

CASE REPORT

Unusual association of brain hemorrhage and digestive tract occlusion: about two prenatal cases

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Introduction

Because many prenatal disorders involve more than one organ, an important clue of prenatal diagnosis is to look for multiple anomalies.

Therefore, finding a specific lesion commands detailed examination of the fetus to rule out a possible involvement of other organs. This approach is clinically important as identifying multiple lesions may impact on diagnosis, prognosis, and pregnancy/postnatal management.

The aim of the present work was to report on two cases in which digestive tract occlusion or perforation was found to be associated with brain hemorrhage. In one fetus, finding this association modified the prognosis and the outcome of pregnancy, while in the other, it influenced the postnatal management.

Case 1

A 39-year-old pregnant woman was referred to our multidisciplinary center of fetal medicine at 26 weeks of

Key Clinical Message

We report two prenatal cases of an exceptional association of digestive tract atresia or perforation with brain hemorrhage. This combination worsens the prognosis leading to termination of pregnancy in one case. We outline the importance of a careful fetal brain examination on imaging in cases of prenatal “acute” abdominal insults.

Keywords

Brain hemorrhage, digestive occlusion, fetus, peritonitis.

gestation for re-evaluation of fetal intestinal dilatation and ascites. There was no relevant personal, obstetrical, or family history. An amniocentesis performed at 21 weeks due to mother age was normal for a female fetus.

On ultrasound (US), a meconial pseudocyst in the left flank of the fetus, ascites, and slight bowel dilatation (7 mm) were found (Fig. 1). On a tight US follow-up, no significant change was observed. The Doppler study was normal for the umbilical artery and ductus venosus but it showed slight daily changes in the resistive index of the medial cerebral artery from 0.84 to 0.75. There were no signs of fetal anemia. On US performed at 31 weeks, a sudden increase in bowel dilatation (15 mm) was observed. The MRI performed on the next day, confirmed the presence of meconial pseudocyst, small bowel dilatation, and unused microcolon which likely developed in the context of perforation secondary to small bowel atresia. Because the MRI reference scan showed an abnormal hyperintense T1 cerebral lesion, a focused brain MRI was performed the week after, at 32 weeks. This examination demonstrated intraventricular hemorrhage and extensive intraparenchymal fronto-parietal ischemic lesions,



Figure 1. A 26-week and 4-day axial US abdominal scan of the fetus 1 showing the meconial pseudocyst (between crosses) and the ascites (arrow).

consistent with the diagnosis of grade 4 brain hemorrhage (Fig. 2). These lesions had most probably been overlooked at least on the US scan performed on the day before the first MRI.

After multidisciplinary discussion and complete information of the parents, they elected termination of pregnancy at 34 weeks because of the risk of cerebral palsy and poor abdominal prognosis. The couple refused necropsy.

Case 2

A 25-year-old pregnant woman came to our fetal medicine center for routine second trimester US at 22 weeks.



Figure 2. A 31-week T2 weighted MRI coronal slice of the brain of the same fetus showing the periventricular parenchymal ischemic lesions (arrow), the hemosiderin deposits in the subependymal region (arrow head), and the ex vacuo dilatation of the frontal horn of the lateral ventricle: grade 4 hemorrhage.

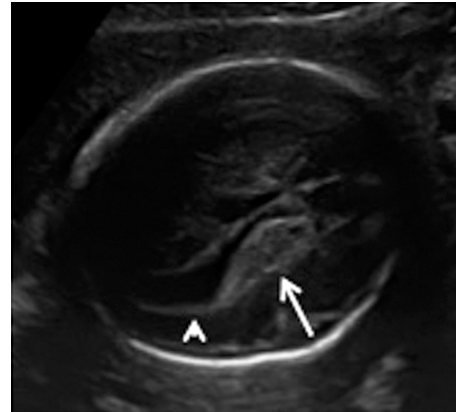


Figure 3. A 25-week axial US scan of the head of the fetus 2 showing subependymal hemorrhage (arrow) and the hyperechogenic wall of the lateral ventricle (arrow head): grade 2 hemorrhage.

There was no relevant familial or personal history except two early miscarriages. Ascites and bilateral pleural effusion were demonstrated without anatomical anomalies or signs of anemia. The amniocentesis revealed neither genetic anomalies nor infection. On subsequent US performed at 25 weeks, an intraventricular brain hemorrhage (Fig. 3) was observed. On further follow-up US, we observed a progressive dilatation of the bowel loops and resolution of the effusions. The colon that should be clearly visible on third trimester US was never seen suggesting proximal bowel atresia. The brain hemorrhage was stable. A comprehensive MRI performed at 31 weeks investigated both regions, the fetal brain MRI confirmed the grade 2 hemorrhage without parenchymal lesions and the abdominal MR sequences demonstrated a distal small bowel atresia with unused microcolon (Fig. 4). On the 37th week US, the loops dilatation increased to 30 mm diameter with heterogeneous content and slight abdominal effusion suggesting parietal damage. A Caesarian delivery was performed on the same day giving birth to a 2620 g baby girl with good Apgar score. The neonatal surgery confirmed an isolated distal small bowel atresia, with proximal intact loops, the atretic segment was resected, and a double ileostomy was done. Neonatal brain imaging confirmed the grade 2 hemorrhage. The digestive tract continuity was restored at day 41.

The clinical neurological and abdominal evolution is eventless at 2 years of follow-up.

Discussion

We report on two cases of prenatal diagnosis of digestive tract occlusion or perforation that were found to be associated with brain hemorrhage; this significantly impacted on prognosis in one case and on postnatal management in the other.

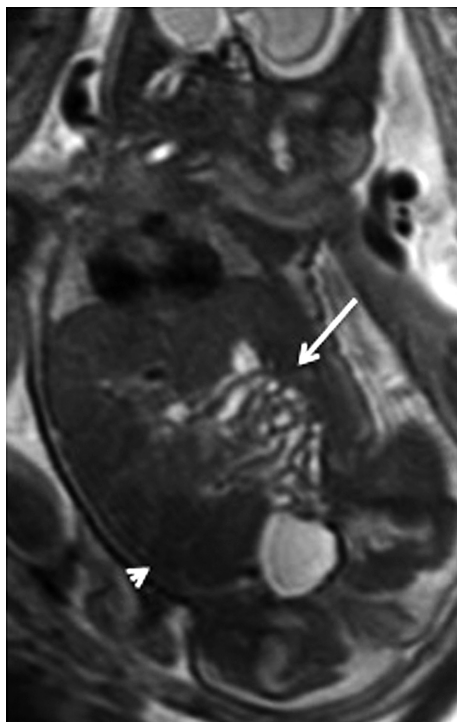


Figure 4. A 31-week T2 weighted MRI coronal scan of the fetus 2 showing normal fluid filled proximal loops (arrow) and dilated intermediate and distal loops attesting of the distal small bowel occlusion. The microcolon is not seen on this image.

Different theories have been proposed to explain digestive tract atresia: for example, lack of vacuolization of the solid cord stage of intestinal development or ischemic insult to the midgut due to mesenteric vascular accident [1, 2]. The atresia can be isolated but may also be associated with meconium ileus, apple peel atresia, volvulus, or abdominal wall defect [3, 4]. Cystic fibrosis should be excluded as it is an associated condition in 7–40% of cases [5, 6]. Concomitant extra-intestinal anomalies are less frequent, though one study reported extra abdominal malformations like congenital heart disease, Down's syndrome, anorectal and vertebral anomalies, neural tube defect, microcephaly, and vesico-ureteral reflux [7].

To our knowledge, the association of meconial peritonitis or ileal atresia with brain hemorrhage has never been described prenatally. A common vascular etiology could be hypothesized as a cause for this rare association. A vascular insult responsible of the gut atresia could be associated with blood flow disturbance leading to cerebral hypoperfusion and subsequent injury [8, 9]. In case 1, the appearance of the brain ischemic lesion on MRI at 31 weeks was compatible with vascular insult going back to a few weeks earlier. Experimental studies in fetal sheep [10] have shown that cerebrovascular immaturity may

impair the ability to maintain constant cerebral blood flow over a wide range of changes in arterial blood pressure; this might also apply to human fetuses. Periventricular and subependymal regions that corresponded to the affected zones in our two fetuses are of greater susceptibility because of high metabolism and watershed vascular regions [11].

Another hypothesis explaining the rare association of meconial peritonitis and brain hemorrhage may be an intravascular meconial dissemination leading to endovascular occlusions by squamous cells. A neonatal case has been published associating meconium peritonitis, periventricular leukomalacia, and pulmonary hypertension leading to neonatal death secondary to respiratory insufficiency. Necropsy disclosed disseminated intravascular occlusions by squamous cells [12]. A case of twin-to-twin transfusion syndrome has also been reported; one twin presented meconium peritonitis and intravascular disseminated coagulation that led to intrauterine death of the other twin. This illustrates that intravascular meconium dissemination may have devastating consequences [13].

These two possible pathogenic mechanisms could cause vascular disruption of the blood supply in multiple organs and hence explain the association we describe in the present report.

These two rare cases associating digestive tract occlusion or perforation and brain hemorrhage give us the opportunity to emphasize the need for careful screening of the fetal brain in such acute abdominal conditions. Because brain lesions may be delayed, neonatologists and pediatricians should also be vigilant and prescribe transfontanellar US whenever a newborn has a history of bowel ischemia and/or perforation.

Conflict of Interest

None declared.

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