A Newborn with a Large Umbilical Cord Pseudocyst with Hemangioma: A Case Report and Review of the Literature

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Umbilical cord pseudocysts in a newborn are very rare. They may be associated with patent urachus and hemangioma. Generally, they are localized in a particular section of the cord. Urachal or vitelline duct cysts, teratoma, omphalocele, umbilical cord hernia and hematoma are considered in differential diagnosis. In this case report, we discussed the clinicopathological findings of a pseudocyst with hemangioma involving the entire cord in a newborn, and in particular the confusing conditions related to the excision of the umbilical cord.

Key Words: Umbilical cord, Hemangioma, Angiomyxoma, Umbilical pseudocyst

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Introduction

Umbilical abnormalities are very rare in newborns.[1] Compared to urachal and vitellin duct abnormalities, omphalocele, and umbilical cord hernias, umbilical cord pseudocysts are very rare. These mass formations may be associated with patent urachus and hemangioma.[2,3]

To date, only eight cases of umbilical cord pseudocyst associated with hemangioma have been reported in the literature, and which involved practically no data on the therapeutical approach.[3] Therefore, we discussed the clinicopathological findings of this disorder surprisingly occurring in a newborn, and in particular, the confusing clinical conditions encountered upon this occurring during delivery.

Case Report

A 25-year-old woman (gravida 2, para 1) attended follow-up visits once every two months during her pregnancy. In the sixteenth gestational week, the maternal serum alpha-fetoprotein, unconjugated estriol and human chorionic gonadotrophin levels were normal. We had no data of nuchal translucency of the fetus. At the end of an unproblematic, 37-week pregnancy, the patient underwent cesarean section due to fetal distress and a female baby weighing 3300 grams was born. The infant had an APGAR score of 7 and 9 in the 1st and 5th minutes, respectively. The umbilical cord was observed to be quite large and edematous; whereas, no umbilical cord mass was detected during pregnancy. The cord was tied with a clamp quite far from the umbilicus with the concern that an intestinal segment could exist in the cord sac (figure 1). After consultation with the pediatric surgery department, the infant was transferred to the neonatal intensive care unit. Vascular access was obtained and the infant was placed under a radiant heater to prevent hypothermia. Systemic examination findings were normal with a normal abdominal ultrasound scan.

The large umbilical mass was rinsed with betadine solution and covered with a sterile covering. After a repeated careful examination, the umbilical cord mass was detected to contain no intestinal segment or any patent duct since it was translucent. At the entrance of the cord to the umbilicus, the diameter was nearly the size of a normal cord. Under local anesthesia with EMLA cream , the umbilical cord was tied to the entrance of the umbilicus using a 2.0 silk suture and was excised by cutting from a part 2-3 mm distal to the tied suture. The infant was given to the mother and began breast-feeding. No karyotyping could be performed because the mother did not agree to such an investigation. The macroscopic examination revealed an umbilical cord of 48-cm in length and 520-gram in weight with a translucent and homogenous structure with the largest section being 11-cm width, containing 1 vein

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and 2 arteries in the edematous Wharton jelly on the transverse section. The histopathological investigation revealed that hemangioma resulted from the umbilical artery and was associated with myxoid degeneration (Figure 2). The examination of the placenta detected no pathology. The infant exhibited normal findings in the system examinations performed at first week, first month and sixth months, and the cosmetic appearance of the umbilicus was perfect .

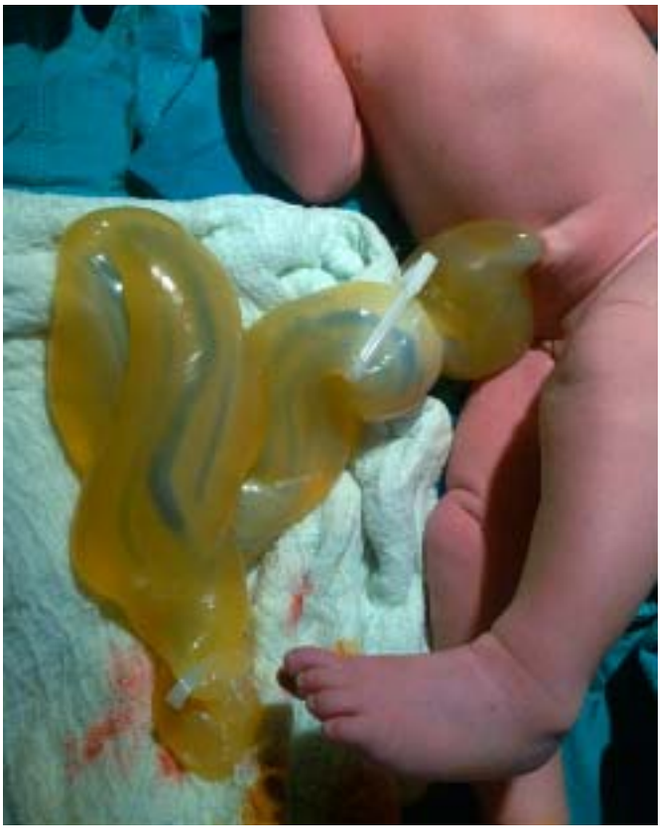

Figure 1: The photo shows the umbilical cord after delivery.

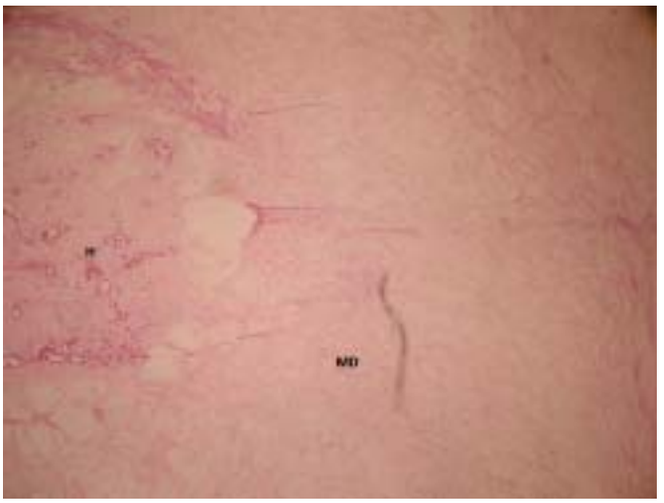

Figure 2: Histopathological examination shows hemangioma and myxoid degeneration (PASX100).

Abbreviations: H: Hemangioma MD: Myxoid Degeneration

Discussion

The congenital cysts observed in the umbilical region of the neonate may be considered as urachal or vitellin duct cysts, or teratoma, omphalocele and umbilical cord hernias, which are rarely occurring real masses. There are also the pseudocysts of the umbilical cord, which result from the myxoid degeneration of the Wharton Jelly. These were also termed as umbilical cord edema, pseudotumor, pseudocyst, Wharton's jelly cyst, angiomyxoma, hemangiofibromyxoma and telangiectatic myxosarcoma.[1,4,5] Generally, they involve a certain section of the cord, and particularly occur in regions close to the umbilicus. As is the case in our patient, the umbilical cord may be very large from the umbilicus to the placenta, edematous, translucent and homogenous.

While the massive edema is believed to be associated with the high permeability of the angiomatous veins located in the soft and gelatinous tissue, the actual mechanism is not established. In the literature, pseudocyst resulting from massive edema was reported in only one case in association with patent urachus.[2] Our case is the first case of large pseudocyst with hemangioma in the literature. This disease may be accompanied by chromosomal abnormalities including trisomia 18, trisomia 13 and Down's syndrome. In addition, it may be associated with omphalocele, cardiac malformation, vertebral defects, radial and renal dysplasia, tracheoesophageal fistula, urachus cyst and hemangioma.[3]

Association of pseudocyst and hemangioma has been reported in eight cases in the literature to date. Hemangioma is generally located in the placental end of the umbilical cord and less commonly in the fetal part. As far as we know, a case of pseudocyst with hemangioma involving the entire cord is published for the first time. Generally, it originates from a single vein or multiple veins, and particularly from the umbilical artery.[3,6]

A mortality of 35% was reported in cases of umbilical cord hemangiomas . They may lead to maternal obstetrical complications, including hydrops fetalis, intrauterine growth retardation, severe fetal hemorrhage and intrauterine fetal death . Diagnosis with prenatal ultrasonography is important in preventing prenatal and postnatal complications.[7] While it may particularly be diagnosed by Color Doppler ultrasonography, it may also not be detected, as is the case in our patient. Such a pathology is surprising for the obstetrician, neonatologist and the midwife assisting in the labor/cesarean section, particularly if it is not detected on prenatal ultrasonography. In case of such an unusual event, there may be hesitation in clamping the cord immediately after labor because such a massive mass is considered to potentially contain an intestinal segment. Concerns arise for the potential occurrence of severe compli-

cations such as intestinal obstruction and peritonitis caused by clamping-cutting of the cord containing an intestinal segment. However, in this case, we were lucky that the lesion was translucent and homogenous; with careful examination, a formation other than the umbilical veins can be observed that does not, for example, represent an intestinal segment. After ensuring that there is no intestinal segment, such massive umbilical mass can be excised after clamping the site of entrance to the umbilicus under local anesthesia, with no need for general anesthesia, which may involve various complications, as is the case in our patient. We present this case report to underline the fact that this very rare disorder can be successfully treated on an outpatient basis with a perfect cosmetic outcome. In case of such pathology, the labor staff should carefully and rapidly examine the umbilical cord, and clamp the umbilical cord as usual unless there is an intestinal segment or patent duct inside the mass with a reduced diameter at the site of entrance to the umbilicus.

Hemanjiomlu Geniş Umbilikal Kord Psödokisti ile Beraber Olan Yenidoğan: Olgu Sunumu ve Literatür Işığında Tartışılması

Fetüste umbilikal kord psödokistleri nadirdir. Patent urakus ve hemanjioma ile ilişkili olabilirler. Genelde kordun belirli bir bölümünde lokalizedir. Urakal veya vitellin kanal kistleri, teratoma, omfalosel, umbilical kord hernisi ve hematom ayırıcı tanıda düşünülür.

Bu vakada yenidoğandaki tüm kordu içeren hemanjioma ile birlikte olan psödokistin klinikopatolojik bulgularını ve umbilical kordun eksizyonu ile ilgili durumu tartıştık.

Anahtar Kelimeler: Umbilikal kord, Hemangioma, Angiomiksoma, Umbilikal psödokist

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