Prenatal diagnosis of **umbilical cord hemangioma** associated with chorioamnionitis: **the "wave sign"** as new potential ultrasonographic marker

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Objectives

Umbilical cord hemangiomas are rare benign vascular neoplasms arising from endothelial cells, with less than 45 cases reported in literature and, among these, less than 30 prenatally diagnosed. Sonographically, an umbilical cord hemangioma appears as a homogeneous solid echogenic mass of variable size, usually located near the placental insertion of the cord. Close to the mass there may be cystic degeneration of Wharton's jelly, therefore the lesion may appear quite heterogeneous. In some cases, color Doppler ultrasound can be useful to demonstrate low-flow blood vessels within the echogenic component of the tumor. The differential diagnosis includes pseudocysts, teratomas of the umbilical cord, allantoic cyst, omphalomesenteric duct cysts, anterior abdominal wall defects. We present a case of umbilical cord hemangioma diagnosed in a pregnant woman at 32 weeks of gestation with ultrasonographic finding of extensive detachment of the amniotic leaflet from the placental surface and from the uterine wall (called by us "wave sign"), and histological diagnosis of acute grade 2 chorioamnionitis.

Methods

A 46-years-old woman, gravida 3 para 1, presented to our Obstetrics And Gynecology Department at 32 weeks and 3 days of estimated gestational age for routine third trimester scan. She had no medical history and the current pregnancy was unremarkable except for gestational hypothyroidism in hormone replacement therapy. Preliminary ultrasound assessment showed an heterogeneous mass with solid and cystic areas located within the cord (fig. 1-2), situated near the placental insertion, which measured 7.4 x 2.9 x 4.6 cm. Doppler analysis showed normal umbilical artery pulsatility index and middle cerebral artery peak-systolic velocity. Fetal ductus venous had a regular waveform with a positive a-wave. Fetal size was appropriate for gestational age as well as amniotic fluid assessment. Unexpectedly, we observed an extensive detachment of the amniotic leaflet from the uterine wall, including the placental surface. It appeared like a floating membrane with underlying anechoic fluid (amniotic fluid? Serous fluid? Blood?), whose echogenicity seemed slightly different from intra-amniotic fluid. For that floating appearance, we decided to define it as the "wave sign" (fig. 3-4).
Uterine contractions were recorded during non-stress test with normal fetal heart rate. For these reasons, our patient was admitted to our gynecological department, receiving later intramuscular corticosteroids for fetal lung maturation and magnesium sulfate for fetal neuroprotection. At hospitalization, both vital signs and routine blood tests resulted in range, except for mild leucocytosis (15.85 x 10^3 µL). Rapid nasal swab resulted negative for SARS-CoV2.
Considering uterine contractions associated with the ultrasonographic signs described above, our patient underwent an urgent cesarean section after a few hours after admission due to suspected subclinical placental abruption. The operating record shows blood and clots leakage after hysterotomy and the amniotic sac widely adherent to the fetal body. A vital male fetus in breech

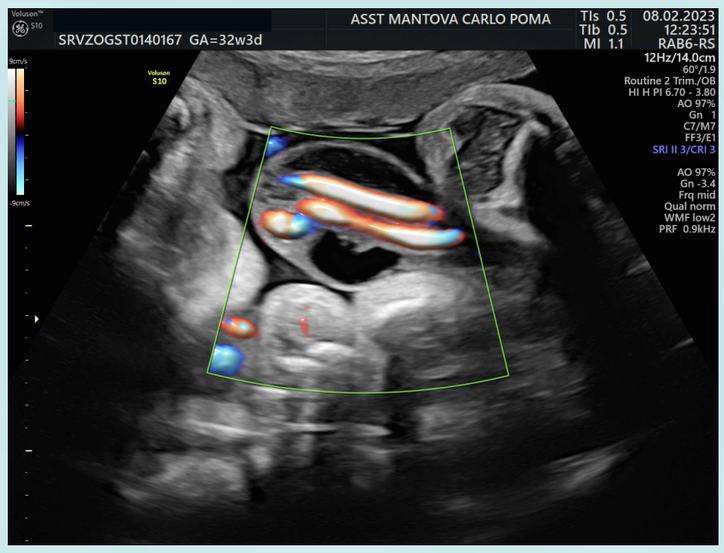

Fig. 1-2 Umbilical cord hemangioma

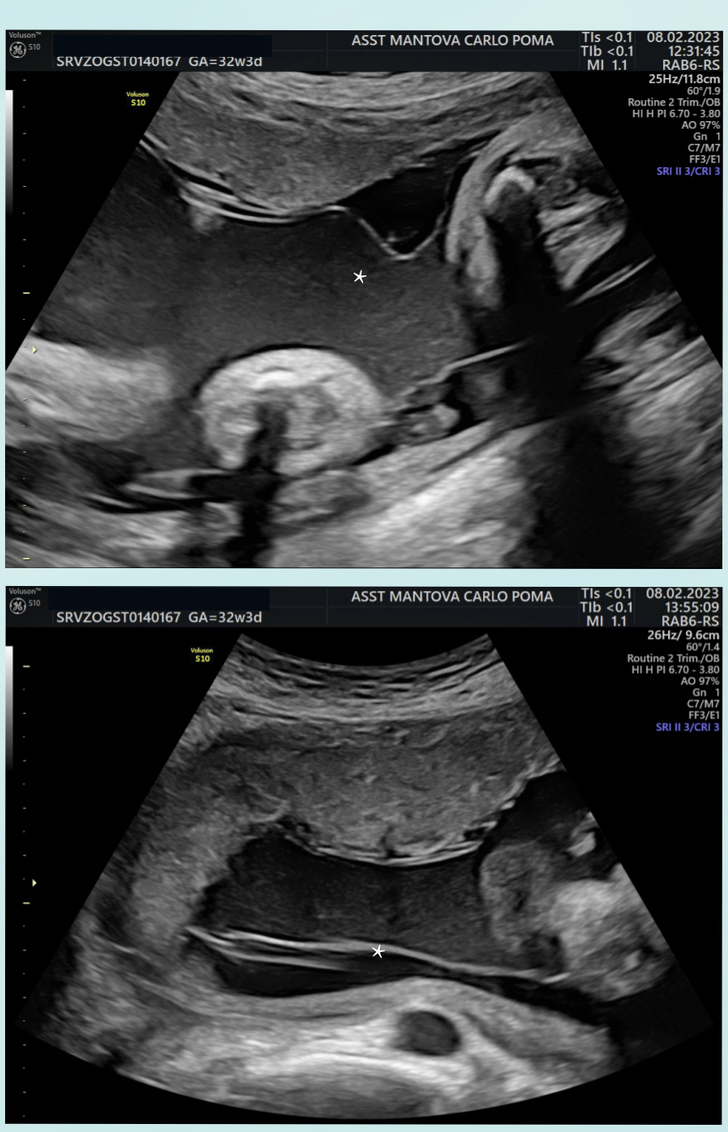

Fig. 3-4 The wave sign (*)

presentation weighing 1840 grams, was then extracted and entrusted to pediatricians' care. Umbilical cord blood gas analysis showed normal fetal oxygenation values. Macroscopic examination of placenta and fetal adnexa (fig. a) showed a paracentral insertion of the umbilical cord, which appeared extremely edematous and thickened; in section, three vessels with fibrotic appearance were recognized. In addition, a blood collection of about 2 cm was detected at the placental insertion of the cord. Maternal side of the placenta showed a spongy cotyledons' appearance, congested chorionic vessels and opaque membranes. Histological examination of the placenta detected fibrous and edematous stem villi with intimal and medial hyperplasia, hyporamification and congestion of the intermediate and terminal villi; a subchorionic fibrinoidosis with congested and ectatic chorionic vessels was also reported. On histological examination, the umbilical cord was markedly and widely edematous, with Wharton's jelly myxoid degeneration (fig. b). In addition, the histological examination confirmed our ultrasound diagnosis, reporting the presence of a hemangioma involving the terminal tract of the umbilical cord (fig. c) associated with a widespread concentric fibrous perivascular thickening of the arteries and umbilical vein (fig. d). Interestingly, a not clinically suspected acute grade 2 chorioamnionitis, maternal stage 2 (fig. e) was also histologically detected.

Results

Due to the rarity of hemangiomas, it is hard at present to define their association with pathological fetal features as well as the weight of these associations. In our report is described, for the first time, the presence of an extensive detachment of the amniotic leaflet from the uterine wall and the placental surface, the so-called "wave sign", together with a hemangioma of the umbilical cord of significant dimensions and typically located near to the placental insertion, site reported as more frequent in the cases available in literature. Furthermore, it is worth noting the incidental finding, in the histological examination of the placenta, of a setting of acute grade 2 chorioamnionitis on the maternal side of the placenta. In absence of microbiological or clinical evidence of an infectious damage, we could speculate this situation as a consequence of an inflammation induced by the hemangioma. Obviously, this hypothesis requires further studies to be confirmed, but it opens up new potential paths of research in the context of a disease on which we currently have limited information.

Conclusions

Hemangiomas of the umbilical cord are extremely rare tumors often prenatally undiagnosed. Their detection is especially based on distinctive features, recognized in global literature. In addition, we reported the first case of umbilical cord hemangioma associated with chorioamnionitis and presenting with an original ultrasonographic sign, never described before. Further studies are needed to investigate this correlation and shed light on still uncertain pathogenesis of these tumors.

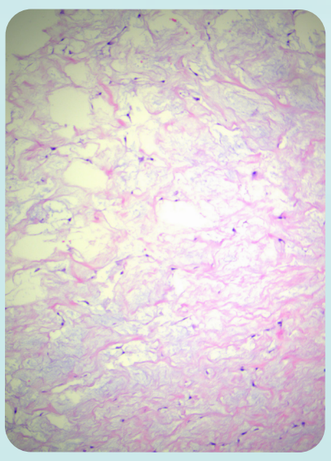

Fig. b Wharton's jelly myxoid degeneration

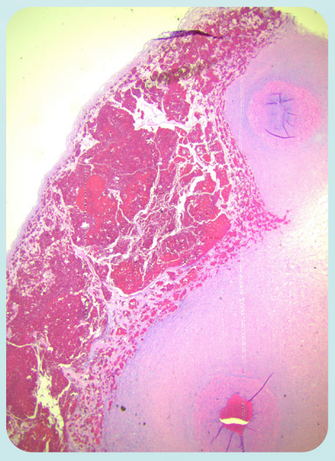

Fig. c Umbilical cord hemangioma

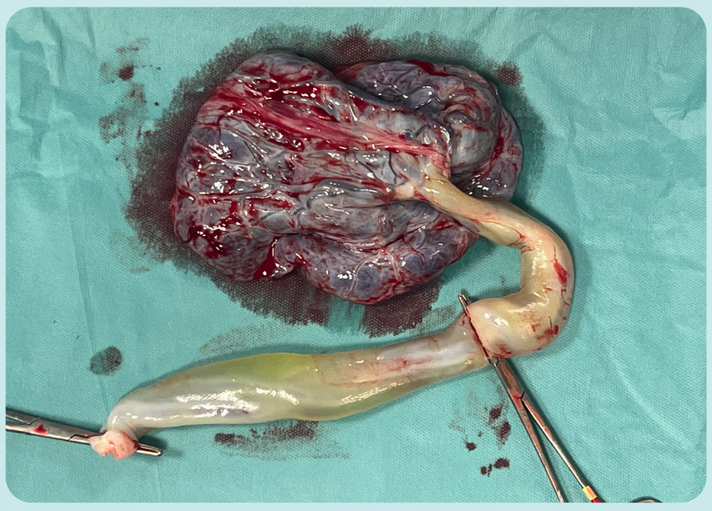

Fig. a Placenta and fetal adnexa

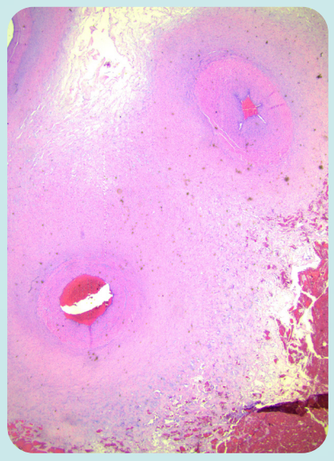

Fig. d Concentric perivascular fibrosis

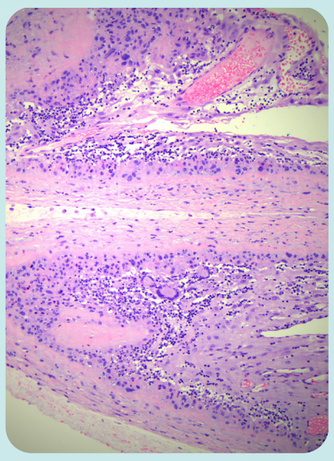

Fig. e Acute grade 2 chorioamnionitis