

Ectopia Cordis, Cleft Lip and Palate in a Patient with Gastroschisis - A Clinical Case

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Abstract

Gastroschisis (GQS) and omphalocele are congenital defects of the abdominal wall; The GQS is a defect usually on the right side of the umbilical cord, where there is exposure of the intestinal loops and there is no membrane that covers them; The omphalocele is a defect through the umbilical cord, in which there is a herniation of the intra-abdominal organs, which are covered by a membrane. GQS is generally associated with minor abnormalities of the digestive tract and generally does not have an association with severe congenital abnormalities. The diagnosis can be made prenatally by Ultrasound (USG). The objective is to present a clinical case of gastroschisis associated with severe midline congenital anomalies, such as ectopia cordis, which is not frequent in these patients [1-4].

Keywords: Gastroschisis; Ectopia Cordis; Cleft Lip; Palate

Introduction

The presence of ectopia cordis and defects of cleft lip and palate are not frequent malformations in patients with gastroschisis, due to its high morbidity and mortality, we consider it important to present this clinical case, in which there were major congenital malformations of the midline, which they prevented offering a surgical treatment to their congenital malformation, leading to the death of the patient in the first 12 hours of life. Ultrasound is a tool for prenatal assessment that can provide us with important information about intestinal loop conditions and if there are associated congenital malformations, which help us to predict the prognosis of the patient at birth [1-3].

Clinical Case

This is a male patient born to a 25-year-old mother, who reports passive smoking during pregnancy, as well as a history of psychoactive drug use and smoking prior to pregnancy, but denied the use of drugs during pregnancy, the mother had adequate prenatal control, took folic acid and iron during pregnancy, was diagnosed with gastroschisis at 28 weeks of gestational age by ultrasound (USG), the USG prenatal only diagnosed gastroschisis, no other anomaly was reported congenital. The mother received two doses of betamethasone as an inducer of lung maturity at 28 weeks of gestation, the patient during prenatal control presented premature rupture of membranes at 33 weeks of gestational age, which is why an emergency caesarean section was performed.

At birth a premature newborn with low weight is obtained, the amniotic fluid is clear without meconium, the newborn was given the basic maneuvers of neonatal resuscitation, during the physical examination multiple congenital malformations are diagnosed, such as cleft lip and palate, neck short, with defect of the sternum presenting ectopia cordis, with defect of the abdominal wall presenting gastroschisis with a defect of 2 cm to the right of the umbilical cord, with exposure of the intestinal loops, without membrane, without data of chemical peritonitis or intestinal atresia, with bilateral cryptorchidism, extremities without abnormalities (Figure 1) [4-11].

The patient was placed a silo to cover the handles and the heart defect, and was transferred to the NICU for conservative treatment.



Figure 1: Premature newborn 33SDG, with low weight, with abdominal Wall defect presenting gastroschisis with exposed liver and defect in stemum presenting ectopiacordis with cleft lip and palate

Result

The patient died at 72 hours of age, due to heart failure and sepsis, secondary to their congenital alterations, it was not possible to perform an echocardiogram to rule out associated cardiac alterations, no genetic tests or necropsy were performed. Conclusion: The association of gastroschisis to major malformations is presented with a low incidence, the prenatal ultrasound can help us to rule out associated congenital malformations.

Discussion

Gastroschisis (GQS) is a congenital defect of the abdominal wall, located on the right side of the umbilical cord, where there is exposure of the intestinal loops and there is no membrane that covers them; The amniotic fluid stained with meconium creates an inflammation of the wall of the intestine, called perivisceritis, the dilatation of the intestinal loops or the gastric chamber diagnosed by prenatal USG, are factors of poor functional prognosis for the patient. The GQS alone in 10% of cases is associated with congenital anomalies, most of the anomalies are of the digestive tract as: Stenosis or intestinal atresia, generally not usually associated with serious congenital defects or chromosomal abnormalities as in the omphalocele.

The differential diagnosis of a gastroschisis and omphalocele is in the presence or absence of membrane that covers the intestinal loops, the omphalocele usually has a reserved prognosis with high mortality since it is associated with congenital abnormalities in up to 70% of the cases, there are defects cardiac and chromosomal abnormalities such as trisomy 13 or Patau syndrome, mosaic trisomy 14, trisomy 15 or Prader Willi, trisomy 18 or Edwards syndrome and trisomy 21 or Down syndrome, Beckwith-Wiedemann syndrome, there is also greater association to Wilms tumor, hepatoblastoma, neuroblastoma, usually suffer from caudal or midline defects, cranial defects, defects of the caudal tube with bladder exstrophy or cloacal exstrophy, Cantrell's pentalogy is characterized by sternum defects, diaphragmatic hernia, alterations in pericardium, heart disease and ectopia cordis, with poor prognosis for life.

The presence of ectopia cordis and midline defects are not frequent in patients with gastroschisis, due to its high morbidity and mortality, we consider it important to present this clinical case, in which there were major midline congenital malformations that prevented us from offering surgical treatment to his congenital malformation, leading to death to the patient in the first 12 hours of life [6-13].

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