Chorangiosis placenta with 5-vessel umbilical cord with omphalomesenteric duct remnant: An unusual association

Omfalomezenterik kanal kalıntıları olan 5-damarlı umbilikal kordon ile koranjiyozis plasenta: Sıradışı bir ilişki

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Abstract

Placenta is an organ that is responsible for nourishing and protecting the fetus during pregnancy. Histologic examination of the placenta can yield significant information about pre-uterine and uterine conditions affecting fetal growth. Chorangiosis is defined as the presence of ≥10 terminal villi, each containing ≥10 capillaries per terminal villus in ≥10 low power (x10) fields in at least 3 or more random, non-infarcted cotyledons of the placenta. Chorangiosis is an adaptive response to in-utero hypoxia and its presence signifies better pregnancy outcomes. Abnormalities that lead to multiple cord vessels are rare with the majority of reported cases highlighting four vessels due to a persistent right umbilical vein. We report here a case of chorangiosis of placenta associated with a rare 5-vessel umbilical cord and omphalomesenteric duct remnant. To the best of our knowledge, this is the first case report to document such an association.

Keywords: Placenta, umbilical cord, chorangiosis

Öz

Plasenta, gebelik sırasında fetüsün beslenmesinden ve korumasından sorumlu bir organdır. Plasentanın histolojik incelemesi, fetal büyümésini etkileyen ön uterin ve uterinin durumları hakkında önemli bilgiler verebilir. Koranjiyozis, büyümede düşük güç (x10), plasentanın en az 3 veya daha fazla rastgele, enfarkt olmayan kotiledonlardaki ≥10 alanda, her biri ≥10 kapiller içeren ≥10 terminal villus bulunmasıyla tanımlanır. Koranjiyozis iç uterin hipoksisin uyaran bir yanıtıdır ve varlığı daha iyi gebelik sonuçları anlamına gelir. Çoklu kordon damarlarına yol açan anormallikler nadirdir, bildirilen vakalarda çoğunlukla persistan sağ umbilikal vene bağlı dört damar vurgulanmaktadır. Bu çalışmada, nadir görülen 5 damarlı umbilikal kordon ve omfalomezenterik kanal kalıntıları ile ilişkili bir koranjiyozis plasenta vakası sunulmuştur. Bildiğimiz kadardan bu çalışma, böyle bir ilişkiye belgeleyen ilk vakası raporudur.

Anahtar Kelimeler: Plasenta, umbilikal kordon, koranjiyozis

Introduction

The placenta is an organ that is responsible for nourishing and protecting the fetus during pregnancy. Histologic examination of placenta can yield significant information about pre-uterine and uterine conditions affecting fetal growth. One of the histo-morphologic features to be evaluated is the vascularity of chorionic villi. The number of vascular channels present in terminal villi ranges from 2 to 6 and if the number increases beyond this then it is termed as 'villus hypervascularity(1)'. The term 'chorangiosis' was introduced by Altshuler in 1984 and is defined as the presence of ≥10 terminal villi, each containing ≥10 capillaries per terminal villus in ≥10 low power (10x) fields in at least 3 or more random, non-infarcted cotyledons of the placenta(2).

PRECIS: We report here a case of chorangiosis of placenta associated with the rare 5-vessel umbilical cord and omphalomesenteric duct remnant.

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Chorangiosis is known to occur as an adaptive response to chronic low-grade placental hypoxia\cite{1}. It has been found to be associated with various maternal, fetal and placental disorders\cite{1}. Rarely, its association has also been demonstrated with umbilical cord anomalies such as true and false knots, long cord, umbilical vein dilatation or thrombosis, nuchal cord, and single umbilical artery\cite{3}. Herein, we report a case of chorangiosis of placenta associated with the rare 5-vessel umbilical cord.

**Case Report**

A 27-year-old female, para-3-live-1, with gestational hypertension and oligohydramnios presented to the gynecologic emergency at 34 weeks’ gestation in labor. Emergency lower segment cesarean section was performed in view of fetal distress. The child was born with a birth weight of 1.6 kg and was stable. There were no congenital anomalies in the child. The placenta was sent for histopathologic examination. On gross examination, the placenta was complete and measured 12×11×3 cm with the attached umbilical cord measuring 18 cm in length. The umbilical cord contained 5 blood vessels (Figure 1a). On microscopy, sections from the placenta revealed ≥10 capillaries each in ≥10 terminal villi in ≥10 non-infarcted areas examined in ≥3 low power (10×) fields of placenta (Figures 1b, c). Immunohistochemically, the capillary endothelial cells showed uniform positivity with CD34, demonstrating more capillaries than were easily discernible using hematoxylin-eosin staining (Figure 2a), and staining for smooth muscle actin (SMA) was negative (Figure 2b). There was no evidence of increased cellularity or fibrosis in the stroma. Sections from hemorrhagic areas showed ischemic necrosis (Figure 1d). Sections from the umbilical cord showed 5 blood vessels; 4 arteries and 1 vein (Figure 2c) and an omphalomesenteric duct remnant (Figure 2d). Placental membranes were histopathologically unremarkable. A diagnosis of chorangiosis placenta with 5 blood vessels and omphalomesenteric duct remnant in the umbilical cord was given.

**Discussion**

Chorangiosis is a very rare entity found in 5-6% of placentas\cite{2}. Its incidence increases with gestational age with more cases found in late preterm (32 to 37 weeks) and term (>37 weeks) pregnancies\cite{1,2}. Its presence has been correlated with fetal morbidity and mortality and congenital malformations as high as 42% and 39%, respectively\cite{1}.

The proposed pathogenetic mechanism of chorangiosis is that chronic hypoperfusion or tissue hypoxemia causes elaboration of vascular endothelial growth factor, platelet-derived growth factor, and transforming growth factor-β by mesenchymal and trophoblastic cells\cite{3}. An alternative hypothesis highlighting the role of macrophage-derived tumor necrosis factor-α has also been suggested\cite{3}. Increased intramural pressure due to umbilical vein obstruction is also thought to play a role in the development of chorangiosis in cases associated with cord anomalies, such as long umbilical cord and thrombosis of vessels\cite{3}.

Chorangiosis is a histopathologic diagnosis. It must be differentiated from placental congestion, tissue ischemia,
chorangioma and chorangiomatosis\textsuperscript{3,4}. In placental congestion, the vasculature is numerically normal. In tissue ischemia, there is shrinkage of the villi. Chorangiomatosis is a heterogeneous, less well-defined lesion with intermediate features between chorangioma and chorangiosis. It has hyperplastic capillaries surrounding larger vessels in the central core of stem villi with increased numbers of loose, poorly cohesive lattices of perivascular bundles of reticulin fibers and circumferential layers of pericytes. These pericytes stain positively for SMA. Also, both chorangioma and chorangiomatosis are seen before 32 weeks of gestation and involve more proximal elements of villous structures, whereas chorangiosis is more common in late preterm and term pregnancy, and is a diffuse process involving the terminal villi.

Its etiology is poorly understood, but it has been found to be associated with various maternal and fetal disorders\textsuperscript{(1)}. Placental disorders such as placental megaly, chronic villitis, acute chorioamnionitis, amnion nodosum, and placenta previa have also been reported in association with chorangiosis\textsuperscript{1,3}. However, contrary to Altshuler's initial description, it is now suggested to be associated with much improved pregnancy outcomes. Recently in 2016, Stanek reported that chorangiosis is an adaptive and protective mechanism against in-utero hypoxia, and abnormal pregnancy outcomes are not a consequence of chorangiosis per se\textsuperscript{(1)}. In 2017, Petersen et al.\textsuperscript{(5)} concluded that chorangiosis is a placental marker of antepartum chronic low-grade hypoxia. In the present case study, the patient had gestational hypertension, oligohydramnios, and delivered a preterm stable child. The indication for cesarean section was decreased fetal heart rate with fetal distress, which might have been due to chronic hypoxia resulting from pre-eclampsia.

Also, in the present case, an association of chorangiosis with a multiple vessel umbilical cord, having 5 blood vessels with an omphalomesenteric duct remnant was identified. Abnormalities that lead to multiple vessels in the cord are rare with the majority of the reported cases highlighting four vessels due to a persistent right umbilical vein\textsuperscript{(6)}. However, occasional case reports mention five or more vessels in the cord in association with conjoined twins\textsuperscript{(6)}. The child in the present case had no congenital anomalies. Also, the literature regarding their possible mechanism of occurrence and significance is sparse.

The omphalomesenteric (vitelline) duct connects the midgut lumen with the yolk sac in the developing fetus. It is often associated with remnants of vitelline vessels, seen in about 7% of umbilical cords\textsuperscript{(7)}. Microscopically, they are lined by cuboidal to columnar epithelium with an intestinal phenotype and may have a surrounding smooth muscle layer.

To conclude, chorangiosis is an important histopathologic sign of fetal injury. It is an adaptive response to in-utero hypoxia and its presence signifies better pregnancy outcomes. To the best of our knowledge, this is the first case report to document the association of chorangiosis with a 5-vessel umbilical cord, whose presence does not always herald an adverse perinatal outcome, and an omphalomesenteric duct remnant.

**Ethics**

**Ethical Committee Approval:** The study was approved by ethical committee of the institute.

**Informed Consent:** Consent form was filled out by all participants.

**Peer-review:** External and internal peer-reviewed.

**Authorship Contributions**

Concept: P.D., Design: P.D., N.G., Data Collection or Processing: N.G., S.A., Analysis or Interpretation: P.D., N.G., Literature Search: N.G., S.A., J.H.G., Writing: N.G., P.D.

**Conflicts of Interest:** There is no conflict of interest among the authors.

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**References**

1. Stanek J. Chorangiosis of Chorionic Villi: What Does It Really Mean? Arch Pathol Lab Med 2016;140:588-93.
2. Altshuler G. Chorangiosis: an important placental sign of neonatal morbidity and mortality. Arch Pathol Lab Med 1984;108:71-4.
3. Gün BD, Barut F, Tanriverdi HA, Özdamar SU, Barut A. Placental Chorangiosis: An Important Pattern of Placental Injury. Gynecol Obstet Reprod Med 2006;12:176-9.
4. M Mathew, S Garg, L Rai, P Kustogi. Placental Chorangiosis – A report of two cases with unusual associations and review of literature. Internet J Gynecol Obstet 2008;11.
5. Petersen SS, Khangura R, Davydov D, Zhang Z, Sangha R. Placental Chorangiosis: Increased Risk for Cesarean Section. Case Rep Obstet Gynecol 2017;2017:1-5.
6. Singh N, Rao S, Sobti P, Khurana N. Multiple vessels in the umbilical cord: a report of four cases. Indian J Pathol Microbiol 2012;55:597-8.
7. Jauniaux E, De Munter C, Vanesse M, Wilkin P, Hustin J. Embryonic remnants of the umbilical cord: morphologic and clinical aspects. Hum Pathol 1989;20:438-62.