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Umbilical cord hemangioma associated with placental chorangiomatosis: a case report and literature review

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Summary

A case of umbilical cord hemangioma associated with placental chorangiomatosis is reported. A premature male infant was delivered vaginally at 34 weeks of gestation. Congenital abnormality and visceral hemangioma were not detected after birth. The placenta did not exhibit any macroscopic abnormalities. In the middle of cord, an enlarged portion with 3 cm in length and 2.5 cm in diameter was identified. Microscopical examination revealed an umbilical cord hemangioma associated with segmental chorangiomatosis in the placenta. Among all the reported cases of umbilical cord hamangioma, only two cases were found associated with placental pathological changes, and neither of them was diagnosed with chorangiomatosis. As the first case reported umbilical cord hemangioma accompanied by placental chorangiomatosis, the pathological changes of umbilical cord hemangioma and associated placental changes are discussed in detail, and the relevant literature is reviewed in this report.

Key words: Umbilical cord; Hemangioma; Chorangiomatosis; Pathology.

Introduction

Hemangioma of umbilical cord is unusual. A total of 38 cases have been reported according to current literature [1-9], and only two of them were found associated with placenta pathological changes [1, 7]. One case was diagnosed with placental chorangiosis [1], and the other was reported to have avascular villi [7]. Here the authors present a case with umbilical cord hemangioma associated with placental chorangiomatosis. To the best of their knowledge, it is the first case of umbilical cord hemangioma accompanied by such pathological changes of placenta.

Case Report

A 26-year-old Chinese woman, G1, P0, was sent to the present hospital emergency department due to premature rupture of fetal membrane. She was at 34 weeks gestation, and fetal movement had increased for three days before the emergency of fetal membrane rupture. No fetal intrauterine growth retardation (IUGR) and fetal distress were diagnosed before admission. Ultrasound examination identified a cystic mass of 2.5 cm in diameter located in the middle of the umbilical cord. The premature male infant weighing 2,100 grams was born vaginally. The baby was then followed with neonatal healthcare examination routinely after birth. No congenital abnormality and visceral hemangioma were developed during the follow-up for three years.

The placenta weighting 540 grams and measured 20×17×4.5 cm in size. No macroscopic abnormalities were observed. The umbilical cord was inserted eccentrically and measured 24 cm in length. Around 10 cm from the placental insertion, an enlarged portion with 3 cm in length and 2.5 cm in diameter was identified (Figure 1). The rest of the cord was slightly edematous and swollen. Microscopically, a well circumscribed nodule was identified. The nodule was composed of numerous blood vessels of varying sizes embedded in myxoid matrix resembling Wharton's jelly (Figure 2A). The lumen of vessels collapsed with few blood cells observed. Most blood vessels were thin-walled capillaries, and a few of them were relatively thick-walled with layers of smooth muscles surrounded (Figure 2B). The original placental arteries and veins were largely compressed and pushed to one side by the mass, and they were found only at the margin of the cord

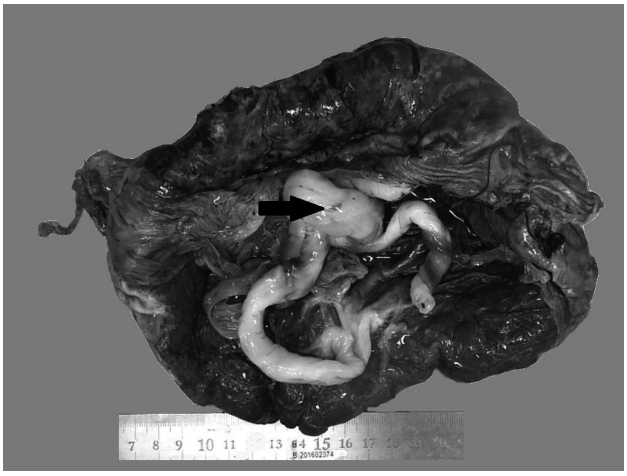

Figure 1. — Macroscopic appearance of the placenta and umbilical cord. An enlarged portion (arrow) can be noticed on the umbilical cord.

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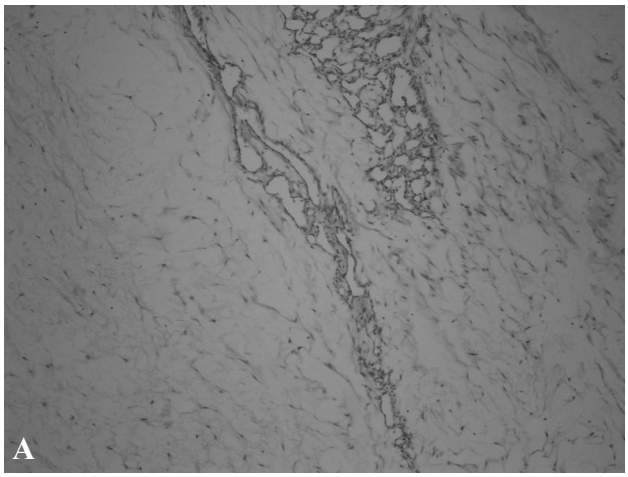

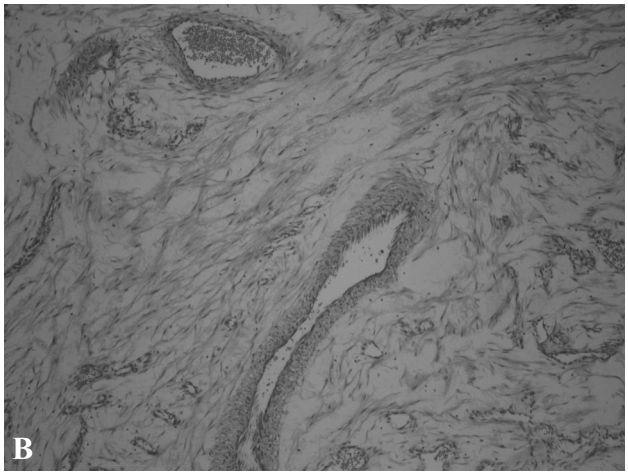

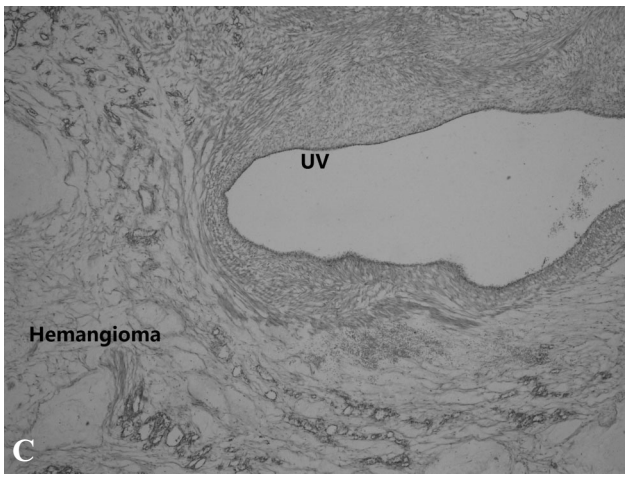

Figure 2. — Microscopic appearance of the umbilical cord hemangioma. (A) Numerous blood vessels of varying size embedded in myxoid matrix resembling Wharton's jelly. (B) A few of blood vessels are thick-walled and surrounded by smooth muscles. (C) Umbilical vein is compressed by the hemangioma (UV: umbilical vein).

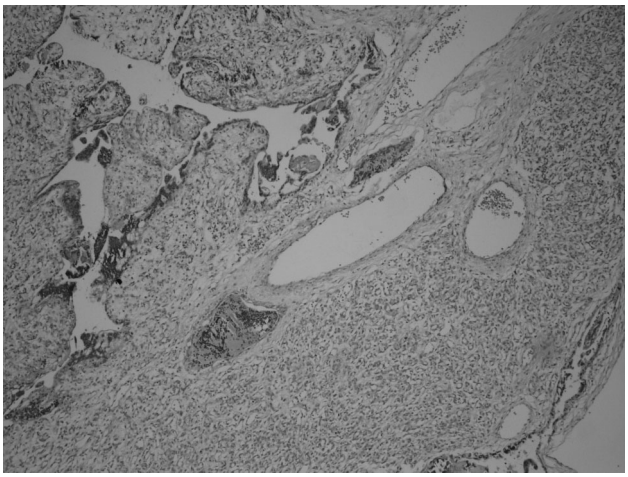

Figure 3. — Segmental chorangiomatosis of placenta.

sections. Fibrosis was clearly identified in the nodule matrix (Figure 2C). No thrombosis, hemorrhage, necrosis, and calcification were seen in the stroma. The placental sections demonstrated abnormally vascularized mature stem villi in two slides and in total, eight stem villi were affected. These stem villi were enlarged, multiple vascular channels were noticed in them (Figure 3), and the placental lesions were not in contact with the umbilical cord lesion described above. Immature villi development was also observed in the placental sections. Placental calcification and chorioamnionitis were absent in this case. The diagnosis of umbilical cord hemangioma accompanied with segmental chorangiomatosis of placenta was then set up based on these pathological findings.

Discussion

Hemangioma of umbilical cord is a benign tumor with extremely low occurrence. In total, 38 cases have been reported until now [1-9]. The pathogenesis of the tumor remains unclear. There is a hypothesis that the hemangioma is essentially a malformation of primitive angiogenic mesenchyme of developing cord. It is in fact a hamartoma, rather than a true tumor [3]. It is suggested that they originate from one or more umbilical major vessels, and most frequently, from the arteries. This was verified by the microscopic findings of direct signals between these vessels [1]. In rare cases, hemangiomas at the fetal end of cord are separated from the main umbilical vessels. They may arise from the vitelline capillaries [4].

Umbilical cord hemangioma can be detected during pregnancy by ultrasound and manifested with a hyperechogenic mass around the base of the cord, however, due to the interference of the cord edema and the degeneration of Whaston's jelly, the prenatal diagnosis can be tricky [4]. Meanwhile, the fact that the hematomas and teratomas of

the umbilical cord share similar features in ultrasound scan should be taken into consideration for differential diagnosis prenatally [4]. Macroscopically, the cord hemangioma consists of an angiomatous nodule ranging from 0.2 to 18 cm in size. In rare cases, it involves the full length of the cord [5]. The nodule is clearly delineated from surrounding tissue and accompanied by edema and cystic degeneration of Wharton's jelly [4]. Microscopically, the nodule is composed of thin-walled anastomosing blood vessels embedded in myxoid stroma. The vascular lumen may be dilated and filled with fibrin thrombi, which is associated with the degeneration and necrosis of endothelium. The vessels are often surrounded by a variable amount of smooth muscle. Calcification and osseous metaplasia of stroma can be observed occasionally, and acute inflammation may be present, sometimes resulting in further degeneration and cystic change of the stroma [3, 4]. Furthermore, although reported to be rare, the tumor may press against the stroma, leading to significant thinning of umbilical vessels [1]. Rupture of hemangioma was reported to cause the massive fetal hemorrhage and the intrauterine fetal death [3, 6]. In the present case, the umbilical arteries and veins were noticeably pressed by the hemangioma, which may impede the hemodynamics of cord. The obstruction of blood circulation in umbilical cord may further lead to the fetal restriction, which was indicated by abnormal fetal movement at early stage.

The pathological changes of the placenta accompanied with umbilical hemangioma are uncommon. Most cases reported so far had no pathological findings in placenta, and only two cases demonstrated chorangiosis [1] and avascular villi [7], respectively. There was a hypothesis that the changed intramural pressure caused by cord hemangioma led to the development of chorangiosis and avascular villi [1,10]. In the present case, placenta chorangiomatosis was observed, which is an extremely rare finding and has not been reported in the literature. Placenta chorangiomatosis is vascular abnormality in chorionic villi, demonstrated by multiple vascular channels in abnormally vascularized mature stem villi [11]. Although the pathogenesis is still unclear, it has been discussed in some literatures that chorangiomatosis may be considered as an intravillous arteriovenous-capillary malformation, which is reported to be associated with several maternal factors, such as multiple gestation, hypoxia, and gestational hypertensive disease [11]. It can be further divided into three groups, focal if only one to five villi are involved, segmental if more than five villi are affected, and multifocal if it affects multiple areas of placenta [11]. In the present case, around eight villi were affected across sections, so the diagnosis of segmental chorangiomatosis was confirmed. Placental chorangiomatosis and cord hemangioma, developed in the early stage of vasculogenesis of placenta and umbilical cord respectively, are both considered as vascular malformations. It is suggested that the chang in blood pressure of placenta caused by the hypertensive disorder of pregnant women may play a role in the development of chorangiomatosis [12]. It is possible that the cord hemangioma may compress the umbilical vessels and change placental hemodynamics, and therefore contribute to the development of chorangiomatosis.

The umbilical cord hemangioma has been reported to be associated with hydramnios, premature delivery, and increased perinatal mortality, especially intrauterine deaths [4]. However, the clinical significance of umbilical cord hemangioma combined with segmental chorangiomatosis has not been established due to the deficiency of cases. Hypothetically, the mechanical compression of umbilical blood circulation by the mass resulting in the reduction of blood supply to the fetus may be the cause of the preterm labor and fetal demise. Moreover, the cord hemangiomas were reported to associate with infantile hemangiomas in some cases, suggesting a possible existence of an underlying congenital predisposition to the vascular malformations in fetuses [4, 9]. While the clinical significance of chorangiomatosis is still obscure, some researchers suggested that chorangiomatosis had no clinical correlations [11]. Meanwhile, according to other literatures, focal and segmental chorangiomatosis were associated with premature delivery, while multifocal chorangiomatosis was related to some adverse clinical outcomes of fetus such as congenital anomalies, IUGR, and even fetal demise [8, 11]. In the present case, premature delivery may be considered as a result of both cord hemangioma and segmental chorangiomatosis. However, no evidence of infantile vascular malformations was found during the follow-up for three years.

Given the possible intrauterine and post-natal complications of infant caused by cord hemangioma, the ultrasound examinations combined with color flow Doppler should be emphasized in the initial diagnosis to detect the umbilical circulation abnormalities. Once the increase of blood flow resistance is identified, the case should be treated with caution. If the maturity of infantile pulmonary is assumed, induced labor should be taken into considerations to avoid of possible intrauterine vascular accidents [4].

Conclusion

Umbilical cord hemangioma is a benign tumor with extremely low occurrence. The placental pathological changes accompanied with cord hemangioma are even rarer. As far as it is known, this is by far the first case reported to be umbilical cord hemangioma associated with placental chorangiomatosis. The present authors offer a hypothesis that the changed placental hemodynamics caused by the existence of cord hemangioma may be responsible for the development of chorangiomatosis, which needs to be proved by more cases and more intensive studies in the future. Meanwhile, due to the possible intrauterine and post-natal complications caused by the cord hemangioma,

early diagnosis and close monitoring of umbilical circulation are vital, which requires high vigilance and deep understanding of the obstetrician of this rare disease.

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