Resection of a large cystic cervical lymphatic malformation complicated by severe postoperative necrotizing enterocolitis

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A B S T R A C T

Here, we report on a patient with severe postoperative intestinal complications after the resection of a prenatally diagnosed large cervical lymphatic malformation. To elucidate the pathogenesis of this life-threatening complication, we analyzed this case as part of a matched pair. The mature index patient was delivered by Caesarean section, and the postnatal adaption was unimpaired. Surgical resection was performed on the 11th day of life. Intralesional bleeding was identified as the most important risk factor in the development of protracted shock and postoperative necrotizing enterocolitis.

Lymphatic malformations (LMs) are rare congenital dilatations of vascular channels lined by endothelial cells and filled with proteinaceous lymph fluid [1]. Functional and esthetic problems as well as a rapid increase in size triggered by an infection may lead to serious problems in these patients. The complication profile includes locoregional morbidity (e.g., nerve and vascular injury, infection) as well as systemic complications [2]. We would like to add one case who developed deleterious systemic morbidity after resection of a large macrocystic cervical LM (cystic hygroma) due to postoperative necrotizing enterocolitis. To encapsulate the characteristic pathophysiology of the index case, we compared it with data from another comparable infant with an early resection of an LM.

1. Case report

The prenatal ultrasound (US) during the third trimester revealed a large macrocystic protrusion on the right side of the neck in an infra and suprahyoid position. At the age of 39 gestational weeks, the infant boy was delivered by Caesarean section (CS) with a birth weight of 4 kg (90th percentile). Apgar scores were 9, 10 and 10 at 1, 5 and 10 min, respectively. The umbilical arterial pH (UABpH) was 7.29. The newborn showed a large cervical protrusion in the posterior triangle of the left side. MRI confirmed the presence of a large cystic LM of 9 × 6 × 8 cm (stage I according to de Serres et al. [3]). The infant experienced an unimpaired postnatal course, and surgical resection was performed at the age of 2 weeks. Additional malformations were not present, and echocardiography excluded any cardiac anomalies. Preoperatively, the newborn was in a good clinical condition with a blood hemoglobin (Hb) level of 13.2 g/dL (age-related normal...
Abdominal patch-plasty was performed (Tutopatch abdomen. To prevent an abdominal compartment syndrome, an severe edema of the gut wall did not allow primary closure of the be removed, and a terminal ileostomy was created. Furthermore, protein (CRP) level bleeding. Throughout the operation, the patient (NV: 230 compensated metabolic alkalosis (pH, 7.38; pCO₂, 52 mmHg; beginning of the operation revealed a mild, respiratory-tory function was stable. An acid-base analysis (ABA) at the within the bowel wall, was con
prompted a laparotomy. Surgery revealed a complete hemor-

3.5 mmol/L. Broad-spectrum antibiotic therapy was initiated early, and intubation and artificial ventilation continued. After a brief period of improvement under antibiotic therapy, a pro-
gressive protrusion and reddening of the abdominal wall prompted a laparotomy. Surgery revealed a complete hemor-

Of the specimen revealed a cyst lined with endothelium and with associated lymphocellular inflammation. For definitive surgical treatment, an ileorectostomy with J-pouch was established which resulted in continence and regular bowel movements. Ten months after the surgery, the patient is very well developed. No remnants of the LM can be observed.1

1. Reference case

This female patient was delivered in the 35th week after a prenatal ultrasound diagnosis of an extended cervical LM with accompanying polyhydramnios. Her birth weight was 2470 g (50th percentile). Nasotracheal intubation on placental support was performed (UABpH 7.35; Apgar scores 7 and 8). An MRI on the fourth day of life confirmed a large left-sided LM of 6 × cm (stage I according to de Serres et al. [3]) with a broad retropharyngeal extension to the contralateral side. On the 11th day after birth, the LM was surgically removed. Extubation was performed 2 days postoperatively, and the infant’s postoperative course was uncomplicated. Important physiological and perioperative data from this patient served as reference values (Table 1).

### Table 1

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Presented case</th>
<th>Reference case</th>
</tr>
</thead>
<tbody>
<tr>
<td>Position</td>
<td>Right, retropharyngeal</td>
<td>Left, retropharyngeal</td>
</tr>
<tr>
<td>Stage</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Age at resection [days]</td>
<td>14</td>
<td>11</td>
</tr>
<tr>
<td>Hemoglobin [g/L]</td>
<td>13.2</td>
<td>16.3</td>
</tr>
<tr>
<td>Operation time [h:min]</td>
<td>4:10</td>
<td>4:40</td>
</tr>
<tr>
<td>Systolic BP at OP [mm Hg] (start – minimum – maximum)</td>
<td>55 – 60 – 48</td>
<td>55 – 58 – 48</td>
</tr>
<tr>
<td>Blood pH; BE; lactate [mmol/L] at OP</td>
<td>7.4; –1.3; 2.1</td>
<td>n.a.</td>
</tr>
<tr>
<td>Postoperative course</td>
<td>Necrotizing subtotal colitis</td>
<td>Residual retropharyngeal cyst</td>
</tr>
</tbody>
</table>

* Oscillometric measurement.

n.a. – not available.

The treatment of LM is not standardized, and no consensus ex-
ists with regard to the optimal treatment modality. Treatment consists of surgical excision of the LM, but it may also involve sclerotherapy, medical treatment and intralosomal laser applica-
tion. The most serious complications reported were respiratory insufficiency due to infection of the LM, obstruction of the trache-

ostomy tube, aspiration pneumonia, pulmonary embolism, obstruc-
tion of the superior vena cava, and cardiac arrest [2]. Locoregional and postoperative complications include nerve palsy, wound infection, and seroma. According to the appearance, symptoms, and clinical course, the age at which therapy is initiated ranges from the neonatal period up to adulthood [2]. The reported cases were comparable with regard to the prenatal ultrasound diagnosis, delivery via CS, the underlying type of LM, stable pre-
operative condition, and normal preoperative infection parameters. During surgery, both infants showed stable and nearly identical cardiorespiratory parameters (Table 1). However, the initial

![Fig. 1. Intraoperative appearance of the LM. Note the dark color of the LM.](image-url)
The hemoglobin concentration was below normal, and the pulse rate was constantly elevated in the index patient.

Generally, NEC predominantly affects premature infants with body weights below 1.5 kg. However, approximately 1 out of 10 babies who develop NEC are full-term, and the colon is the part of the bowel that is predominantly affected in this age group. The etiology of NEC is multifactorial. The primary risk factors include immaturity of the gut and the mucosa-associated immune system, pathological bacterial colonization of the bowel, perinatal and postnatal cardiorespiratory events, persistent fetal circulation, arterial hypotension, and shock. Both patients fulfilled 3 potential risk factors in the development of NEC, i.e., delivery by CS, preoperative surveillance in the neonatal intensive care unit, and a tendency to become anemic [4–6]. However, NEC developed only in the index patient. During the preparation, we observed nearly bloodless surgical planes as a sign of hemodynamic centralization. In addition, an increase in dark staining of the LM was suspicious for relevant intraluminal bleeding (Fig. 1). Consequently, protracted hemodynamic shock with splanchnic hypoperfusion and bowel necrosis developed in this patient.

Furthermore, it remains speculative whether biologically active proinflammatory mediators (e.g., cytokines, tumor necrosis factor) from the LM may have entered the systemic circulation of the infant. Parallels to amniotic fluid embolism, a rare complication of pregnancy, were observed [7,8].

3. Conclusion

In conclusion, the reduced bleeding during preparation and the increasingly bloody appearance of the lymphatic fluid were the first signs of relevant blood loss. Sufficient and early substitution of fluid volume and red blood cells may be required under these circumstances.

References