Isolated Umbilical Cord Cyst: A Case Report
C Lilaia, C Ribeiro dos Santos, L Gonçalves, J Siles Cadilla, C Santos, B Cabrita

Citation

Abstract
The authors report a case of an isolated giant umbilical cord cyst in an healthy newborn, which was not identified in prenatal ultrasound.

INTRODUCTION
Umbilical cord cysts are usually classified as true cysts and pseudocysts. Although they can occur at any location along the umbilical cord, they are usually located towards its fetal insertion. They are irregular in shape and located between the vessels. Cord cysts are found in 3% of first trimester pregnancies and often resolve.

True cysts are derived from the embryological remnants of either the allantois or the omphalo-mesenteric duct when partial or complete absence of obliteration of this structures occurs. True cysts have an epithelial lining, actually a flat or cuboidal uroepithelium. Usually microscopic and with no clinical relevance, they can grow up to several centimetres but more often range between 4 and 60 mm in size. They are located between the umbilical vessels without an edematous structure of Wharton’s jelly. There is a reported association between allantoic cysts and omphalocele, patent urachus, hydronefrosis and Meckel diverticulum. Allantoic cysts are also reported to be associated with aneuploidies.
Pseudocysts are more common than the true cysts, have no epithelial lining and represent localized edema of Wharton’s jelly caused by degeneration or liquefaction. Pseudocysts are also associated with chromosomal anomalies, omphalocele and hemangiomas.

The prenatal diagnosis of umbilical cord cysts can be done during the first trimester, at 7th to 13th weeks' gestation, when cysts can be visualized most easily with color doppler studies because umbilical vessels are smaller. The natural history of these cysts is to resolve by the end of the first trimester. The longer the cyst persist, the more likely it is to be associated with a congenital anomaly. However, if no other anomaly is found, the prognosis is excellent.

The prenatal differential diagnosis includes pseudocysts, omphalo-mesenteric duct cysts, vascular disorders, abdominal wall defects, bladder extrophy and urachal anomalies. The prenatal ultrasound differential diagnosis between cysts and pseudocysts is impossible but also not important since both have been associated with fetal anomalies.

Nearly 20% of cord cysts of any type are associated with structural or chromosomal anomalies, especially with trisomy 18 and 13. Often the structural anomalies associated with the cord cyst require surgical intervention. The recommended management of persistent cyst in the second trimester of pregnancy is detailed investigation into possible fetal anomalies, including ultrasonographic examination and amniocentesis with fetal karyotype.

Intrauterine fetal death is the reported complication due to cord cysts, because of compression of umbilical vessels causing intrauterine compromise of blood flow. If a large cyst is present, delivery can be undertaken as soon as fetal lung maturity is achieved.

CASE REPORT

A 30 years old female, G5/P3, delivered a term 39-week male child. At completion of delivery, the umbilical cord broke spontaneously at its fetal insertion, only a few millimetres above the skin, and the neonate had a significant hemorrhage. This was probably provoked by the tension exerted on the fragile umbilical cord during the delivery of the fetal abdomen. It was very difficult to place a small plastic clamp in such a tiny and fragile tissue and also to control the bleeding. However, the neonate had an APGAR score of 9 at 1 and 10 at 5 minutes of life. His birth weight was 3685g. He had no apparent congenital anomalies on physical examination.
The pregnant patient was a depressive woman under anti-depressive medication for ages. Her family and obstetric history was unremarkable (no history of consanguinity or congenital malformations). She had obstetric surveillance in our hospital. Three ultrasound were done (at 12, 21 and 30 gestational weeks): in none of them an umbilical cord cyst was identified nor any congenital anomaly. She refused biochemical screening in view of personal beliefs. There was no formal indication for amniocentesis for fetal karyotype. The only intercurrence during pregnancy was hyperemesis gravidarum in the first trimester.

On clinical examination the umbilical cord had a very large cyst, 5 centimetres in largest diameter and 10 centimetres in length. The total length of the umbilical cord was normal and so was the placenta and the fetal membranes.

The stump was sent for histological examination, which confirmed the presence of allantoidal duct cyst histology and of the epithelial lining of the cyst.

The newborn had acute anemia and received red blood cell transfusion on the first 24 hours of life. An urological ultrasound was made to exclude a patent urachus. Further pathologies of the urinary tract such as posterior urethral valves or vesico-ureteral reflux were also excluded. The newborn did well during the first 15 days of his life. The dried umbilical stump detached on the 10th day, leaving a granulomatous structure. However, 5 days later the neonate was brought to the hospital because of an umbilical purulent discharge. At physical examination the purulent discharge was confirmed and no relation between the discharge and urination or Valsalva manoeuvres was found. The neonate had oral antibiotherapy (amoxicillin) for 10 days and the umbilicus had a normal cicatrisation. There were no further intercurrences.

DISCUSSION

This is a case of a giant umbilical cord related with the presence of a true umbilical cord cyst. Fortunately it was an isolated finding and the neonate had no congenital anomaly. Unfortunately the giant cyst was not identified in the fetal ultrasounds. However, this case confirms that whenever no other fetal anomaly is found, the prognosis of this umbilical cord anomaly is excellent.

References

Author Information

Carla Lilaia  
Gynecology Resident, Gynecology and Obstetrics Department, Hospital Distrital de Faro

Conceição Ribeiro dos Santos  
Specialist nurse in Obstetrics, Gynecology and Obstetrics Department, Hospital Distrital de Faro

Luís Gonçalves  
Specialist in Paediatrics, Neonatology Department, Hospital Distrital de Faro

Jesús Siles Cadilla  
Specialist in Anatomo-Pathology, Hospitalar Assistant, Pathology Department, Hospital Distrital de Faro

Celeste Santos  
Specialist in Gynecology and Obstetrics, Hospitalar Assistant, Gynecology and Obstetrics Department, Hospital Distrital de Faro

Beatriz Cabrita  
Gynecology and Obstetrics Department Director, Gynecology and Obstetrics Department, Hospital Distrital de Faro