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Hemangioma of the Umbilical Cord: A Case Report

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ABSTRACT

Background: Hemangioma is one of the umbilical cord anomalies and it is arising from the remnants of embryo hemangioblasts. It is associated with increased perinatal morbidity and mortality.

Case report: We describe an umbilical cord hemangioma that was identified in antenatal period. The patient was referred to our hospital for further evaluation of omphalocele that was noticed at gestational age of 29 week. The mass, measured 80*75*50 mm diameter, was surrounded by edema of the Wharton jelly. Color Doppler imaging revealed arterial and venous flow within the mass. The neoplasm was composed of organized and non-organized capillary vessels within the myxoid stroma. Neoplasm was covered with squamous epithelium focally but the epithelium was widely eroded. The immunohistochemical examination positive staining was obtained for vimentin, desmin, CD34 and SMA.

Conclusion: Umbilical cord anomalies are rare. Early detection and close follow-up can reduce the risk of perinatal morbidity and mortality.

KEYWORDS
Hemangioma; angiomyxoma; umbilical cord tumor.

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Introduction

Umbilical cord anomalies are rare and the prenatal differential diagnosis is generally difficult. Hemangioma is one of the umbilical cord anomalies and it is arising from the remnants of embryo hemangioblasts. While it can be visualized as an abnormal vascular pattern during the ultrasonographic examination, in most cases the definitive diagnosis cannot be made antenatally. It is associated with increased perinatal morbidity and mortality.[1-3] Herein, we describe an umbilical cord hemangioma that identified antenatal period.

Ethic statement

The data collection and reporting of this case study were carried out in accordance with the Helsinki Declaration. Written informed consent was obtained from the patient for the publication of the data and the use of related materials.

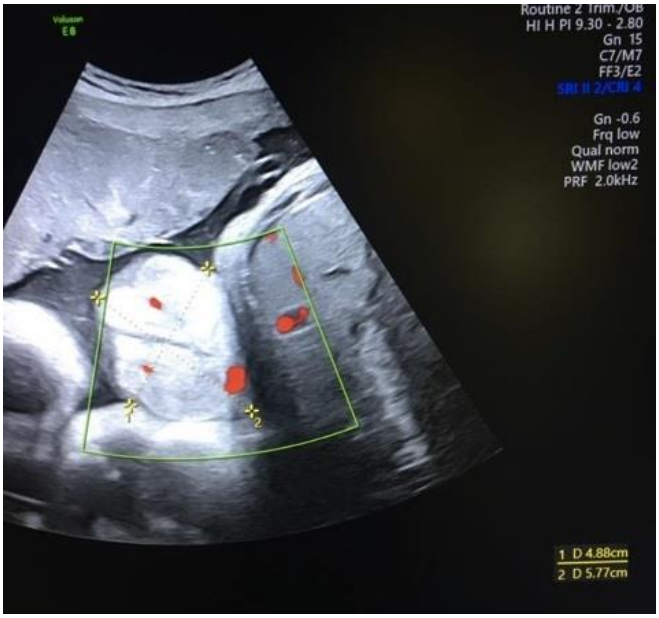
Case report

A 32-year-old, gravida 1, parity 0-0-0 was referred to our hospital for further evaluation of omphalocele that was noticed at gestational age of 29 week. Her medical history was unremarkable. The first trimester screening revealed low combined risk for trisomy 21. Second trimester alpha fetoprotein level was 1.27 MoM. No anomaly was detected on the second trimester ultrasound screening. Our sonography examination at 29-week of pregnancy revealed a heterogenous and complex mass, composed of cystic-solid parts, that was arising from the umbilical cord entrance to the fetal abdomen. The mass, measured 48*57*50 mm diameter, was surrounded by edema of the Wharton jelly (Figure 1). Color Doppler imaging revealed arterial and venous flow within the mass (Figure 2). Except polyhydramnios, the fetal sonographic examination was normal. An umbilical cord angiomyxoma was considered based on the present findings. The patient was followed-up periodically and at 39th gestational week, she was hospitalized with the complaints of amnion leakage and uterine contractions. She gave birth a male neonate (4015 g, APGAR scores 6 and 8) with caesarean section (Figure 3).

The neonate was admitted to intensive care unit and the umbilical mass was excised on the second day after birth. After 20 days, baby was discharged from hospital.

On the pathologic examination, umbilical vessels consisting of 2 umbilical arteries and 1 umbilical vein are observed adjacent to the neoplasm and within the tumor in focal areas. The neoplasm is composed of organized and non-organized capillary vessels within the myxoid stroma. Neoplasm is covered with squamous epithelium focally but the epithelium is widely eroded. The immunohistochemical examination positive staining is obtained for vimentin, desmin, CD34 and SMA (Figure 4-9). For S100, CAM5.2, ER, PR and AR, there is no immune reaction detected. No thrombus was reported. Based on these pathologic findings, umbilical cord hemangioma was reported as the final diagnosis.

Figure 1. Ultrasound images of the umbilical cord hemangioma.



Note: Hyperechogenic mass delineated from the edematous Wharton jelly.

Figure 2: Doppler flow image and measurements of the tumor.

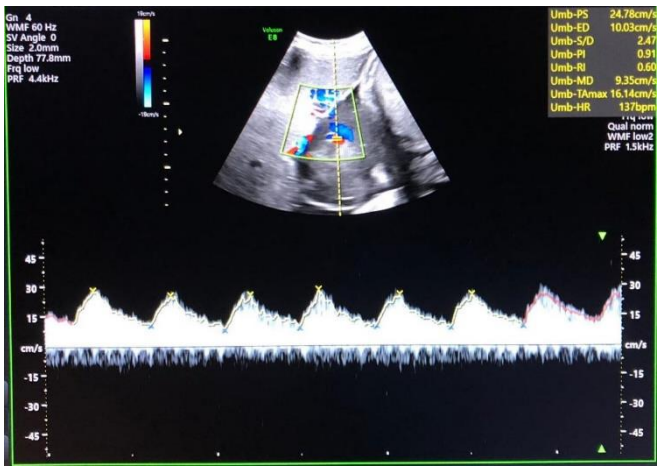
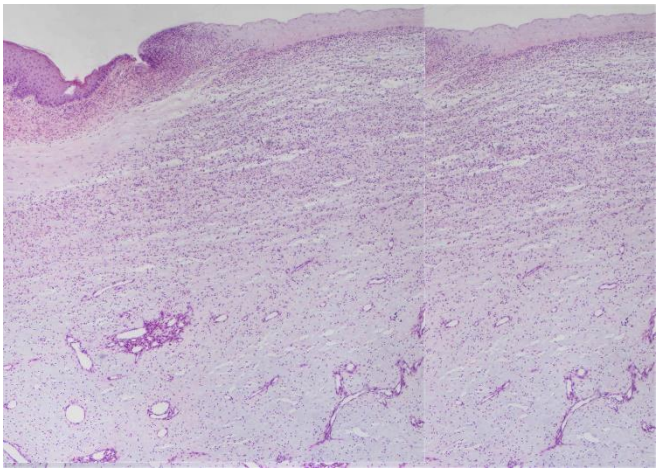


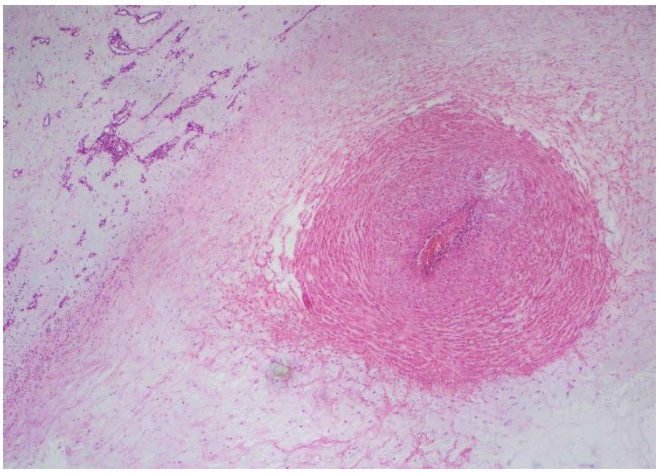
Figure 3: Appearance of the umbilical cord shortly after birth

Figure 4: The neoplasm consisting organized or non-organized capillary vessels within the myxoid stroma, covered by squamous epithelium focally is observed.



Note: The epithelium is widely eroded. The lesion is infiltrated by numerous neutrophils in the stroma and at the surface. (HE, x40)

Figure 5: Umbilical vessels are observed adjacent to the neoplasm and within the tumor in focal areas.



Note: A few neutrophils attacking the umbilical artery is seen. (HE, x40)

Figure 6: The neoplasm is composed of organized and non-organized capillary vessels within the myxoid stroma (HE, x40).

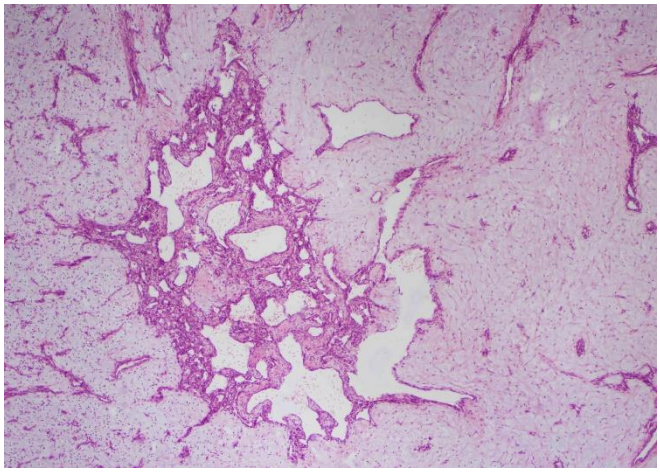


Figure 7: There are numerous neutrophil leukocytes in the stroma and at the surface of the neoplasm and these neutrophils attack the vascular walls. (HE, x200)

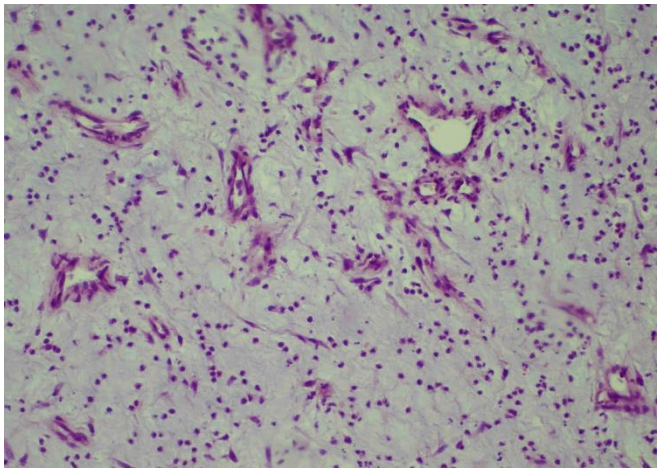


Figure 8: Positive staining is obtained at vascular endothelial cells for CD34 immunohistochemical staining.

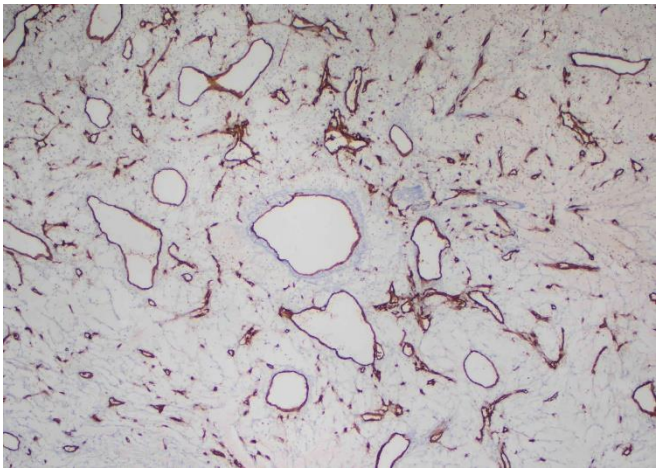
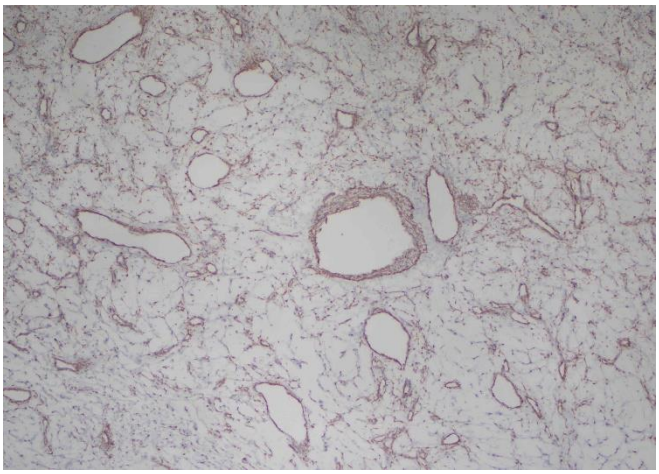


Figure 9: Positive staining is obtained at the vessel walls for SMA immunohistochemical staining.



Discussion

Hemangioma is a rare anomaly of the umbilical cord. These benign tumors can usually be an isolated anomaly but sometimes they can be described in association with cutaneous lesions, cardiac and central nervous system malformations, and syndromes.[2,4] Early detection and close follow-up can reduce the risk of perinatal morbidity and mortality. In differential diagnosis umbilical vein

varices, aneurysm, hematoma, teratoma and abdominal wall defects should be considered.

Hemangiomas are known as benign tumor that is arising from allantoic or omphalomesenteric vessels. It is generally located in distal part of the cord and because of this it can be misdiagnosed as abdominal wall defect.[2,5,6] Therefore, the integrity of the abdominal wall and presence of any fetal organ in the mass should be carefully examined on the ultrasound screening.

The antenatal diagnosis of an umbilical cord hemangioma is considered when a hyperechogenic mass is detected in the umbilical cord. As the similarities of sonographic finding, distinction among umbilical cord teratoma, hematoma and omphalomesenteric duct cyst may be difficult.[7] Histopathological examination provides differential diagnosis. While hemangioma generally originates from umbilical vessels endothelium, the teratoma is derived from germ cell layers.[6-8] In our case, all of the umbilical vessels were covered by the tumor. In immunohistochemical exam, positive staining for CD 31 and negative staining for S 100 also correlated with hemangioma. Sonographic examination of umbilical cord hematoma reveals usually septate and hypoechoic unless acute hemorrhage.

Elevated AFP, polyhydramnios, umbilical vessels thrombus, preterm delivery and hydrops may be associated with umbilical cord hemangiomas. In some cases, placental pathology may also accompany but its clinical significance was not shown.[5-9] In our case, any placental pathology or thrombus were detected and AFP level was normal. A previous study reported that elevated AFP has been found in 60% of the hemangiomas and this was probably due to the impairment of the amniotic fluid barrier.[10] Although it occurs less frequently than in placental hemangioma, polyhydramnios may also be seen in umbilical hemangioma.[11] It is thought that, increased transudation into the amniotic cavity from the elevated vascular pressure in the hemangioma can be the reason of the polyhydramnios.[12] In our case, large tumoral mass with increased vascular pressure and transudation of fetal serum through the thinned walled vessel of the tumor may cause polyhydramnios without elevated AFP level.

Umbilical cord hemangiomas are highly related with poor perinatal outcomes such as preterm delivery, hydrops, intrauterine asphyxia and fetal death. Umbilical cord torsion, thrombus or bleeding within the tumor and mechanical vessel compression are the causes of these adverse outcomes.[3,13] Amniotic fluid volume, Doppler flow studies of umbilical vessels and tumor size should be closely monitored by serial ultrasound examination.[3,14] Thinking about the possibility of a sudden vascular accident, delivery should be considered when pulmonary maturity is assumed.[7]

Conclusion

This case represents a rare benign tumor of umbilical cord that is complicated with polyhydramnios. Because of the relationship with increased perinatal morbidity and mortality, it is imperative to close follow-up fetal well-being.

Conflict of interest

The authors declare that they have no conflicts of interest.

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