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Case report

An unexpected association – cord hemangioma and patent urachus

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Abstract

A patent urachus is the presence of structures found early in pregnancy, which could be associated with other abnormality in 46% cases.

Hemangiomas are benign tumors and are more common in the skin. Umbilical cord hemangiomas are very rare, with fewer than 50 cases described in the literature.

Here we report a clinical case of a neonate with a prenatal diagnosis of an umbilical cord hemangioma and a patent urachus. We would like to draw attention to the differential diagnosis of cord masses, as well as to the importance of early diagnosis and the appropriate management of the newborn. To our knowledge, this is the first case reported of a newborn diagnosed with this association.

Keywords

Patent urachus, hemangioma, benign tumor, umbilical cord, prenatal diagnosis, cord mass.

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Introduction

A patent urachus is the persistence of structures seen early in pregnancy. Where the connection between the bladder wall and the umbilical cord persists, this may result in different types of urachal remnants [1]. Patent urachus can be associated with other anomalies in 46% cases, such as abdominal wall defects, bladder exstrophy, and lesions of the cord like hemangiomas [2].

Hemangiomas are benign endothelial cell tumors that can appear in several organs, although they occur more frequently on the skin [3]. On the other hand, umbilical cord hemangiomas are very rare, with less than 50 cases described in the literature [3, 4].

We describe a clinical case of a newborn with a prenatal diagnosis of a concomitant umbilical cord hemangioma and a patent urachus, who survived after the neonatal period without sequelae. Although a cord hemangioma is a rare condition, it should be considered in the differential diagnosis of cord masses.

We would like to highlight the value of accurate prenatal monitoring, because early diagnosis and proper management of the newborn can change the prognosis. To our knowledge, this is the first case report of a newborn diagnosed with these conditions.

Case report

A female newborn of a 37-year-old primigravida with an uneventful prenatal follow-up until 30 weeks of gestational age. The ultrasound assessment revealed an echogenic mass (29 x 29 mm) in the umbilical cord (**Fig. 1**, gray arrow), likely including intestinal loops inside. Also, at a more proximal position, another structure (with 36 mm or greater diameter) with hematoma features was identified (**Fig. 1**, black arrow). Fetal karyotype was 46,XX.

An elective C-section was performed at 38 weeks of gestational age. The Apgar score was 8/8/10 at $1^{st}/5^{th}/10^{th}$ minutes of life, respectively. The newborn was adequate for gestational age (weight 2,885 g; length 46 cm and head circumference 32.5 cm).

The physical examination did not present any relevant findings besides a firm, non-reducible mass (3 x 3 cm), in the proximal portion of the umbilical cord (**Fig. 2**).

A few hours following birth, the newborn underwent surgical intervention due to the possibility of the presence of incarcerated intestinal loops. A hernia of the umbilical cord, associated with persistence of the urachus, and a solid lesion externalized by the defect and extending through the anterolateral wall of the bladder were described.

The extra-bladder lesion was removed, and the injury was closed (**Fig. 3**). An intraoperative

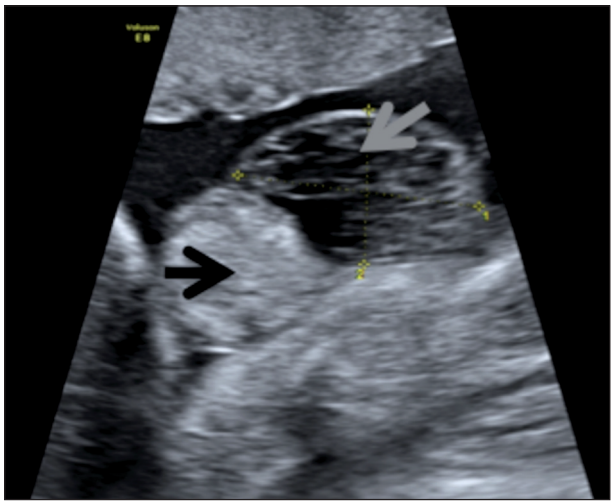

Figure 1. Ultrasound image at 30 weeks.

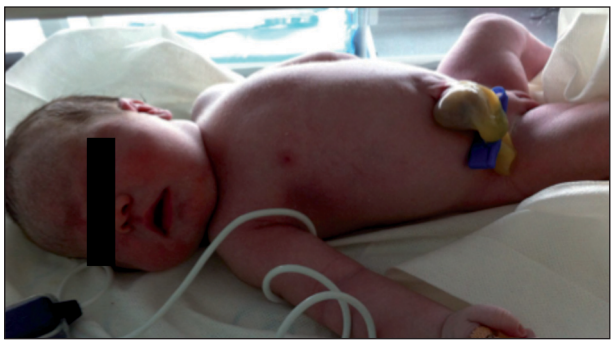

Figure 2. The newborn in the Neonatal Intensive Care Unit (NICU) with a visible mass in umbilical cord.

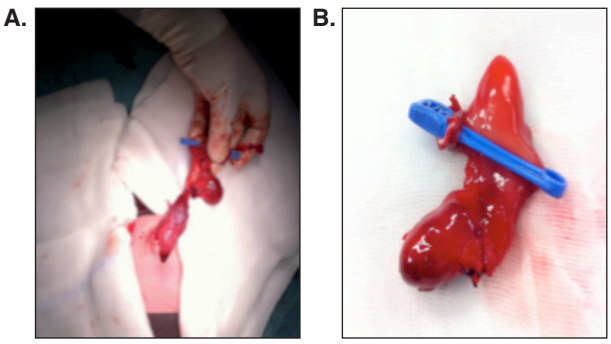

Figure 3. Excised surgical part.

cystography was completed, and no impairment was identified.

The newborn was later admitted to the Neonatal Intensive Care Unit (NICU) and had a good clinical evolution, with normal urine output.

Laboratory tests, including renal function and alpha-fetoprotein, were normal. Transfontanellar, cardiac, renal and bladder ultrasounds were unremarkable. She was discharged from the NICU at 16 days old with good healing of the surgical wound.

The histologic examination of the surgical specimen was consistent with an umbilical artery hemangioma and a patent urachus.

At 24 months of follow-up, the patient was asymptomatic, alpha-fetoprotein levels remained normal and no renal or bladder abnormalities were identified in the ultrasonographic evaluation.

Discussion

Embryologically, the urachus derives from the allantois and allows communication between the bladder wall and the umbilical cord. Usually, this connection is obliterated by 6 weeks of gestational age and gives rise to the median umbilical ligament [1, 5]. When this does not occur, different types of urachal remnants can be identified, such as complete patency or vesicoumbilical fistula, vesicourachal diverticulum, urachal sinus and urachal cyst [1]. The clinical manifestations of patent urachus vary from asymptomatic to recurrent omphalitis, urinary tract infections, calcifications and infrequently carcinoma [5].

Patent urachus has an estimated prevalence of 0.25:10,000 births, with boys being affected twice as much as girls [5].

Umbilical cord hemangioma is a rare condition [3, 4], with an estimated incidence of 1 in 5,500-11,000 deliveries [6]. Prenatal diagnosis is possible, but not always easy [3, 7], as reflected in this case. The fetal ultrasound described an echogenic mass, possibly containing intestinal loops, supporting the hypothesis of an abdominal wall defect. Additionally, a formation on the more proximal segment of the umbilical cord was also reported, raising the question of whether it was a cord hematoma.

Thus, in the context of an umbilical mass, the differential diagnosis must include hemangiomas, hematomas of the umbilical cord, teratomas, metastatic neuroblastomas, placental masses, herniation into the cord, varicose veins, aneurysms, omphalomesenteric duct cysts, and abdominal wall defects [3, 7].

The prenatal identification of a hyperechogenic cord mass associated with cord edema usually occurs in prenatal care [3, 7, 8]. Close prenatal follow-up is required to monitor the growth of the mass and the well-being and growth of the fetus [4, 7].

After birth, an attempt was made to reduce the umbilical cord mass, thought to be a hernia. During the surgery procedure, the solid mass was described macroscopically as a hemangioma associated with the persistence of the urachus. This histologic report confirmed the clinical suspicion.

Usually, these neoplasms occur as isolated abnormalities, but they have been known to present in association with other malformations, such as cardiac malformations, anencephaly, and syndromes with cutaneous or systemic hemangiomas [3, 7].

Hydramnios, fetal hydrops, and increased levels of the serum alpha-fetoprotein are also associated with cord hemangiomas [3, 7]. These findings can increase the risk of perinatal death and morbidity. The prognosis of a cord hemangioma is not fully understood [7].

Reduced umbilical circulation, spontaneous bleeding, and umbilical cord torsion are suggested as possible causes of death [3, 7, 8]. Morbidity and mortality around 35-50% have been reported [6, 9]. In this case, no complication was detected.

Management of these patients is not consensually established. They should have a close follow-up because of the risk of development of other vascular lesions [9]. The need for prophylactic excision of the patent urachus is controversial, although these lesions are usually removed, taking into account the potential risk of neoplastic transformation [1, 5].

The authors hypothesize that the patency of the urachus allowed the growth of the hemangioma to the bladder or, on the other hand, the presence of a growing hemangioma prevented the obliteration of the urachus.

With this case report, we underline the challenge of the prenatal diagnosis of umbilical cord hemangiomas. Excision of the lesion and histopathological examination may be necessary to confirm the diagnosis in some cases. Despite the benign nature of a hemangioma, we should be aware that potential life-threatening complications might occur.

Declaration of interest

The Authors declare that there is no conflict of interest.

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