literature with a reported incidence ranging from 1 in 10,000 to 1 in 40,000. The severity of symptoms and the age of onset vary according to the size and location of the web, and if the obstruction is total or partial. Therefore, the diagnosis may be delayed until late childhood or even to adulthood.[1],[2]

Duodenal web is a rare cause of duodenal obstruction found across all age groups, but most commonly seen in infants. These webs are thin, consisting of mucosa and submucosa, usually lacking a muscular layer. Peristaltic causes the diaphragm to balloon distally, resulting in the classic appearance of a wind sock. Duodenal webs are thought to arise from incomplete recanalization of the duodenal lumen during the fourth and fifth weeks of gestation. Presentation is typically early in life with evidence of proximal bowel obstruction, however, presentation late in adulthood has been reported. [3]

Duodenal web is type I of duodenal obstruction. By virtue of being a congenital disorder, duodenal webs are more commonly seen in infants and children. Such a web consists of a mucosa and submucosa but does not have a muscular layer. The second part of the duodenum is the most common site, representing 85% to 90% of all duodenal web cases. The third and fourth parts of the duodenum represent 20% and 10% of WD cases, respectively.[3],[4],[5],[6]

The characteristics of a gastrointestinal web may take the form of complete obstruction owing to an intact membrane or partial obstruction owing to a fenestrated membrane. The presenting symptoms for patients with any form of gastrointestinal web are consistent with bowel obstruction or partial obstruction, including vomiting (or typical presentation with bilious vomiting in an obstruction distal to the opening of the ampulla of Vater), abdominal distension and failure to pass meconium in instances of lower obstruction. Retrospective study by Sarin et al (2012), from 18 cases the commonest presentation of duodenal web was vomiting after feeding, which was bilious in majority of the children. Two patients had vomited as long as 6 months and 1 year before medical help was sought. Two patients presented with imperforate anus. More than 3/4 of children (76.9%) had weights >3 SDs below normal at the time of presentation.[1],[4]

Signs and symptoms suggesting a duodenal obstruction differ widely depending on the underlying defect. Between 30-59% of patients with proximal intestinal obstruction have a maternal history of polyhydramnios during pregnancy.

Vomiting shortly after initiation of feeding, feeding intolerance and failure to thrive are among the first symptoms babies present with duodenal web. Dehydration and changes in electrolytes are possible complications. In 85% of all cases, the obstruction is distal to the papilla of Vater, and therefore gastric aspirates and vomiting will be bilious. Abdominal distention, in contrast, is rare as the obstruction is proximal. [5]

In more than half of the patients with duodenal atresia, associated anomalies and syndromes are present. Down syndrome is present in 30% of cases, malrotation in 20%, and congenital heart diseases in 20% of cases, nevertheless, other congenital anomalies of alimentary tract are also. Kumar et al. showed congenital duodenal obstruction associated with anomalies were noted in 19/31 (61%) patients: 6 cases Down's syndrome, 5 cases anorectal malformation, 5 cases annular pancreas, 4 cases cardiac anomalies, 3 cases esophageal atresia with tracheoesophageal fistula. [7]

Plain X-rays and ultrasonography are often unremarkable. In the majority of cases, diagnosis can be established by plain abdominal radiography. Although the classical "double-bubble" appearance is pathognomonic of duodenal obstruction, symptoms may not be specific in patients with fenestrated duodenal web. A typical finding during upper gastrointestinal series is the so called "windsock sign" because of peristalsis of the proximal part of the gut resulting in a prolapse of the membrane into the distal part of the lumen.[5]

There are, in essence, two therapeutic options: surgery entailing resection of the obstruction followed by duodenoduodenostomy is well established. In some cases, endoscopic resection of duodenal webs has been described. However, this latter procedure is being discussed controversially. Supporters promote this procedure because of its minimally invasive character, opponents criticize the high risk of injury to the papilla of Vater. [5]

The objective of this case was to present a duodenal web causing duodenal obstruction. When the obstruction is incomplete, as it was in our case, the clinical symptoms are variable and the diagnosis can be difficult.

2. Case

A 1 month old girl Barat was admitted to hospital with history of vomiting since she was born. Frequency of vomiting about 2 to 3 episodes per day with volume was 3-5 spoons without bilious color. There were no history of fever, jaundice, bloating.

She was born in general hospital by caesarean section and vigorously at aterm gestational age. Birth weight was 3100 grams and 45 cm of length. Mother's age was 29 years old when she had the first pregnancy. Antenatal care was performed by an obstetrician every 3 month and antenatal ultrasound revealed within normal. There were no risk factors of infection during pregnancy and bad obstetric histories. There were no complains during pregnancy and no histories with other disease that associated with pregnancy. The patient was the first child and there were no history of miscarriage.

On physical examination the patient was alert. Respiratory rate was 42 times/minute with regular heart rate of 140 beats/minute. Her axillary temperature was 37^0 C. Examination of the head and face was normal shaped. There were no conjunctiva injection, anemia nor icteric sclera. The pupil light reflect was normal. The ear, nose, and throat examination were in normal limit. It was no cyanotic lips. The chest was symmetrical both on rest and movement, breath sound was bronchovesicular without rales or wheezing. The first and second heart sound was normal, regular, systolic murmur in upper left sternal border grade 3/IV. In the abdomen, bowel sound was normal. There were was no enlargement of liver and spleen. The extremities were normal. Patient's body weight was 2,5 kg and her body length was 55 cm, which was between the 0 and the 2nd percentiles for both weight and height for age according to the WHO growth charts.

Patient underwent several laboratory examination. Complete blood count revealed hemoglobin level was 10,25 /dL, hematocrit 31,07 %, leukocytes 26,23/mm^3, thrombocytes 307,80/mm3. Liver function were within normal limit, serum aspartate transaminase 18,2 U/L, alanine aminotransferase 7,40 U/L, albumin 2,9 g/dl, and alkali phosphatase 195 U/L, total bilirubin 6,48 mg/dL, direct bilirubin 3,19 mg/dL, indirect bilirubin 3,29 mg/dL. Electrolyte examination showed level of natrium 137 mmol/L and low level of kalium 1,9 mmol/L. Kidney function test revealed BUN 3,0 mg/dL and serum creatinine 0,35 mg/dL. Coagulation profile showed international normalized ratio (INR) of 1,46 with increased partial thromboplastin time (PTT) 17,0 (control 14,4) and normal level of activated partial thromboplastin time (APTT) 33,9 (control 36). Babygram radiography revealed "single bubble appearance" in cavum abdomen that suggested to Hypertrophic Pylorus Stenosis. Upper gastrointestinal study showed partial stenosis in pylorus and distal bulbus duodenum, duodenal web can not excluded and not supported to hypertrophic pylorus stenosis (Figure 2). Echocardiography examination revealed Moderate Atrial Septal Defect, Small Patent Ductus Arteriosus, Mild Mitral Regurgitation and did not need cardiac treatment.

The working diagnosis was Suspect Duodenal Obstruction, Moderate Atrial Septal Defect, Small Patent Ductus Arteriosus, Mild Mitral Regurgitation, Cholestasis Extrahepatic differential diagnose with Intrahepatic.
Patient underwent laparotomy exploration and web excision. A transverse incision at supra umbilical and abdominal exploration was conducted. Patient underwent exploratory laparotomy, which revealed a partial obstruction cause of duodenal web hole type at duodenal pars II pre ampula vater then web excision and duodenoplasty with primary hecting was conducted.

After surgery patient was admitted in the Pediatric Intensive Care Unit (PICU). The enteral feeding was postponed until next 2 days. After surgery laboratory examination was

performed, revealed: the leucocyte count was 27.85 K/uL with absolute neutrophil count 12.13 (43.54%), hemoglobin 9.84 g/dL, platelet count 488 K/uL. Based on this condition, addition of intravenous antibiotic therapy were Ceftriaxone 125 mg every 12 hours and Metronidazole 35 mg every 8 hours. Two side blood culture performed before antibiotic administration.

After 3 days after antibiotic administration evaluation was performed, revealed the leucocyte count was 16.81 K/uL with absolute neutrophil count 6.75 (40.18%), hemoglobin 9.35 g/dL, platelet count 378 K/uL, procalcitonin 0,37 ng/mL and antibiotic was continued. A weeks after that the leucocyte count was 13.78 K/uL with absolute neutrophil count 8.27 (60.0%), hemoglobin 8.15 g/dL, platelet count 336.50 K/uL, procalcitonin 1.15 ng/mL. Based on this condition antibiotic was changed to Cefepime 160 mg every 8 hours and Amikacin 60 mg every 24 hours.

On the second day after surgery, she was in good condition, no fever, no bloating and the bowel sound quite good. She tried to drink and well tolerated. And she was admitted to ward.

On the 21th days post operation, she was discharged from hospital in good condition. Patient was scheduled for follow up in one week.

3. Discussion

Congenital web of the gastrointestinal tract is a rare anomaly causing gastrointestinal obstruction in infants and children. It may develop at any site of the gastrointestinal tract, but is most commonly found in the stomach and small intestine. The most common site of intestinal webs is the second portion of the duodenum. It is a rare condition with approximately 100 cases reported in the literature with a reported incidence ranging from 1 in 10,000 to 1 in 40,000. Duodenal web is a rare cause of duodenal obstruction found across all age groups, but most commonly seen in infants.

The presenting symptoms for patients with any form of gastrointestinal web are consistent with bowel obstruction or partial obstruction, including vomiting (or typical presentation with bilious vomiting in an obstruction distal to the opening of the ampulla of Vater), abdominal distension, and failure to pass meconium in instances of lower obstruction. More than 3/4 of children (76.9%) had weights >3 SDs below normal at the time of presentation.[1,4] Signs and symptoms suggesting a duodenal obstruction differ widely depending on the underlying defect. Vomiting shortly after initiation of feeding, feeding intolerance and failure to thrive are among the first symptoms babies present with duodenal web. Dehydration and changes in electrolytes are possible complications. In 85% of all cases, the obstruction is distal to the papilla of Vater, and therefore gastric aspirates and vomiting will be bilious. Abdominal distention, in contrast, is rare as the obstruction is proximal.[5] In this case, patient with history of vomiting since born, volume of vomiting about 3-5 spoons without bilious color. Patient also have failure to thrive and electrolytes disturbance.

More than 50% of affected patients may be associated with other congenital anomalies, with Down syndrome, annular pancreas, congenital heart disease and malrotation being among the most common associated anomalies. Additionally, polyhydramnios presents in 33% to 50% of cases, 45% of which are premature. In this case, from physical examination patient found with systolic murmur in upper left sternal border grade 3/IV and echocardiography revealed atrial septal defect and patent ductus arteriosus.

In the majority of cases, diagnosis can be established by plain abdominal radiography. Although the classical “double-bubble” appearance is pathognomonic of duodenal obstruction, symptoms may not be specific in patients with fenestrated duodenal web. In this condition, the barium meal is the ideal procedure of choice for diagnosis. Moreover, in older children exible endoscopy can reveal the dilatation of the duodenum and the protrusion of the web in the lumen. In this case, babygram radiography revealed “single bubble appearance” in cavum abdomen that suggested to Hypertrophic Pylorus Stenosis. It is difficult to make a diagnosis preoperatively in cases with partial obstruction. For duodenal webs, diagnosis may be achieved in most instances of complete obstruction by plain abdominal radiography showing the characteristic double bubble representing the distended stomach and proximal duodenum, but no gas distal to this.

An intestinal web is a membranous structure within the lumen and consisting of the mucosa, submucosa and the incomplete vacuolization and recanalization of the duodenum. The resulting barrier may be complete, or less frequently may have a small orifice which forms a duodenal membrane with fenestration. The continuous peristaltic activity causes the membrane to prolapse distally, forming the wind sock sign. This wind sock is typical for the duodenal web in contrasted imaging. However in this case, the upper gastrointestinal study showed partial stenosis in pylorus and distal bulbus duodeni, duodenal web can not excluded and not supported to Hypertropic Pylorus Stenosis**.**

There are, in essence, two therapeutic options: surgery entailing resection of the obstruction followed by duodenoduodenostomy is well established. In some cases, endoscopic resection of duodenal webs has been described. However, this latter procedure is being discussed controversially. Supporters promote this procedure because of its minimally invasive character, opponents criticize the high risk of injury to the papilla of Vater.[5] In this case, patient underwent laparoscopic exploration and web excision. A transverse incision at supra umbilical and abdominal exploration was conducted. The patient underwent exploratory laparotomy, which revealed a partial obstruction cause of duodenal web hole type at duodenal pars II pre ampula vater then web excision and duodenoplasty with primary hecting was conducted.
Chen et al (2014) found postoperative parenteral nutritional support was achieved over a period of 9.32 ± 3.45 days. Patients started enteral feeding after after average of 8.5 ± 3.17 days and mean postoperative hospital stay was 14.87 ±

5.60 days. In this case, patient started the enteral feeding on the second day after surgery, he was in good condition, no fever, no bloating, and the bowel sound quite good. She tried to drink and well tolerated.

Sarin et al. found there were 4 deaths in the postoperative period giving a mortality rate of 22%. The causes of death were sepsis with refractory shock (n=2), aspiration (n=1), and necrotizing enterocolitis necessitating reexploration (n=1). Three out four deaths were seen in preterm babies. Only one of the 4 preterm babies in this series survived. In this case, patient was observed after discharged from hospital. She was doing well, tolerating her normal diet and had no vomiting episodes. She was vitally stable and her height was 50 cm and weighed 3,4 kg and (increased by 400 grams since discharge).

The mortality rate of duodenal obstruction has gradually decreased and the survival rate now exceeds 90%. The risk factors for death are complex cardiac anomalies, prematurity, sepsis, pneumonia, and surgical complications. Multiple congenital anomalies and sepsis are still the main risk factors for mortality, accounting for nearly 80% of all postoperative deaths. In this case, patient with congenital heart defect, but follow up observation showed was in good condition. These situation could be avoided or minimized if the parents paid more attention to neonatal vomiting and if the primary care physician fully recognized the disease. Although multiple congenital anomalies and prematurity are the major factors influencing the prognosis for duodenal obstruction, which is an established fact that can not be altered artificially. Beyond this prerequisite, we should put more emphasis on the importance of early diagnosis and urgent surgical treatment.

The parent concern about the possibility of having serious long term complication after surgery a journal search was conducted and showed “Duodenal Atresia and Stenosis: Long Term Follow Up Over 30 Years” from Journal of Pediatric Surgery 2004. The conclusion was late complications occur in 12% of patients with congenital duodenal anomalies, and the associated late mortality rate is 6%, which is low but not negligible. So, we could explain to the parent that the complication and mortality was low but we still need to follow up of the patient into adulthood to identify and address these late occurrences.[8]

4. Summary

A 1 month old girl was hospitalized with history of vomiting since born. Volume of vomiting about 2 to 3 episodes of vomiting per day, 3-5 spoons without bilious color. There were no history of fever, jaundice, bloating.

Laboratory investigation showed hypokalemia and cholestasis. Babygram radiography revealed “single bubble appearance” in cavum abdomen that suggested to Hypertrophic Pylorus Stenosis and upper gastrointestinal study showed partial stenosis in pylorus and distal bulbus duodeni, duodenal web can not excluded and not supported to hypertrophic pylorus stenosis**.**

Patient underwent exploratory laparotomy, which revealed a partial obstruction cause of duodenal web hole type at duodenal pars II pre ampula vater then web excision and duodenoplasty with primary hecting was conducted. On the 21th days post operation, she was discharged from hospital in good condition.

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