First Report of Fatal Secondary Abdominal Compartment Syndrome Induced by Intestinal Gas Accumulation without Organic Occlusive Intestinal Lesion in a Child with Sepsis

Patient: Female, 2
Final Diagnosis: Abdominal compartment syndrome
Symptoms: Abdominal distention • fever • respiratory distress
Medication: —
Clinical Procedure: —
Specialty: Pediatrics and Neonatology

Objective: Unusual clinical course
Background: Abdominal compartment syndrome (ACS), characterized by an increased intra-abdominal pressure and new-onset organ dysfunction, is a critical and potentially fatal condition, with no case of ACS caused by intestinal gas without intestinal lesion being reported to date.

Case Report: A 2-year-old girl with a chromosomal abnormality of 1p36 deletion presented with fever and diarrhea following upper-gastrointestinal series for the evaluation of gastroesophageal reflux. After 20 days, she experienced septic shock and multiple-organ failure, accompanied with rapidly growing, severe abdominal distension. A marked increase in the intra-abdominal pressure was indicated by the complete loss of elasticity in the extremely hard and distended abdomen. She died 14 h after the onset of shock. Her autopsy examination revealed extensive pneumonia and excessive intestinal gas, despite no occlusive intestinal lesion present.

Conclusions: It is critical to be aware that secondary ACS can occur following sepsis due to the accumulation of extensive intestinal gas, without an occlusive intestinal lesion.

MeSH Keywords: Chromosome Aberrations • Disseminated Intravascular Coagulation • Intra-Abdominal Hypertension • Sepsis

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Abdominal compartment syndrome (ACS) is defined by a sustained increase in the intra-abdominal pressure (IAP), accompanied with new-onset organ dysfunction [1,2]. ACS can cause multiple-organ failure due to an increase of IAP and can thus be fatal [3]. ACS can be subdivided into primary, secondary, and recurrent types. Although the primary causes of primary ACS are intra-abdominal complications such as peritonitis and trauma, the main causes of secondary ACS are sepsis, pneumonia, and burns accompanying intra-abdominal fluid collection [4] or intestinal edema and/or ischemia [3]. We performed a PubMed search of the English language literature from January 1, 1966, to December 31, 2018, using the keywords “intra-abdominal hypertension”, “abdominal compartment syndrome”, and “fatal”. We could not find any case of fatal secondary ACS caused by intestinal gas accumulation without an organic occlusive intestinal lesion in children with sepsis. Here, we report a case of pediatric fatal secondary ACS caused by excessive intestinal gas without apparent intestinal lesion during the clinical course of pneumonia and septic shock.

Case Report

A 2-year-old girl with chromosomal abnormality of 1p36 deletion syndrome, the most common (about 1 in 5000 newborns) terminal chromosomal deletion, characterized by developmental delay, intellectual disability, seizures, vision and/or hearing problems, distinctive facial features, brain anomalies, orofacial clefting, congenital heart disease, cardiomyopathy, and renal anomalies [5], was admitted to our clinic for evaluation of gastroesophageal reflux [6]. Previously, she had undergone a pulmonary arterial banding at 4 weeks of age for ventricular septal defect and Ebstein anomaly, and she had lip alveolus and palate cleft, laryngomalacia, hypothyroidism, psychomotor retardation, and gastroesophageal reflux. She was administered home oxygen therapy and tube feeding. After performing an upper-gastrointestinal series on day 2 of admission, the patient experienced fever and diarrhea, which necessitated use of high-flow nasal cannula [7] for the treatment of respiratory distress. Her respiration and circulation were subsequently stabilized, diarrhea was resolved, but fever persisted. Between days 16 and 22, she developed a higher fever (>39°C) and tachycardia (140–180 bpm), exhibited mild lactate level elevation (2–4 mmol/L), showed a mild decrease of percutaneous oxygen saturation (Spo2) to a value in the high

Figure 1. Progression of abdominal distention caused by the accumulation of intestinal gas on X-ray. (A) At 5 days before shock (anteroposterior only). There is relatively less intestinal gas. (B) Just before shock (anteroposterior only). There is a significant increase in the amount of intestinal gas. (C) Three hours after shock (anteroposterior only). The amount of intestinal gas is further increased. (D) Seven hours after shock (lateral only). The abdominal distention has progressed and the abdomen has become much stiffer.
70% range (while her usual SpO\textsubscript{2} was in the mid-80% range due to a right-to-left shunt), and had elevated levels of blood urea nitrogen (39–54 mg/dL). Although her fever and tachycardia were unexplained by her mild dehydration, her general condition and peripheral circulation was preserved. We did not administer antibiotics because her general condition was preserved, and multiple blood examinations performed consistently revealed marginal elevation of C-reactive protein level (the highest value was 2.0 mg/dL on day 2, with a gradual decrease to 0.3 mg/dL on day 24) and negative cultures that did not indicate any bacterial infection. On day 23 of admission, she abruptly presented further higher fever (40–41°C), respiratory distress, and abdominal distention, resulting in hypotensive shock (blood pressure could not be measured). Her laboratory data revealed thrombocytopenia, renal and liver dysfunctions, coagulation disorder, and mixed acidosis, indicating the development of multiple-organ failure (MOF) and disseminated intravascular coagulation (DIC). Accordingly, we initiated intensive care, including mechanical ventilation, inotropes, and antibiotics. Despite continuous decompression of the stomach gas using nasogastric tube and additional decompression of the intestinal gas using Nélaton’s catheter via the anal approach, the abdominal distention caused by the accumulation of the intestinal gas (Figure 1) led to rapid and extreme stiffening of the abdomen, with complete loss of elasticity. Simultaneously, skin reddening was observed on the lower extremities and lower abdomen, indicating the circulatory failure in these regions and further progression of MOF and DIC. No urine was produced after this onset of shock, but urine output was 1.7 mL/kg/h during the 8 h prior to shock onset, and more than 3.0 mL/kg/h during the previous 3 days. Although we assessed that performing open decompressive laparotomy might be necessary to stabilize her condition because of the apparent contribution of the severe abdominal distention to the circulatory failure, we were unable to perform it because of the advanced bleeding tendency associated with DIC. She died 14 h after the onset of acute deterioration.

An autopsy was performed 23.5 h after death. Severe abdominal distention and extreme skin reddening on the lower abdomen and lower extremities were noted (Figure 2A). Although only a
small amount of ascites was observed, the intestinal dilation due to gas retention was remarkable (Figure 2B). However, no obvious organic disease that could cause an intestinal tract obstruction was identified. The intestinal mucosa was macroscopically unremarkable, except for localized dot hemorrhage in the colonic mucosa (Figure 2C). Histological studies revealed congestion, with no edema or inflammation, in the intestinal wall (Figure 2D). The extensive infiltration of neutrophils into the alveolar space in the left upper, right upper, and middle lobes of the lung noted were indicative of extensive pneumonia. However, the cause of the accumulation of extensive intestinal gas remained unclarified.

Discussion

Our literature search revealed that this is the first case of fatal ACS induced by intestinal gas accumulation (without an organic occlusive intestinal lesion) secondary to septic shock in a child. Excessive intestinal gas elevated IAP and caused secondary ACS during the clinical course of pneumonia and sepsis in the present case.

Systemic inflammation and abdominal distention

This infant died on day 24 of her admission and after 14 h of the clinical onset of shock and abdominal distention. During the period between days 16 and 22, the patient demonstrated high fever and tachypnea as well as a mild elevation of lactate and mild decrease of SpO₂. These were not so particularly severe during the clinical course of this child, and her general condition was preserved. Although we could not identify the cause of high fever at that time, the autopsy results suggested systemic inflammation induced by pneumonia that progressed after day 16 of her admission, thereby triggering septic shock and DIC at day 22. Antibiotics should have been administered much earlier despite the marginal elevation of CRP level. Abdominal distention was indicated several hours before the shock, and her illness rapidly progressed to death 14 h after the clinical onset of shock. Such a clinical course indicated prior sepsis and secondary progression of abdominal distension.

ACS caused by intestinal gas secondary to septic shock

Until the day before her death, no findings indicative of impeded blood flow on the lower extremities were noted. Skin reddening on the lower abdomen and lower extremities appeared approximately half a day before death and it rapidly progressed with the abdominal distension. The peak abdominal distention in the present patient was more severe than that indicated by Figures 1D and 2A, 2B for the following 2 reasons: (1) abdominal distention further progressed following the final X-ray of lateral view (Figure 1D), and (2) because an autopsy was performed 23.5 h after death, the photos were acquired at autopsy (Figure 2) after the body was stored in the cooler for over 1 day and after the volume of the intestinal gas had reduced.

Although the IAP measurement using intravesical pressure is important for the diagnosis of ACS [8], it is not always measured for the diagnosis of ACS. Considerable increase of IAP was confirmed solely by external physical finding with percutaneous needle decompression for tension pneumoperitoneum [9] and with a decompression laparotomy [10] in previously reported ACS cases. In the present case, muscular defense by peritonitis was unlikely based on the autopsy findings, and the severely distended abdominal surface was extremely stiff despite the use of a muscle relaxant. Therefore, although IAP was not measured, these physical findings in our case strongly suggested that her IAP was far higher than the cutoff of 10 mmHg [3] or 12 mmHg in pediatric ACS [1] and even higher than the cutoff of 25 mmHg observed in adult severe ACS [3]. The extreme skin reddening was observed on the lower extremities and lower abdomen (Figure 2A), suggesting that the site impeding the venous flow was at the inferior vena cava level rather than at the site of femoral vessel puncture. Furthermore, IAP may be as high as the impeding arterial flow under the hypotensive state considering the lack of apparent edema, despite the extreme skin reddening observed on the lower extremities. The diagnosis of ACS requires new-onset organ dysfunction/failure [3]. Within 1 day until death, MOF progressed with the progression of the abdominal distension. Thus, we diagnosed the present patient with secondary ACS.

The factor responsible for the extremely stiff distended abdomen was the accumulation of the extensive intestinal gas. Although sepsis is a major cause of secondary ACS, ascites, accumulation of intestinal liquid or contents, or intestinal severe edema induced by volume resuscitation [11] are typically responsible for secondary ACS. The autopsy performed in this case, however, revealed neither organic occlusive intestinal lesion nor edema. These findings were completely different from those of typical cases of ACS secondary to sepsis. Overall, autopsy results could not identify the mechanisms for the accumulation of such excessive intestinal gas despite the best efforts to reduce it.

The present report has certain limitations. Except for several negative blood cultures, the assessment for bacterial or other pathogen was not performed, and the causative pathogen for this clinical course remained unidentified. However, it was evident that there was extensive, severe pneumonia but no occlusive or organic intestinal lesion accounting for the ACS progression, even on the autopsy examination. The relationship between this clinical course and her chromosomal abnormality

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remains unclear because we did not find any reported association between them except for a few case reports of sudden unexplained death [12].

Conclusions

Despite the lack of high-quality evidence to support decision making [3], IAP should have been monitored using a urinary catheter [8] for the assessment of disease progression and timely therapeutic decision-making in the present case. Furthermore, this case revealed that secondary ACS can progress due to the accumulation of excessive intestinal gas in cases without organic occlusive intestinal lesion. These findings should be of importance for pediatric intensivists.

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Conflict of interests

None.

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