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Perinatal hemorrhage from ulceration of the umbilical cord: A potentially catastrophic association with duodenal and jejunal obstruction

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ABSTRACT

Purpose: The purpose of this study is to review published reports and contribute new cases of umbilical cord ulceration (UCU) with perinatal hemorrhage into the amniotic cavity in the setting of duodenal or jejunal obstruction because knowledge of this sequence is poorly disseminated and could be lifesaving.
Methods: Published reports of UCU with hemorrhage associated with congenital duodenal or jejunal obstruction were reviewed. Chart review was conducted for the cases encountered at our institutions between January 2008 and March 2017. We noted perinatal complications, method of delivery, gestational age, birth weight, gender, number, location, and pathologic description of umbilical cord ulcers, and outcome.
Results: Thirty-one reports and 7 new cases were studied. Perinatal complications included: preterm labor or preterm premature rupture of membranes: 63%; fetal distress: 95%; mean gestational age: 33 weeks; premature gestation: 95%; bloody amniotic fluid: 90%. Pathological analysis of UCUs revealed solitary, multifocal, helical and punched-out lesions. There were 12 neonatal deaths (32%), and 12 intrauterine deaths (32%). Survival rate was 37%.
Conclusions: UCU with perinatal hemorrhage is associated with duodenal and jejunal obstruction. Knowing the typical clinical signs of this potential catastrophic complication could prompt lifesaving delivery.
Type of study: Prognostic
Level of evidence: IV

In the era of prenatal diagnosis, the natural history of fetal malformations has become of increasing practical value. Successful management of pregnancy and delivery complicated by a fetal malformation requires knowledge of predictable occurrences, especially of those for which intervention could limit fetal morbidity and mortality. Umbilical cord ulceration (UCU) is associated with both duodenal and jejunal obstruction [1–17]. UCU can cause potentially life-threatening perinatal hemorrhage into the amniotic cavity, with potentially catastrophic fetal consequences. Intensive fetal surveillance and prompt delivery could be life-saving.

Because awareness of this association is not well disseminated, we offer this observational study. We have added 7 cases to the 31 reported cases in the English literature, and present a review of the same.

1. Methods

Our literature review was limited to English language citations. We utilized search terms “umbilical cord ulcer”, “umbilical cord erosion”, “umbilical cord rupture”, and “umbilical cord AND hemorrhage” in PubMed. Reports were excluded if the umbilical cord ulceration was not associated with duodenal or jejunal obstruction.

In addition, after obtaining IRB exemption, we reviewed clinical records from the newborn intensive care units and the pathology departments at our two institutions. From January 2008 to March 2017 we encountered 7 patients with duodenal or jejunal obstruction complicated by UCU with hemorrhage. The relevant clinical records of these 7 patients were studied.

In both the literature and our institutional records, fetal obstructions of the duodenum or jejunum were most often recognized by ultrasounds showing some combination of double bubbles, distended stomachs and distended loops of intestine with or without

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polyhydramnios. The diagnosis of duodenal or jejunal obstruction was confirmed postnatally or at autopsy. The diagnosis of umbilical cord ulceration with hemorrhage was established when bloody amniotic fluid was encountered at delivery with grossly evident erosions of the umbilical cord. Pathologic analyses of the placentas in these cases were reviewed.

From the previously published cases and the 7 we contribute here, we extracted descriptions of fetal sonograms including signs of intestinal obstruction and occasional umbilical cord abnormalities. We recorded the obstetrical presentation at the time of delivery including gestational age, perinatal complications, and method of delivery. We observed birth weight, gender, number, location and pathologic description of umbilical cord ulcerations, additional anomalies, site of intestinal obstruction, and outcome.

2. Results

2.1. Review of the literature and 7 additional cases

A total of 31 cases of duodenal or jejunal obstruction complicated by UCU and hemorrhage were identified in the literature. Congenital duodenal or jejunal obstruction was complicated by UCU and hemorrhage in 7 cases in our 2 institutions from January 2008 and March 2017. During this time period, we located 46 total cases of congenital duodenal or jejunal obstruction. This included 20 cases of duodenal atresia, 6 cases of duodenal web, 3 cases of duodenal stenosis, 15 cases of jejunal atresia, 1 case of jejunal duplication cyst, and 1 case of malrotation with congenital duodenal obstruction from Ladd's bands. Obstruction was complicated by UCU and hemorrhage in 7/46 cases, yielding an incidence of 15%.

The findings of the 31 published reports combined with our 7 cases are summarized in Table 1. Prenatal diagnosis of intestinal obstruction was made in 36/38 cases (95%). There were 17 cases of duodenal obstruction, 16 of jejunal obstruction, and 1 of concomitant duodenal and jejunal obstruction (two atresias). The site of obstruction was not specified in the remaining 4 reports. Of the 34 cases with reported antenatal detection of duodenal or jejunal obstruction, diagnosis was made by prenatal ultrasound at an average gestational age of 29 weeks (range 20–38). Polyhydramnios was observed in 23/38 cases (61%). The average gestational age at delivery was 33 weeks (range 26–38). Premature delivery, defined as <37 weeks' gestation, occurred in 36/38 of cases (95%). Gestational age was not specified in 1 case. Cesarean section was performed in 32/38 cases (84%). Four of the 5 vaginal deliveries occurred in patients with intrauterine fetal demise (IUFD). Initial obstetrical presentation with preterm labor and/or preterm premature rupture of membranes (PPROM) occurred in 24/38 cases (63%). Nonreassuring fetal heart tracings, fetal decelerations or bradycardia occurred in 36 of 38 cases (95%). Bloody amniotic fluid was noted at delivery in 34 of 38 cases (90%). Umbilical cord abnormalities were detected on prenatal ultrasound in 2 cases (5%). There were 27 females and 7 males (79% female). Gender was not specified in 4 cases. IUFD occurred in 12/38 (32%). Postnatal death occurred in 12/38 (32%), 11 of these in the neonatal period and 1 at ten months of age. Overall, 14 of 38 neonates survived, representing a survival rate of 37%.

2.2. Clinical pathology of 7 new cases

The 7 cases from our institutions included 4 with IUFD, 1 who died in the neonatal period, and 2 survivors. Pathological examination of the 7 cases from our institutions showed solitary ulcers in 4 and multiple ulcers in 3 of the umbilical cords. Ulcers ranged from 0.9 up to 5 cm in greatest dimension, either in a helical pattern (Fig. 1) or as punched-out lesions (Figs. 2b and 3a). Gross and histological examination in 6 of the cords revealed exposed umbilical vessels with either absent or attenuated overlying Wharton's jelly. Rupture of an umbilical vessel with overlying deep ulceration of Wharton's jelly was noted in 2 of these specimens (Fig. 3b), and pigment-laden macrophages infiltrating Wharton's jelly were evident in 4 of these specimens. Intestinal obstruction occurred distal to the ampulla of Vater in 6 patients as evidenced by pathology report or the presence of bile in gastric aspirate. The site of obstruction could not be determined in 1 patient.

2.3. Case presentation

We present case 38 as representative of a characteristic clinical course for duodenal and/or jejunal obstruction complicated by UCU and hemorrhage.

A 27 year-old primigravida was admitted to the antenatal high-risk service at Lenox Hill Hospital at 33-1/7 weeks of gestation owing to preterm premature rupture of membranes (PPROM). Her prenatal care was uneventful until 32 weeks of gestation when routine ultrasound detected a markedly distended fetal stomach with a second distended cystic structure ("double bubble" sign) and an apparent narrowing of the duodenum between the stomach and the second cyst (Fig. 2a). In addition, polyhydramnios was detected with an amniotic fluid volume of 34.5 cm. No other structural malformations were detected. The mother reported normal first trimester screen and a normal anatomical survey done by a previous provider. She was counseled regarding the suspicion for duodenal obstruction, the need for postdelivery surgery, and association with aneuploidy. Given the advanced gestational age, NIPT (noninvasive prenatal testing) was performed (which was normal). Upon admission to the hospital she displayed no signs of infection, and fetal heart rate monitoring was reactive and reassuring. Antenatal corticosteroid course and latency antibiotics were administered, and fetal surveillance ensued. Owing to recurrent fetal heart rate decelerations that started on day 4 of admission (33-5/7 weeks of gestation), emergency cesarean section was promptly performed. The intraoperative findings were striking for bloody amniotic fluid. A live female neonate was delivered with Apgar scores of 4 and 7 at 1 and 5 min, respectively. The birth weight was 2040 g. A visible ulceration in the umbilical cord a few centimeters from the fetal abdominal wall was noted (Fig. 2b). The neonate was taken to the neonatal intensive care unit. She was pale and her abdomen appeared slightly distended. Anemia was confirmed with a hematocrit level of 22. Packed red blood cells (45 cm^3/kg) were administered over the first days of life. Bilious return was noted from the orogastric tube. Abdominal radiographs at 1 and 20 h of life showed a normal intestinal gas pattern. An upper GI series at 24 h of life demonstrated a partially obstructed duodenum with malrotation and no evidence of volvulus (Fig. 2c). Emergency laparotomy revealed midgut malrotation, viable intestine without volvulus, and dense bands causing obstruction of the duodenum. A Ladd procedure was performed. The neonate recovered and is doing well several months since discharge.

3. Discussion

Life-threatening hemorrhage can result from UCU associated with congenital duodenal and jejunal obstruction [1–17]. Our literature review identified 31 cases, to which we contribute an additional 7 in this series. Most cases were published in obstetrical journals (17/31), and most of the cases originate from Japan (18/31). A citation in 2010 found more than 50 cases [18]. The number of cases in that citation is different than ours because the authors included references that did not meet our search criteria. Some were in Japanese and some were not listed in PubMed.

Our summary of 38 cases reveals a typical pattern for this clinical scenario. Duodenal or jejunal obstruction is diagnosed by fetal ultrasound. Preterm labor or PPROM occurs at an average gestational age of 33 weeks (range 26–38). Signs of fetal distress ensue, prompting cesarean section. A blood-filled amniotic cavity is encountered at delivery. A visible ulcer or multiple ulcers are seen in the umbilical cord. Hemorrhage is commonly fatal either before delivery (12/38; 32%) or in the postnatal period (12/38, 32%).

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