

CASE REPORT

Abnormal entrance of the umbilical vein into the liver and fetal thrombotic vasculopathy in a fetus: a rare combined case

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Abstract

The left umbilical vein is lodged on the fissure of the round ligament of the liver, from umbilicus to portal vein, just on the inferior margin of the falciform ligament. We report an anomalous course of the umbilical vein in a 39-week-old deceased male fetus, also exhibiting fetal thrombotic vasculopathy (FTV). In the present case, the umbilical vein entered the liver through a tunnel on its anterior part of the diaphragmatic surface, close to the lower free edge of the liver and next to the falciform ligament. The entrance of the tunnel was covered by Glisson's capsule. The round ligament's fissure was absent. The quadrate lobe was not well demarcated and the visceral surface of the liver had an abnormal appearance. The coexistence of FTV was confirmed histologically. The possibility of a tunnel present for the umbilical vein, instead of a fissure, is of great significance for surgical approaches or for radiological evaluations, even though it is not likely to cause intrauterine fetal death. However, the anomalous course of umbilical vein might be a predisposing factor for FTV, which often leads to intrauterine fetal death.

Keywords: umbilical vein, abnormal course, tunnel, fetal thrombotic vasculopathy, round ligament's fissure, quadrate lobe.

Introduction

The left umbilical vein carries oxygenated and nutrient-rich blood from the placenta into the fetus, initially towards its liver [1]. Part of the blood enters the hepatic circulation *via* the main portal vein, while the rest of it is shunted *via* the venous duct (*ductus venosus*) to the inferior vena cava and then to the right atrium. The umbilical vein extends from the umbilicus to the portal vein through the inferior margin of the falciform ligament and then through the fissure of the round ligament (*ligamentum teres*). Normally, the umbilical vein and the venous duct are obliterated one week after birth and are replaced by the round ligament and venous ligament (*ligamentum venosum*) of the liver, respectively [2].

The round ligament is often used for cannulation in various therapeutic or diagnostic procedures. It is reported that under extreme pressure, such as portal hypertension, it may reopen to allow the passage of blood, but some authors suggest that the vessel actually involved is an enlarged paraumbilical vein [3]. An abnormal course of the umbilical vein is a rare anomaly [4]. We document the course of the umbilical vein through an anomalous tunnel in the liver of a deceased fetus and discuss the coexistence of fetal thrombotic vasculopathy (FTV) [5, 6]. Although an abnormal course of umbilical vein alone is

probably not the cause of intrauterine fetal death, it may be correlated to FTV, a combination which may lead to intrauterine fetal death.

Aim

The aim of the study is to present a case of abnormal course of the umbilical vein through an anomalous tunnel in the liver of a deceased fetus and discuss its correlation with the coexistent FTV. The possibility that this combination could be responsible for intrauterine fetal death is investigated.

Case presentation

Post-mortem examination and autopsy were performed on a 39-week-old male fetus after intrauterine death of unspecified etiology. It was the mother's first pregnancy. She was 30-year-old and initially carried twins but at the 11th week of gestation, there was a missed abortion of one embryo. Ultrasound examinations up to the time of fetal death, revealed no abnormalities of any kind. The mother did not suffer from thrombophilia or antiphospholipid syndrome (APS). No amniocentesis or karyotype were performed.

Post-mortem examination revealed that the umbilical vein entered the liver weighing 65 g, through a tunnel

on its anterior part of the diaphragmatic surface. It was situated next to the inferior margin of the falciform ligament. The ligament was shorter than normally expected and did not extend to the inferior free edge of the liver (Figure 1). The notch of the round ligament (*incisura ligamenti teretis*) and its fissure were not present. It seems that the fissure was transformed into a tunnel, whose entrance was covered by Glisson's capsule and run within the liver to reach the left branch of the portal vein. The demarcation of the quadrate lobe of the liver was not clear (Figure 2). Eventually, the morphological appearance of the liver surface was affected. Despite the abnormal entrance of the umbilical vein in the liver, the dissection revealed its normal connection with the portal vein and the venous duct (Figure 3). No other congenital anomalies were noticed.

The placenta weighing 490 g revealed marginal

insertion of the umbilical cord. The cord was 40 cm in length and was located 1.5 cm from the placental margin. In the maternal surface of the placenta, there were small, sporadic hemorrhagic foci, less than 1 cm in diameter. In the fetal surface, a hematoma, 1 cm in diameter, was observed within an area with congested vessels.

Microscopic examination of multiple sections from the placenta revealed occlusive and organized thrombi within some large chorionic and stem villi vessels. Foci of avascular villi were observed. Occasionally, villi appeared with calcification and endothelial damage. Fibroblast hyperplasia, causing bulging into the lumen, and hemorrhagic endovasculitis were detected (Figure 4). In the somatic part of the umbilical vein and in several somatic vessels, extravasation of red cells and occlusive thrombi were observed (Figure 5). The described findings indicate FTV.

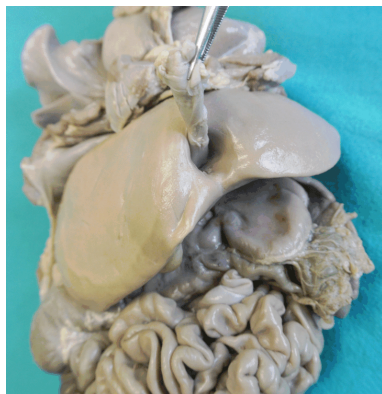


Figure 1 – The umbilical vein enters the liver through a tunnel on its anterior part of diaphragmatic surface. The tunnel is covered by Glisson's capsule.

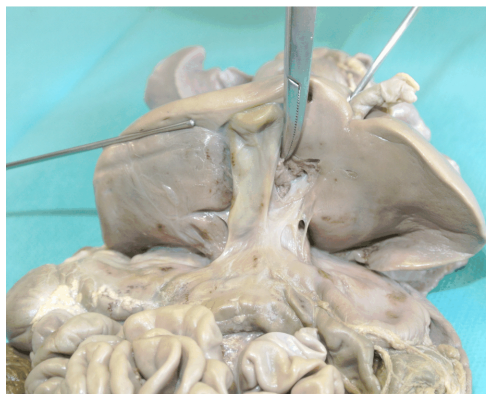


Figure 2 – The notch and the fissure of the round ligament are not present. The demarcation of the quadrate lobe of the liver is not clear. The morphological appearance of the liver surface is generally affected.

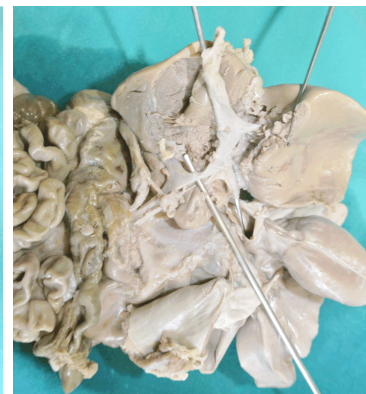


Figure 3 – Normal connection of the umbilical vein with the portal vein.

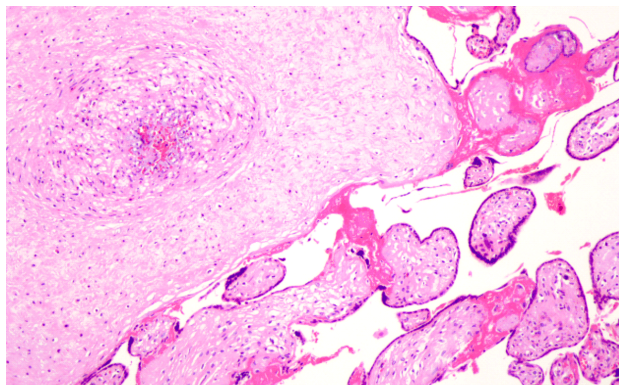


Figure 4 – Hemorrhagic endovasculitis within stem villi vessel, with endothelial damage, fibroblast hyperplasia, causing bulging into the lumen and foci of avascular villi are observed. Hematoxylin–Eosin (HE) staining, ×40.

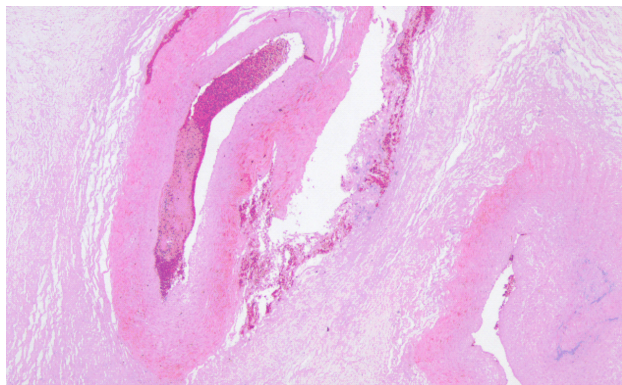


Figure 5 – Extravasation of red cells and occlusive thrombi in an umbilical cord vessel. Wharton's jelly between two umbilical vessels. HE staining, ×40.

Discussions

Few reports exist on anomalies of the umbilical vein's entrance in the liver. These rare reports usually involve a persistent right umbilical vein [7].

Reported anomalies, after birth, on the liver's ligaments or fissures are also very few [2]. Some of them, as in the present case, describe the complete absence of the fissure for the round ligament [8–11]. There are even fewer reports

describing the transformation of the fissure for the round ligament into a tunnel, as in the current case. In one of them, there was no quadrate lobe present and the tunnel was situated on the diaphragmatic surface of the liver [12]. In another report, the round ligament coursed to its final destination through a tunnel, situated exactly where the fissure should be and with the quadrate lobe present, although not clearly delimited due to the tunnel's existence [2].

The liver is normally separated into lobes during the second month of gestation. The presence of the tunnel instead of a fissure for the umbilical vein might result from incomplete separation and fusion of lobes, during the embryonic period [2].

The important role of the round ligament in the management of diseases and in clinical procedures cannot be overlooked [2]. The umbilical vein may remain open long enough after birth, to serve for cannulation in various therapeutic or diagnostic procedures [13]. It may be used for exchange transfusion of blood in cases of fetal erythroblastosis, to prevent severe complications, such as mental retardation, heart failure or even death [14]. It can also be used for medical administration of drugs or chemotherapy [15]. The possibility of a tunnel being present instead of a fissure in the area is of great significance for surgeons and for radiologists who might be confused noticing a cavity in the liver.

The coincidental discovery of the tunnel after birth in a few described cases [2, 12], suggests that it is compatible with life if no additional vascular or other anomalies are present. Although the fetal tunnel was a unique and fascinating discovery, its isolated presence most probably does not explain fetal death.

In the current case, the fetus presented with FTV. Thrombosis of the large chorionic vessels or of the stem villi vessels, cause obstruction in the capillary circulation inside the corresponding villi. Endothelial ischemia and reduced or complete lack of blood flow follow, which probably result in disruption of the capillaries' integrity. Vascularization gradually disappears and chorionic villi are left avascular [5]. These alterations are accompanied by karyorrhexis of the endothelial cells and of the nucleated embryonic blood cells. In addition, the ruptured erythrocytes are extravasated [16].

Similar histological findings may arise from another condition called intrauterine fetal demise. The obstruction of blood circulation in the placenta that follows embryonic death creates diffuse and simultaneously developing histological changes in the placental vessels. The severity of the findings is related to the time interval between fetal death and childbirth [17]. In FTV cases on the other hand, pathological microscopic findings are local and appear asynchronously, depending on the time and extent of thrombosis of large vessels of the umbilical cord, as observed in the presented case. In addition, intrauterine fetal demise does not present with endothelial cushion bulging into the lumen of vessels in chorionic or stem villi. Clots in the fetal vessels' branches have also been described in cases of marginal or membranous insertion of the umbilical cord [5]. Even though the presented case appears with a marginal insertion, this alone cannot be responsible for the severity of the microscopic findings.

FTV has been associated with both thrombotic episodes in neonates and maternal thrombophilia [18, 19]. In our case, the parents did not have thrombophilia or APS, diabetes or polycythemia, which may be implicated with FTV. Redline suggested that fetal vascular stasis related to chronic umbilical cord abnormalities might be a predisposing factor for FTV [6]. According to Redline's hypothesis, the documented marginal insertion of the umbilical cord and the anomalous course of the umbilical

vein could both be the predisposing factors for FTV, as in the present case. To our knowledge, this is the first case reported on a fetus with an anomalous course of umbilical vein through a tunnel, which could be responsible for the presence of FTV, a combination likely leading to intrauterine death.

Conclusions

The anomalous course of the umbilical vein through a tunnel towards the left branch of portal vein within the liver could be of great significance for surgical approaches or for radiological evaluations. Moreover, it may be a predisposing factor, which may lead to FTV, which often leads to intrauterine fetal death.

Conflict of interests

None to declare.

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