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CASE REPORT

Intestinal Lymphangiomatosis in a Newborn with Lower Gastrointestinal Bleeding: Diagnostic Challenge

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ABSTRACT

Congenital cystic lymphatic malformations are rare disorders of the lymphatic tissue lining the soft tissues, lungs, abdominal organs, and bones. We report a case of a female neonate who presented with bloody stools and was subsequently diagnosed with a jejunal lymphangioma. An interesting aspect of our case was that the severe active hemorrhage could not be controlled with medical management and blood transfusion, necessitating urgent surgical intervention by the pediatric surgery team. During the operation, two cystic structures filled with lymphatic fluid were identified in the jejunal bowel loop and were removed by resection. The diagnosis of congenital cystic intestinal lymphangioma, a life-threatening condition presenting with rectal bleeding in the neonatal period, should be considered.

Keywords: Newborn. Congenital cystic intestinal lymphangioma. Bloody faeces. Cystectomy.

Alt Gastrointestinal Kanaması Olan bir Yenidoğanda Bağırsak Lenfanjiyomatozu: Tanı Zorluğu

ÖZET

Doğumsal kistik lenfatik malformasyonlar, yumuşak dokular, akciğerler, abdominal organlar ve kemiklerin lenfatik dokusunu etkileyen nadir bozukluklardır. Biz, doğum sonrası kanlı dışkı ile başvuran ve daha sonra jejunumda lenfanjiyom tanısı konan bir yenidoğan kız olguyu sunuyoruz. Olgumuzun ilginç yönü, şiddetli aktif kanamanın medikal tedavi ve kan transfüzyonu ile kontrol altına alınamaması ve bunun pediatrik cerrahi ekibi tarafından acil cerrahi müdahale gerektirmesiydi. Operasyon sırasında jejunal bağırsak segmentinde lenf sıvısı ile dolu iki kistik yapı tespit edildi ve rezeksiyon ile çıkarıldı. Doğumsal kistik intestinal lenfanjiyom tanısı, yenidoğan döneminde rektal kanama ile başvuran, yaşamı tehdit eden bir durum olarak düşünülmelidir.

Anahtar Kelimeler: Yenidoğan. Konjenital kistik intestinal lenfanjiom. Kanlı dışkı. Kistektomi.

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Although congenital cystic lymphatic malformations, which are due to changes in the embryological development of lymphatic structures, are uncommon and benign formations, they may sometimes be life-threatening[1]. The typical localization is in the head and neck, but they have also been identified in the abdomen[2]. In the literature, they have been described in soft tissue, the lungs, abdominal organs, and bones[3,4]. The incidence of mesenteric cystic lymphangiomas is approximately 1 in 20,000 to 250,000 individuals[5]. Lymphangioma is thought to be associated with obstruction of the lymphatic system, efferent canals or dilatation of the afferent canals as a result of failure, beginning in the sixth week of gestation and resulting in a developmental disorder[6].

Varying forms can be seen, from small lymphangioma which are generally asymptomatic, to large lymphangioma that emerge with various clinical symptoms[4,7]. Lymphatic malformations in children can be seen in any part of the body but are most often determined in the head and neck region (2, 4). Gastrointestinal involvement has been reported to be uncommon[4,5,8] and the most common symptoms seen in these cases are abdominal distension, gastrointestinal bleeding, anemia, protein loss, intestinal perforation, sepsis, and abdominal pain[2,4,5,8,9]. The case reported here presented with bloody faeces of a life-threatening degree, and was determined to be a very rare neonatal case of congenital cystic intestinal lymphangioma.

Case Report

A female infant delivered by caesarean section at 39 weeks with a birthweight of 2850 g, as the second pregnancy of a 29-year-old mother, was transferred to our unit at 27 days old with complaints of bloody faeces (Figure 1), restlessness, and difficulty feeding.

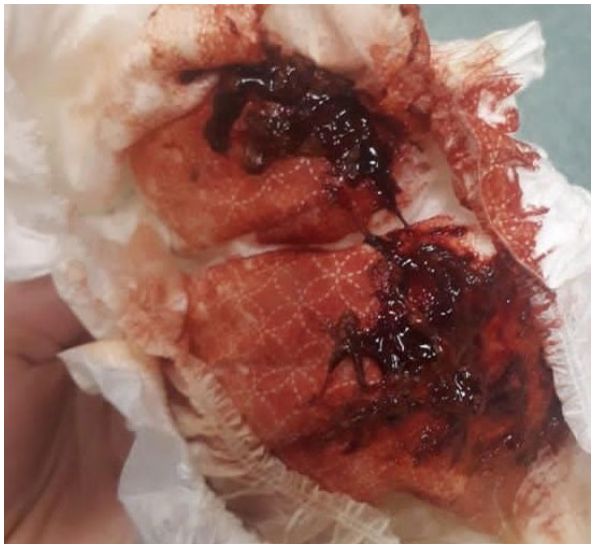

Figure 1.
Bloody stool in baby's diaper

No hyperechogenic bowel was observed during prenatal follow-up. Vitamin K was administered at birth, and the infant has been exclusively breastfed since birth. There was no history of bleeding disorders or allergies in the patient's birth or family history. There was no parental consanguinity.

In the physical examination, the general condition was average with a slight tendency to restlessness and sleep. Other than blood in the diaper of the infant, no other findings were determined in the history.

The body temperature was 34.5 °C (normal range 36.5-37.5 °C), heart rate 160/min, respiratory rate 36/min, and blood pressure was 100/60 mmHg (upper normal limit; 97/64 mmHg). On the postnatal 27th day, the body weight of the infant was 4500 gr (97th percentile) and length was 54 cm (97th percentile). In the initial examination, the respiratory and circulatory systems were normal. The abdominal examination revealed no abnormalities, the skin appearance was normal, and there were no petechiae or/and ecchymoses. The infant was admitted for observation with the initial differential diagnoses of infection, anal fissure, infantile proctocolitis, necrotising enterocolitis, duplication cyst, Meckel's diverticulum, hemorrhagic disease of the newborn, and congenital anomaly of the intestines. The laboratory test results were as follows: white blood cell count: 2000/mm^3, hemoglobin: 10.5 gr/dl, thrombocyte count: 358000/mm^3, C-reactive protein: 123 gr/dl. No eosinophilia was detected in the laboratory. All coagulation parameters were within normal limits. Blood gas analysis revealed, pH: 7.34, PCO_2: 34 mmHg, HCO_3: 18,9 mmol/L, base deficit: 6.8 mmol/L, and lactate: 2.6 mmol/L. The biochemical parameter results were glucose: 265 mg/dl, BUN: 4.4 mg/dl, creatinine: 0.23 mg/dl, uric acid: 2.5 mg/dl, total protein 3.2 g/dl, albumin: 2 g/dl, ALT: 10 U/L, AST: 19 U/L, lactate dehydrogenase: 320 U/L, creatinine kinase: 396 U/L, gamma glutamyl transferase: 30 U/L, total bilirubin: 3.87 mg/dl, direct bilirubin: 0.49 mg/dl, chlorine: 104 mmol/L, sodium: 133 mmol/L, potassium 4.4 mmol/L, phosphorus: 4.30 mg/dl, calcium: 7.73 mg/dl, magnesium: 1.81 mg/dl.

According to the history, physical examination, and laboratory results, neonatal sepsis was considered and treatment with ampicillin, gentamicin, and metronidazole was initiated. Enteral feeding was stopped and intravenous fluid treatment was started. Tests were planned to determine the cause of the lower gastrointestinal bleeding. In the tests performed for the etiology, *Giardia* antigen, enteric bacteria panel, adenovirus, *Entamoeba histolytica*, direct parasite examination, *Cryptosporidium* antigen, COVID-19 PCR test, campylobacter antigen, and anti-HIV tests were all negative. No specific findings were observed on the direct abdominal radiograph taken to determine ileus and other causes.

On ultrasonographic examination of the whole abdomen, septations were observed in the left upper quadrant and cystic lesion formations containing 11 mm septations in the left lower quadrant. In the right lower quadrant of the abdomen, a multiseptate-multiloculated cystic expansile lesion 54 x 21mm in size extending laterally, and fluid localisations between the intestinal loops were observed. Based on these ultrasound findings, the case was initially evaluated as a mesenteric lymphangioma. Contrast-enhanced abdominal Magnetic Resonance Imaging (MRI) was planned for lesion characterization and differential diagnosis, but it could not be performed because the patient was unstable due to ongoing

bloody stools. As a result of the severe bleeding, a significant decrease to 8.1 g/dl was observed in the hemoglobin value of the unstable patient. There were respiratory problems and tachycardia due to anemia so the patient was administered 20ml/kg erythrocyte suspension transfusion. The patient was then admitted for an emergency operation as the active bleeding was continuing. During the operation, two cystic lesions measuring approximately 10×7 cm and 9×7 cm, filled with lymphatic fluid, were identified on a jejunal loop about 20 cm distal to the ligament of Treitz. Since the cysts could not be separated from the adjacent bowel wall and mesentery, a segmental jejunal resection with primary anastomosis was performed (Figure 2).

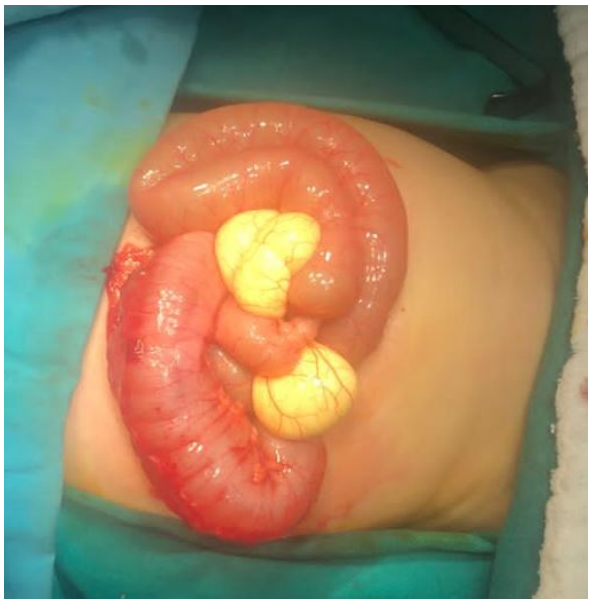

Figure 2.
A cystic structure of approximately 10 x 7 cm containing lymphatic fluid was observed to be tightly adhered to the intestinal ans, and immediately adjacent, there was observed to be a second cystic structure, 10-9 cm in size, containing serous fluid.

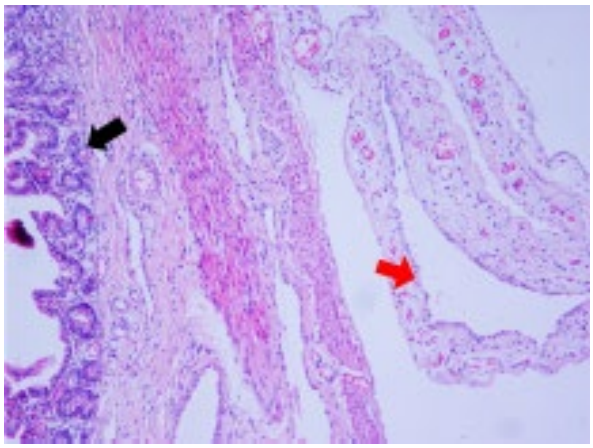

Figure 3.
İntestinal mucosa and muscularis mucosa (black arrow), dilated lymph vessels (red arrow) were seen in a H&E stained section on microscopic magnification of 4x.

The pathological diagnosis of the biopsy sample was confirmed as intra-abdominal lymphatic malformation (lymphangioma). Dilated lymphatics covered with a single layered flat epithelium were observed under the microscope (Figure 3,4). Eosinophilia was not observed. On postoperative day 6, minimal enteral feeding was initiated via nasogastric tube. During the hospital stay, the patient achieved full enteral feeding and had no additional complications. The patient was discharged on the 45th day after birth.

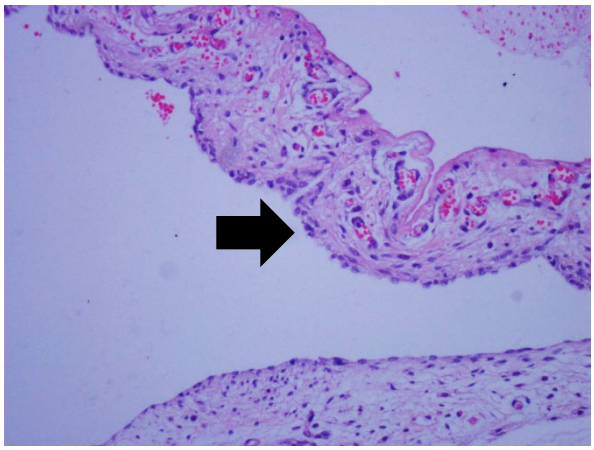

Figure 4.
Dilated lymphatic vessel walls; covered with a single layer flat epithelium (black arrow) (H&E, 20x).

Discussion and Conclusion

Intestinal lymphangioma are clinically concerning congenital malformations that can be seen in a spectrum extending from benign to life-threatening conditions. To our knowledge, this is a very rare case of intestinal lymphangioma seen in a newborn presenting with life-threatening rectal bleeding. A newborn presenting at the polyclinic with lower gastrointestinal bleeding is a rarely seen event, and the most significant complication is severe anemia and findings of failure. As the blood volume is low in newborns, early diagnosis and a rapid start to treatment are important.

In the differential diagnosis of lower gastrointestinal bleeding in the neonatal period, infections, anal fissure, infantile proctocolitis, necrotising eneterocolitis, duplication cyst, coagulation disorders, Meckel's diverticulum, and neonatal late hemorrhagic disease should be considered. In addition to these conditions, rarely seen vascular anomalies and inflammatory intestinal diseases should also be taken into consideration.

Bacterial or viral infections can lead to bleeding by causing damage and inflammation in the intestinal wall. Fever, vomiting, diarrhea, bloody faeces, and general condition deterioration are seen. Diagnosis is made from blood culture, faeces analysis, and other

infection markers. Antibiotics and supportive treatments are administered[10].

Anal fissure in children is a frequently seen clinical problem, and is defined as a tear along the length of the anal canal. Typical presentation is painful defecation and rectal bleeding. The etiology of anal fissure is not known, but the passing of hard faeces and increased internal anal sphincter pressure are accepted as important factors. Treatment aims to eliminate the internal anal sphincter spasm which prevents healing of the fissure. Conservative treatment with faeces softeners, topical analgesics, and sitting baths, is recommended as the first treatment option. The treatment of constipation is important to prevent recurrent tears in the anal canal[11].

Allergic proctocolitis due to food protein is a condition characterised by inflammatory changes in the distal colon as a result of immune-mediated reactions as a response to one or more foreign food proteins. Mucosal and bloody defecation, restlessness, and sometimes skin rashes and vomiting can be seen. The diagnosis is based on food provocation tests and the presence of eosinophils in the faeces. Milk and other dairy products are removed from the diet of the infant, and if the mother is breastfeeding, cow's milk in her diet is restricted. Some clinical characteristics may not be specific and the etiology of rectal bleeding in childhood can be heterogenous. Therefore, it is important to discount other potential causes of rectal bleeding in the paediatric age group, including anal fissure, intestinal intussuception, necrotising eneterocolitis, and very early onset inflammatory intestinal disease in infants[12].

Necrotising enterocolitis is seen more often in premature infants. Necrosis develops in the intestine due to ischaemia. Abdominal swelling, vomiting, food intolerance, and bloody faeces are seen. This condition can worsen and may be life-threatening. Diagnosis is made from the presence of air observed on a standing direct abdominal radiograaph (pneumatosis intestinalis) and from clinical findings. Treatment includes antibiotics, terminating enteral feeding, and when there is perforation surgical intervention is required[13].

Meckel's diverticulum causes bleeding from the diverticulum, which is an embryological remnant in the intestinal wall. Painless bright red bleeding is seen together sometimes with abdominal pain or symptoms of intestinal obstruction. Meckel scintigraphy detects the diverticulum. The treatment for Meckel's diverticulum is surgical removal[10].

Vitamin K deficiency, which is also known as neonatal hemorrhagic disease, can cause significant life-threatening bleeding. It is classified as early presentation, classic or late onset. Early onset occurs within the first 24 hours of life, classic between 1 and 7 days, and late onset between 7 days and 6 months. It is most commonly seen between the 14th day and 3 months. Typical presentation manifests with cutaneous, gastrointestinal, or intracranial hemorrhage most often in infants fed completely on maternal milk[14,15]. The American Paediatric Academy recommends parenteral vitamin K as the most effective means of reducing the risk of severe bleeding and bleeding due to vitamin K deficiency in newborns and young infants[15].

Duplication cyst is a rarely seen type of congenital cyst that develops from the respiratory tract or the digestive system during embryonic development. They may be asymptomatic in the neonatal period. However, depending on the size and localisation, symptoms such as nausea, vomiting, difficulty feeding, abdominal distension, respiratory problems, intestinal obstruction, abdominal pain, and bleeding may be seen. Diagnosis is made with ultrasonography in the prenatal period, and postnatally imaging methods such as ultrasonography, abdominal tomography, or MRI can be used. Cysts that are symptomatic are generally surgically removed. Asymptomatic cysts can be followed up with observation but because of the risk of complications surgery is generally recommended. If an early diagnosis is made, complications (eg., perforation or infection) can be prevented and the prognosis is generally good[16].

Baskin et al. reported the case of a female infant with intestinal cystic lymphangioma diagnosed with prenatal ultrasonography. Intestinal hyperechogeneity and/or dilatation on prenatal ultrasonography and the continuation of these findings throughout the pregnancy suggested pathologies such as meconium ileus, meconium peritonitis and intestinal atresia. Intra-abdominal bleeding and distension occurred in the infant in the postnatal period. On the postnatal 7th day, the infant was exitus. The diagnosis of cystic lymphangioma was confirmed in the postmortem examination[6]. In the current case, there was no intra-abdominal bleeding or distension. However, on the 27th postnatal day, there was severe lower gastrointestinal bleeding.

Lymphangioma may not be evident in the early stages of life and the first symptom may be a complication that puts the life of the patient at risk, such as abdominal swelling, bleeding, or sepsis. Surgical treatment is becoming more discussed and fewer non-invasive techniques are recommended[2]. In the current case, as the bleeding of the patient could not be stopped, the patient was examined for both diagnoostic and treatment purposes.

This case adds to the limited number of reported instances of congenital cystic lymphangioma of the small intestine, particularly in the neonatal period. While lymphangiomas are more commonly found in the head, neck, or axillary region, intestinal

involvement is exceedingly rare, and even more so as a cause of gastrointestinal bleeding in neonates. Most reported cases present later in infancy or childhood with symptoms like abdominal distension or chronic anemia. Our case is unique in that the neonate presented with acute rectal bleeding that progressed to hemodynamic instability, requiring emergency surgical intervention within several weeks of birth. This highlights an important diagnostic consideration for neonatologists and pediatric surgeons when faced with unexplained gastrointestinal bleeding in the early neonatal period. Prompt surgical management led to a favorable outcome, reinforcing the importance of early recognition and intervention in such life-threatening presentations.

In conclusion, this uncommon case of a female infant presenting with bloody faeces in the neonatal period demonstrates that the diagnosis of congenital cystic intestinal lymphangioma must be kept in mind.

Researcher Contribution Statement:
Idea and design: G.T., A.K.; Data collection and processing: G.T., E.K., C.G.; Analysis and interpretation of data: G.T., C.G., N.Y.; Writing of significant parts of the article: G.T., C.G., E.K.
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