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Umbilical cord hemangioma

APRIL 2007



A 35-year-old G3/P3 was referred to the obstetric department at 33 1/7 weeks of gestation for prenatal assessment of an echogenic mass of the umbilical cord detected during a routine ultrasound examination. Chorionic villous biopsy for advanced maternal age at 13 weeks of gestation had revealed a normal 46, XX karyotype. Sonographic assessment of the fetus demonstrated growth parameters appropriate for gestational age. The abdominal wall was closed, with an inhomogeneous mass measuring 43 x 32 x 28 mm near the insertion of the umbilical cord (Fig. 1 and Fig. 2). The cardiotocogram showed no fetal distress. Doppler flow studies of the umbilical cord showed three vessels, displaced by a multilobulated echogenic tumor (Fig. 3). Maternal serum screening showed an elevated level of alpha-fetoprotein (326.2 mcg/l, reference < 10 mcg/l), but no signs of congenital infection. Cervical screening for streptococcus B at 35 weeks gestation was positive, leading to peripartal antibiotic prophylaxis.



Fig. 1

Fetal ultrasound examination: fetal abdominal wall with umbilicus. The arrow marks the umbilical vein passing through the abdominal wall (asterisk: umbilical cord tumor, double asterisk: edematous Wharton's jelly).

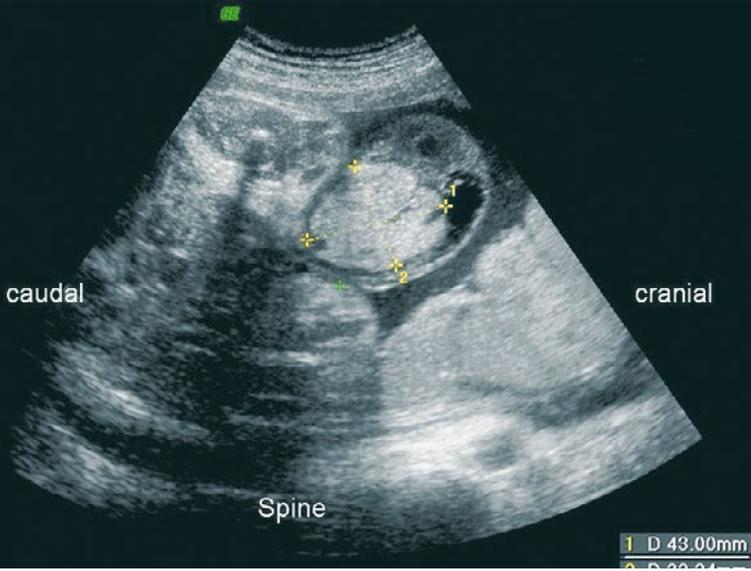


Fig. 2

Fetal ultrasound examination: dimensions of the umbilical cord tumor. (1:D43.0mm, 2:D 32.2mm).

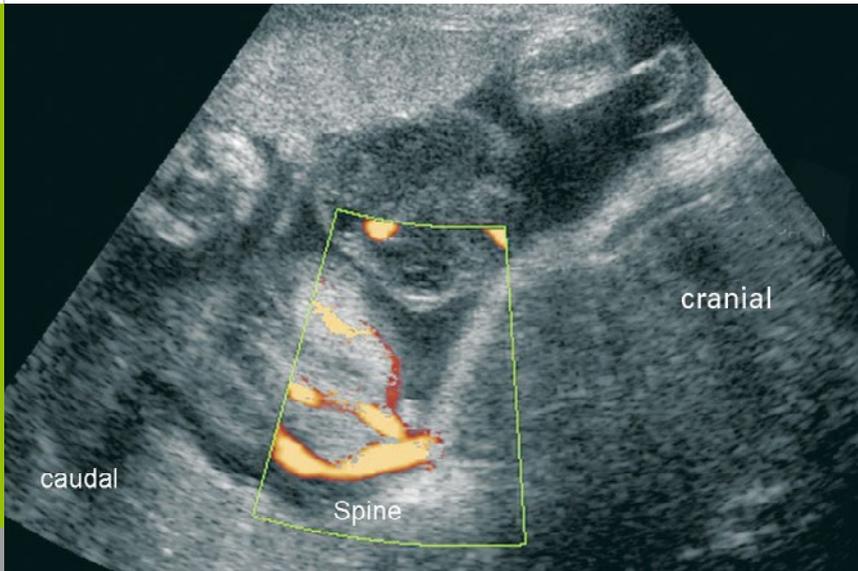


Fig. 3

Fetal ultrasound examination: umbilical tumor with three vessels passing through it.

After induction of labor at 38 5/7 weeks of gestation, a baby girl was delivered vaginally, weighing 2840 g (P 10-25), length 47 cm (P 10-25), head circumference 33 cm (P 10-25). She adapted with Apgar scores of 8 at 1 minute, 9 at 5 minutes, and 9 at 10 minutes. Arterial cord pH was 7.31.

Surgical revision was undertaken for the suspected diagnosis of a "hernia into the cord". However, no defect of the abdominal wall was found. An ordinary surgical resection of the umbilical cord with an umbilicoplasty was performed. The maximum diameter of the surgically removed umbilical cord segment was 3.6 cm. Macroscopically, the

cross section revealed three vessels. Only on microscopic examination did it become clear that Wharton's jelly was mostly replaced by abundant aggregates of thin walled capillaries (Fig. 6). The capillary hemangioma showed a diffuse growth around vessels, with focal dissection of the muscular coat of the umbilical vein. In addition, nodular aggregates were seen, some of which were freshly thrombosed (Fig. 7). Distal to the lesion, the remaining umbilical cord measured 20 cm in length and 1.2 cm in diameter and was unremarkable. Postoperative recovery was uneventful. Subsequently, the girl developed a small supraumbilical hernia which was successfully operated three months later.

Appearance of the umbilical cord shortly after birth.



Fig. 4



Fig. 5

Nodular bulging of the umbilical cord.

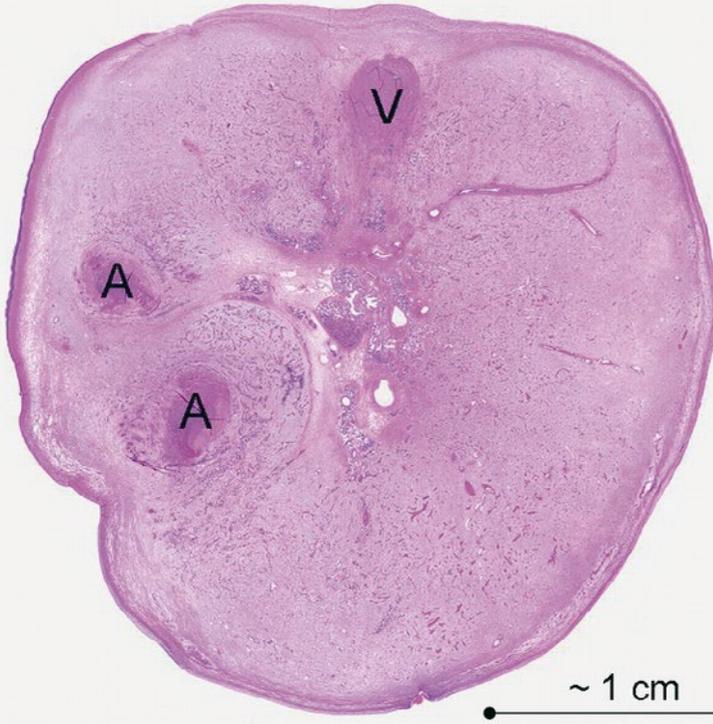


Fig. 6

Cross-section of umbilical cord showing three vessels with muscular coats, two arteries (A) and one vein (V). Most of the Wharton's jelly is replaced by capillaries. Hematoxylin-eosin, magnification x10.

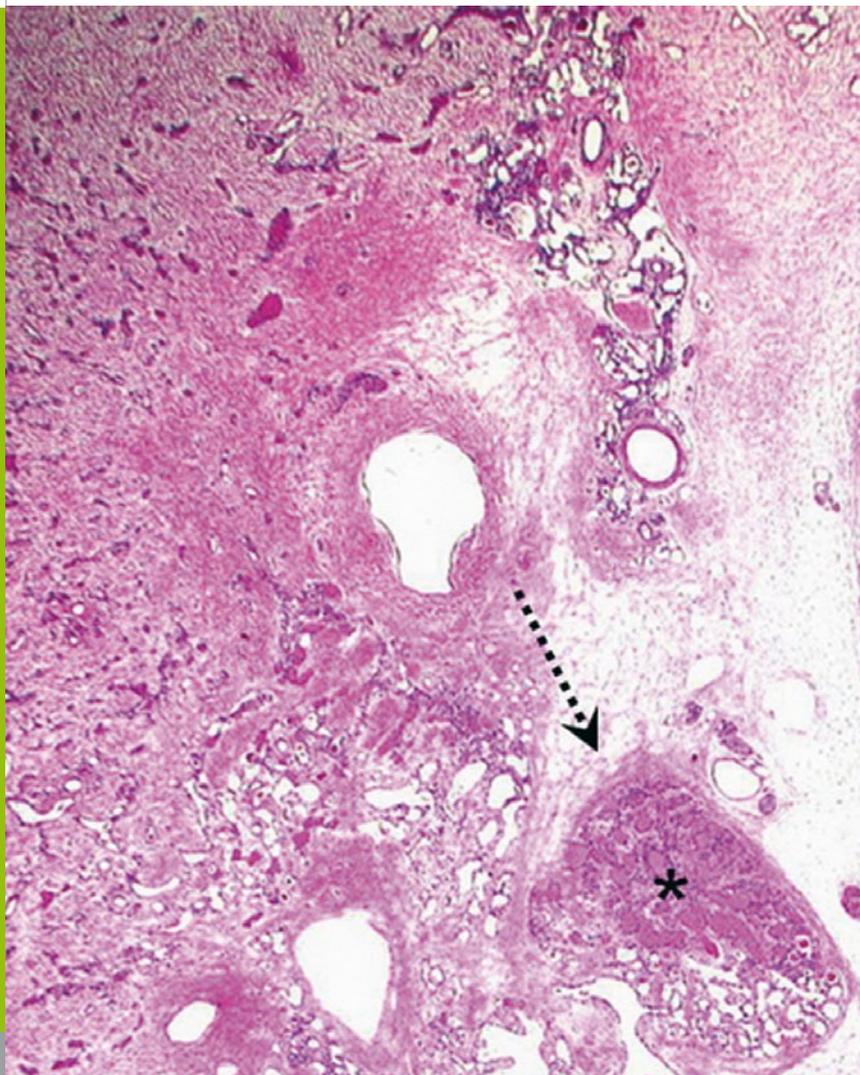
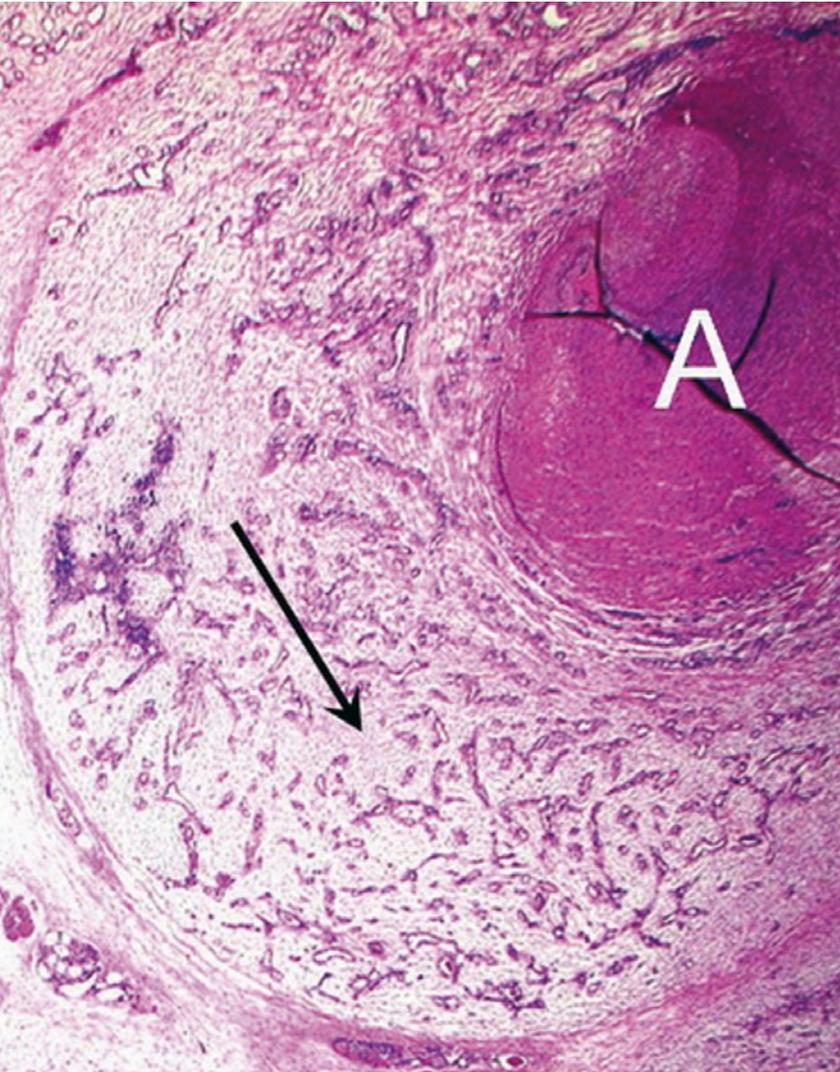


Fig. 7

Cross-section of umbilical cord showing an umbilical artery (A). Note the abundance of tiny capillaries around the artery (thick arrow). Some capillary growth is nodular (dotted arrow) with occasional fresh thromboses (asterisk) (Hematoxylin-eosin, magnification x100).



DISCUSSION

Hemangioma of the umbilical cord is a rare vascular pathology characterized by capillary endothelial proliferation, originating from the umbilical arteries or, in some instances, the umbilical vein or vitelline capillaries (4). The etiology is still not clear; it may represent a true neoplasm or it may be a developmental abnormality (hamartoma). We reviewed 37 reports in the literature from 1951 through 2005, in most of which the lesion was referred to as hemangioma, and rarely as angiomyxoma, myangioma, or hemangiofibromyxoma because of the associated myxoid appearance of the edematous Wharton's jelly. A hemangioma usually consists of a nodular tumor of polymorphous presentation, ranging from 0.2 to 18 cm in diameter, derived from one of the umbilical vessels, and tending to develop proximally to the placenta, which, even though it lacks circumscription or encapsulation, never metastasizes (2), although it has been reported to be associated with additional hemangiomas. An edematous degeneration of Wharton's jelly is commonly observed proximal to the lesion (1), occasionally extending distally as well. The abdominal insertion of the umbilical cord is normal. Histologically, although possessing no fibrous capsule, hemangiomas are well-defined aggregates of closely-packed, thin-walled capillary vessels, filled with blood, and separated by scant connective tissue stroma. Some of the capillary lumina may be partially or completely thrombosed.

Diagnosis	Macroscopic appearance	Association with other findings
Umbilical cord teratoma	Nodular or cystic Max. diameter 10 cm Normal umbilical cord origin All along the umbilical cord	50% associated with other malformations
Hemangioma of the umbilical cord	Nodular Diameter 0.2 to 18 cm Normal umbilical cord origin Proximal to the placenta	Mostly isolated finding Edema of Wharton's jelly Complication: intrauterine bleeding Hernia into the cord Nodular
Hernia into the cord	Nodular Diameter < 4 cm Normal umbilical cord origin Proximal to the abdomen Elastic consistence	Mostly isolated finding Associated with Meckel's diverticulum
Omphalocele	Abdominal organ prolapse Diameter > 4 cm Large umbilical cord origin	Associated with other malformations, syndromes, or chromosomal abnormalities
Umbilical cord cyst	Nodular or cystic Most often single Normal umbilical cord origin All along the umbilical cord	Mostly isolated finding
Umbilical cord polyp	Nodular, solid, reddish, exudative, sometimes ulcerated and bleeding	Meckel's diverticulum and umbilical enteric fistula
Umbilical cord hematoma (A) and/or aneurysm (2)	A: nodular, livid B: cystic in US, turbulent, non-pulsatile blood flow	A: fetal distress B: trisomy 18, single umbilical artery, arteriovenous fistula

Table 1

Differential diagnosis of an umbilical cord tumor.

Most often, a hemangioma of the umbilical cord is an isolated anomaly, but large lesions have been described in association with polyhydramnios (3), intrauterine growth restriction (1), elevated maternal serum alpha-fetoprotein (4), fetal malformations such as anencephaly (3), and malformations of the gastrointestinal tract. Furthermore, some authors have also reported its association with fetal death caused by impaired umbilical circulation resulting in nonimmune hydrops fetalis (5), torsion, compression, or stenosis of the umbilical vessels (1,4), fetal hemorrhage (1), thrombosis of an umbilical vessel, and hematoma of the umbilical cord (6). Some of these conditions were also associated with premature delivery. A possible hereditary predisposition to this vascular anomaly is still under discussion.

Nodular bulges of the umbilical cord are rare entities of polymorphous presentation that can be detected prenatally by ultrasound examination. Their differential diagnosis and some of the respective characteristics are listed in Table 1, a more detailed review of which is beyond the scope of this report. The clinical significance common to all of these anomalies is determined by their size, which can potentially cause vascular compromise and affect fetal growth. After birth, umbilical cord clamping should be distal to the lesion to avoid intestinal strangulation. Ultrasound examination of the lesion is recommended and referral of the newborn

to a pediatric surgery department for revision and correction is mandatory but not an emergency.

We report the case of an infant with an umbilical cord tumor which had twice been misdiagnosed previously: antenatally by ultrasound, as a teratoma, based on its inhomogeneous character and the absence of intralesional blood flow, and postnatally as a hernia into the cord. The definitive diagnosis of a hemangioma of the umbilical cord was made by histopathology following surgical resection.

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CONCLUSIONS

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