**Angiomyxoma of the Umbilical Cord**

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**Summary**

**Objective:** Angiomyxoma is a rare tumor of the umbilical cord and is associated with increased perinatal morbidity and mortality. However, the management of these pregnancies in the third trimester is not clearly defined. We present a case of an angiomyxoma of the umbilical cord diagnosed in the second trimester, and highlight the contribution of color Doppler imaging to the early diagnosis of cord anomalies.

**Case Report:** A 29 year-old, gravida 3, para 1, woman had elevated maternal serum α-fetoprotein at 17 weeks of gestation. Ultrasonography at 19 weeks showed a placental mass measuring 2 × 1.5 cm over the insertion site of the umbilical cord. The mass slowly enlarged in size, from 2.72 × 1.09 cm at 21 weeks to 3.9 × 3.9 cm at 33 weeks. Beyond the cord lesion, the development of the fetus was unremarkable. At 38 weeks, a normal female infant was delivered by cesarean section due to previous history of cesarean section. A mass measuring 3.2 cm was found near the insertion site of the umbilical cord to the placenta. Pathologic examination showed proliferation of thin-walled vessels embedded in a myxoid stroma, and the endothelial cells were positive for factor VIII-related antigen.

**Conclusion:** Angiomyxoma is a rare tumor of the umbilical cord and should be considered when using prenatal ultrasound for detection of cystic lesion. Color Doppler imaging can easily and instantly detect perfusion through the umbilical vessels and assess cardiac function. In our case, application of color Doppler imaging for monitoring the relationship between the tumor and the adjacent vessels allowed the fetus to be delivered at term with a favorable outcome. [Taiwanese J Obstet Gynecol 2006;45(4):360–362]

**Key Words:** angiomyxoma, ultrasonography, umbilical cord

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**Introduction**

Angiomyxoma is an uncommon subtype within a group of tumors of the umbilical cord [1]. It has been reported to be associated with increased perinatal morbidity and mortality [2]. The management of these pregnancies in the third trimester is not clearly defined. We describe a case of an angiomyxoma of the umbilical cord, diagnosed in the second trimester, highlighting the contribution of color Doppler imaging to the early diagnosis of cord anomalies.

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**Case Report**

A 29 year-old, gravida 3, para 1, woman had previously had cesarean section once for prolonged labor; she had an otherwise uneventful medical history. Elevated maternal serum α-fetoprotein (436.29 ng/mL) was noted at 17 weeks of gestation. Ultrasonography at 19 weeks showed a placental mass measuring 2 × 1.5 cm over the insertion site of the umbilical cord (Figure 1). At follow-up, the mass had slowly enlarged in size, from 2.72 × 1.09 cm at 21 weeks to 3.9 × 3.9 cm at 33 weeks. Beyond the cord lesion, the development of the fetus was unremarkable. At 38 weeks, a female infant was delivered by cesarean section due to previous history of cesarean section. The infant was normal at birth with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. A mass, measuring 3.2 cm in its greatest
dimension, was found near the insertion site of the umbilical cord to the placenta. Pathologic examination showed proliferation of thin-walled vessels embedded in a myxoid stroma, and the endothelial cells were positive for factor VIII-related antigen (Figure 2).

Discussion

Tumors of the umbilical cord are extremely rare. In 1925, Browne cited eight cases of umbilical cord tumor described in the literature [2]. These included four telangiectatic myxosarcomata, a rather poor term for a lesion that pursues a benign course, one angiomyxoma, and one teratoma. The diagnosis of angiomyxoma was supported by the microscopic appearance, showing that the main tumor consisted of myxomatous tissue with hemangiomatous foci [1]. With the advances in ultrasound, obstetricians are now able to examine the placenta and the cord in detail before delivery. In this case, serial ultrasonographic examinations provided a unique opportunity to analyze the development of the lesion in utero. The tumor was detected at 19 weeks by the finding of elevated maternal serum α-fetoprotein. The application of the color Doppler technique allowed optimal analysis of the perfusion through the umbilical vessels and assessment of cardiac function between the 19th and 36th week of gestation.

Some cases of angiomyxoma were associated with premature delivery, cardiovascular anomalies, non-immune hydrops fetalis, hydatidiform mole, polyhydramnios, disturbance of fetal heart rate, and stillbirth [1,3–7]. Yavner and Redline described an extremely large angiomyxoma of the umbilical cord and a subsequent benign clinical course [8]. In the absence of an arteriovenous fistula, clotting, or an anatomic location impeding blood flow, large angiomyxoma has little impact on fetal hemodynamic status. In some cases, however, fetal morbidity and mortality may result from secondary direct mechanical compression of adjacent blood vessels by the increasing tumor size [1]. In this case, the tumor size was not too large and direct mechanical compression of adjacent blood vessels did not occur, resulting in a favorable fetal outcome.

Although cesarean section is used to manage most cases, Wilson et al reported a case of spontaneous vaginal delivery after ultrasound-guided in utero decompression of a massive cystic angiomyxoma of the umbilical cord [9]. Concerns regarding intrapartum management without cystic decompression include possible risks of dystocia or sudden cystic rupture affecting blood flow through the cord [9]. Furthermore, unusual extension of the lesion with secondary shortening of the cord would make it mandatory to deliver the infant by cesarean section. Controlled aspiration with ultrasonographic guidance may be used to decrease these potential risk factors. However, the theoretical risks of vascular compression with secondary thrombosis and/or shortening of the cord underline the importance of an early prenatal diagnosis of this anomaly.

References