The Turkish Journal of Pediatrics 2022; 64: 935-939
https://doi.org/10.24953/turkjped.2021.5045

Case Report

A unique case of a newborn with a hemangioma on the omphalocele sac

Elif Emel Erten[1], Can İhsan Öztorun[2], Süleyman Arif Bostancı[1], Tuğba Örnek Demir[1], Medine Ezgi Öcal[1], Ahmet Ertürk[1], Sabri Demir[1], Doğuş Güney[2], Müjdem Nur Azılı[2], Emrah Şenel[2]

[1]Department of Pediatric Surgery, Ankara City Hospital, Ankara; [2]Department of Pediatric Surgery, Ankara Yıldırım Beyazıt University Faculty of Medicine, Ankara, Türkiye.

ABSTRACT

Background. Mass lesions of the umbilical cord are rare anomalies. There have been rare reports of hemangiomas of the umbilical cord, but the co-occurrence of omphalocele and hemangioma of the umbilical cord has not been previously reported. Nonetheless, the condition is clinically significant as it may cause the disturbance of intrauterine fetal circulation, retardation of fetal growth and development, non-immune hydrops fetalis, morbidity and mortality.

Case. Here we aim to report a case that was prenatally diagnosed with an omphalocele and that presented after birth with a hemangioma on the omphalocele sac.

Conclusions. When dealing with umbilical mass lesions in the prenatal and postnatal periods, a hemangioma on the omphalocele sac should be considered in the differential diagnosis of patients when an omphalocele is suspected.

Key words: newborn, omphalocele, hemangioma.

An omphalocele can be separated but is more as often as possible related with other innate inconsistencies and disorders, such as Beckwith-Wiedemann disorder and trisomies 13, 18 and 21.[1] The administration of neonates born with an omphalocele comprises of the starting steps of airway stabilization, sterile wrapping of the bowel to conserve warmth in order to diminish insensate fluid privation, inclusion of an orogastric tube for bowel decompression, and the foundation of fringe intravenous access. Hemangiomas are congenital lesions originating from errors in embryonic development and are characterized by the proliferation of the vessels' endothelial cells.[2] In most instances, hemangiomas are associated with an increased risk of fetal anomalies, polyhydramnios, fetal hydrops, and perinatal morbidity and mortality.[3] We aim to report an intriguing case of a newborn with a hemangioma on the omphalocele sac who was referred with a prenatal diagnosis of omphalocele. Although many cases of umbilical cord hemangioma have been reported in the literature, to the best of our knowledge no case of hemangioma on the omphalocele sac has been reported previously.

Case Report

A 3180-gram newborn baby who was the firstborn of the first gestation of a 31-year-old mother was referred to us for a consultation. He was delivered by cesarean at 39 weeks of gestation after being diagnosed with an omphalocele at the 23rd week of gestation. During the pregnancy period, the mother had not undergone an alpha-fetoprotein test. The

✉ Elif Emel Erten
elifemelerten@hotmail.com

Received 17th November 2021, revised 15th December 2021, accepted 28th December 2021.

patient APGAR scores at 1min and 5min were 6 and 8 respectively in the neonatal intensive care unit. The omphalocele sac was 7x8 cm in diameter and had a pale, dark blue/yellow appearance with the umbilical cord on top. A dark red mass lesion, which was about 2x3 cm in diameter and resembled a hemangioma, was noticed over the sac (Fig. 1). Sterile wet gauze was placed on the omphalocele sac with hemangioma. The umbilical cord contained 2 arteries and 1 vein. After a short period of patient stabilization, the patient was taken to the operating room. When the sac was opened, it was found that the sac was completely occupied by the liver (Fig. 2) and that the hemangioma was originating from the sac without any obvious connection with the intraperitoneal space. After the liver was introduced into the abdomen, the defect size measured 6 cm. The intra-abdominal pressure was measured with a bladder catheter preoperatively. It was 15 cm H_2O. Upon this the omphalocele was treated by primary repair. An artificial patch was not used. On postoperative day-5, the patient required endotracheal intubation and mechanical pulmonary support due to respiratory distress and then remained intubated for 10 days. After the surgery, the patient had thrombocytosis that gradually decreased to normal values over a period of days. The patient's cranial and abdominal ultrasonography were normal, but a small ventricular septal defect (VSD) and a small secundum atrial septal defect (ASD) were identified on echocardiography.

The patient was discharged home on postoperative day-30. A histological examination of the lesion revealed large cystic dilated vascular structures with thin walls that contained scarce areas of intravascular thrombosis. On immunohistological examination, the vascular endothelium was found positive for D2-40 and CD31 expression. The final morphological and immunohistochemical diagnosis was determined to be a cavernous hemangioma (Fig. 3). A chromosomal analysis was performed, and the karyotype analysis of the patient was 46XY. Written informed consent for publication of this case was obtained from the parents.

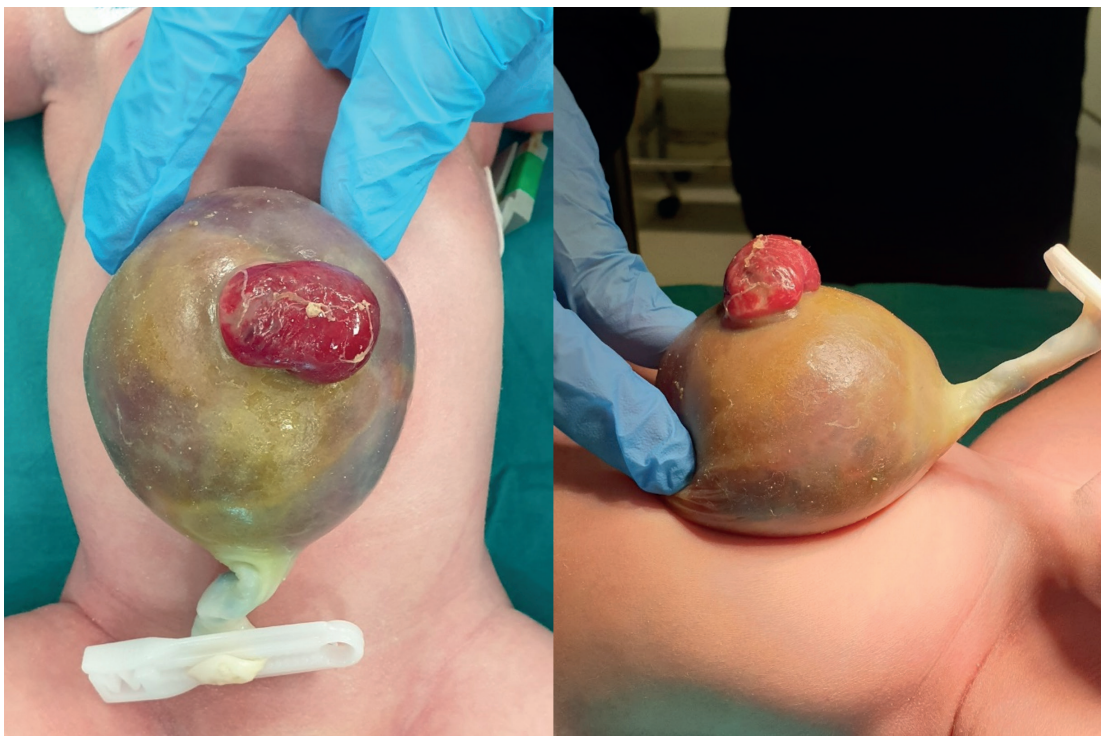

Fig. 1. Omphalocele appearance with the hemangioma and umbilical cord on it.

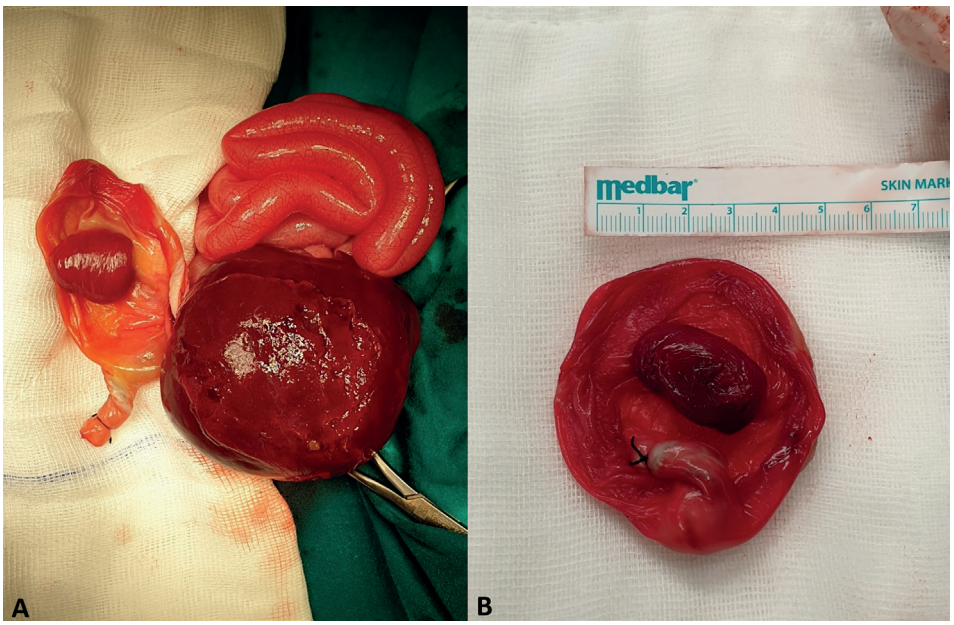

Fig. 2. A. The view of the entire liver after the omphalocele sac wass excised **B.** Macroscopic view of the sac.

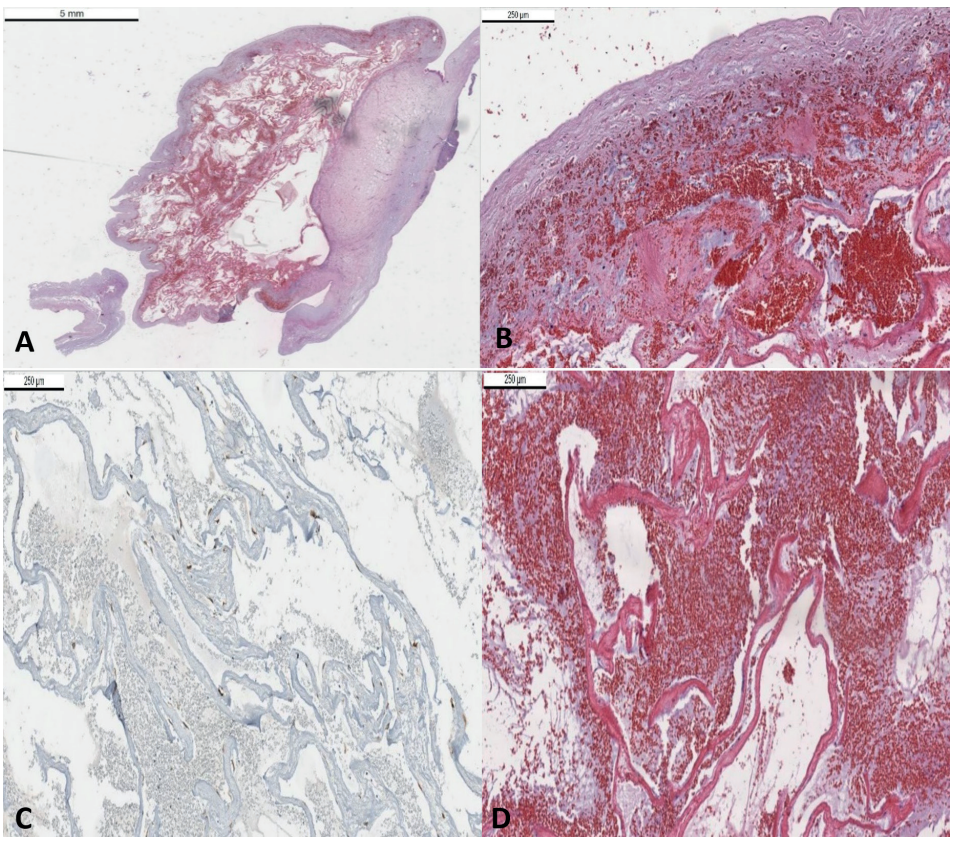

Fig. 3. A. Cystic hemorrhagic lesion with polypoid appearance on the omphalocele sac. **B.** Multiple cavernous cavities filled with blood. **C.** CD34 positivity in the vascular endothelium. **D.** Cystic cavernous structures filled with erythrocytes

Discussion

The omphalocele is a transparent sac connected to the umbilical cord that may contain intestinal structures and/or liver within it. The outer surface of the omphalocele is covered by an amniotic membrane while the inner surface is covered by the peritoneum. Between these layers, umbilical vessels and the embryological remnants of allantois and yolk sac are placed, all of which are surrounded by Wharton's jelly.[1] Associated chromosomal anomalies, such as Beckwith-Wiedemann and pentalogy of Cantrell syndromes, are encountered in about 50 to 70% of the cases.[3] The administration of neonates born with omphalocele comprises of the introductory steps of air duct stabilization, sterile wrapping of the bowel to protect warm to decrease unaware liquid misfortune, inclusion of an orogastric tube for bowel decompression, and the foundation of fringe intravenous entry.

Tumors associated with the umbilical cord are usually reported as isolated malformations without co-existing fetal anomalies.[4] To the best of our knowledge, 12 cases of umbilical cord pseudocyst have been reported to date.[5] Chromosomal anomalies are frequently reported in these cases (53.8%). Trisomy 18 was identified in 6 patients, while only one patient was reported to have trisomy 13.

Increased levels of AFP can be seen in either umbilical cord hemangioma cases or in patients with omphalocele.[6] Mass lesions of the umbilical cord may cause circulatory collapse due to stenosis, thrombosis and torsion of the umbilical vessels via compression of the mass. This process may cause intrauterine growth retardation, non-immune hydrops fetalis and even intrauterine fetal death.[7,8] Loss of the fetus has been reported due to intrauterine bleeding caused by hemangioma of the umbilical cord.[9] Although edema of the umbilical cord is a relatively frequent finding in patients with hemangioma of the umbilical cord, our patient with a hemangioma on the omphalocele sac did not have a such finding.

The presence of a deformity in the abdominal area is a precise complication for pediatric surgeons. Sustaining deficient intra-abdominal force is vital to ensure effective ventilatory procedures. Because modification of this fragile stability is authoritative for the ethics of one-fifth of patients with large omphalocele[10], multistage terminating approaches involving the exertion of patches have been suggested to permit time for the abdominal wall to adjust.[11] Within our, a primary repair was preferred because the intra-abdominal pressure measured preoperatively was not high.

Most cases with an omphalocele can be recognized during prenatal screening and should be differentiated from umbilical cord tumors, omphalomesenteric channel remnants, hemangioma, pseudocyst of the umbilical cord and exstrophy of the bladder. During the differential diagnosis of the omphalocele, it should be kept in mind that these conditions may co-exist with each other.

In our patient, a hemangioma on the omphalocele sac was protruding into the amniotic space and was located remote to the umbilical vessels in the omphalocele sac. Although it was prone to bleeding into the amniotic space due to its location, we did not experience such an event in our patient. The dressing of the patient was done with wet sponges. No additional intervention was applied, as no bleeding was observed. The hemangioma on the omphalocele sac didn't affect the manipulation of the sac and surgical management.

About 45 cases of umbilical cord hemangioma have been detailed within the literature, but to the leading of our information, our case is the primary case of co-existing hemangioma on the omphalocele sac.

When dealing with umbilical mass lesions in the prenatal and postnatal periods, a hemangioma on the omphalocele sac should be considered in the differential diagnosis of patients when an omphalocele is suspected.

Ethical approval

Written informed consent for publication of this case was obtained from the parents.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: EEE, CİÖ, EŞ; data collection: CİÖ, TÖD, MEÖ; analysis and interpretation of results: SAB, AE, SD; draft manuscript preparation: DG, MNA, EEE, EŞ. All authors reviewed the results and approved the final version of the manuscript.

Source of funding

The authors declare the study received no funding.

Conflict of interest

The authors declare that there is no conflict of interest.

REFERENCES

1. Zahouani T, Mendez MD. Omphalocele. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2021.
2. Gasparella P, Singer G, Arneitz C, et al. Rapidly involuting congenital hemangioma of the liver in a newborn with incomplete Pentalogy of Cantrell: description of a new association. J Surg Case Rep 2021; 2021: rjab047. https://doi.org/10.1093/jscr/rjab047
3. Emura T, Kanamori Y, Ito M, et al. Omphalocele associated with a large multilobular umbilical cord pseudocyst. Pediatr Surg Int 2004; 20: 636-639. https://doi.org/10.1007/s00383-004-1247-y
4. Miller KA, Gauderer MW. Hemangioma of the umbilical cord mimicking an omphalocele. J Pediatr Surg 1997; 32: 810-812. https://doi.org/10.1016/s0022-3468(97)90625-4
5. Suzuki T, Yamamoto Y, Nakamura H, et al. Fetal umbilical cord cyst may evolve to omphalocele during pregnancy. J Clin Ultrasound 2020; 48: 181-183. https://doi.org/10.1002/jcu.22786
6. Iglesias-Deus A, Pérez-Muñuzuri A, Urisarri A, Bautista-Casasnovas A, Couce ML. Umbilical cord and visceral hemangiomas diagnosed in the neonatal period: a case report and a review of the literature. Medicine (Baltimore) 2016; 95: e5196. https://doi.org/10.1097/MD.0000000000005196
7. Natalucci G, Wisser J, Weil R, Stallmach T, Bucher HU. Your diagnosis? Umbilical cord tumor. Eur J Pediatr 2007; 166: 753-756. https://doi.org/10.1007/s00431-006-0301-2
8. Caldarella A, Buccoliero AM, Taddei A, Savino L, Taddei GL. Hemangioma of the umbilical cord: report of a case. Pathol Res Pract 2003; 199: 51-55. https://doi.org/10.1078/0344-0338-00354
9. Vougiouklakis T, Mitselou A, Zikopoulos K, Dallas P, Charalabopoulos K. Ruptured hemangioma of the umbilical cord and intrauterine fetal death, with review data. Pathol Res Pract 2006; 202: 537-540. https://doi.org/10.1016/j.prp.2006.02.008
10. Saxena AK, Raicevic M. Predictors of mortality in neonates with giant omphaloceles. Minerva Pediatr 2018; 70: 289-295. https://doi.org/10.23736/S0026-4946.17.05109-X
11. Saxena AK, van Tuil C. Delayed three-stage closure of giant omphalocele using pericard patch. Hernia 2008; 12: 201-203. https://doi.org/10.1007/s10029-007-0264-x