Journal Homepage: - www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/14783
DOI URL: http://dx.doi.org/10.21474/IJAR01/14783

RESEARCH ARTICLE

PENTALOGY OF CANTRELL, AN EXTREMELY RARE MALFORMATION:A CASE REPPORT

A. Oubid, S. Haddout, A. Cherkaoui, M. Jalal, A. Lemrissi and S. Bouhya

Manuscript Info

Manuscript History
Received: 27 March 2022
Final Accepted: 30 April 2022
Published: May 2022

Key words:-
Cantrellpentalogy, Cantrell Syndrome, Congenital Malformation, Highcelosomia

Abstract

Pentalogy of Cantrell (PC) is an extremely rare and usually fatal birth defect. It corresponds to a defect in the closure of the umbilical ring concerning the supra-umbilical portion of the abdominal wall secondary to an anomaly in the development of the mesoderm during the first stage of embryonicdevelopment (1). We report a case of observation of pentalogy of Cantrelldiagnosed in the delivery room in a male newborn on an unattendedpregnancyestimatedat 7 months. The caseisaccompanied by video footage of the newborntakenafterdelivery.

Introduction:-

Cantrell syndrome or Pentalogy of Cantrell (CP) is a congenital anomaly described for the first time in 1958 by Cantrell, and includes five anomalies: supra-umbilical hernia of the abdominal wall, defect of the anterior part of the diaphragm and the pericardium diaphragmatic, anomaly of the lower part of the sternum, heart defects.

The occurrence of this syndrome is extremely rare and usually fatal, its incidence varies from 5.5 to 7.9 per million live births, There is a male predominance with a male/female ratio of 2.7:1 (2)

In this work we report a case of pentalogy of Cantrell diagnosed on a pregnancy estimated at 7 months with a brief review of the literature on the epidemiological, physiopathological, clinical, radiological characteristics and the therapeutic management of this syndrome.

Observation:-

BM, aged 38, with no particular medical or surgical history, no notion of consanguinity or family fetal malformation, primiparousprimigravida, presents to the delivery room for the threat of premature delivery in a pregnancy not followed up within the framework of prenatal consultations, estimated at 7 months, without any notion of specific drug intake. The clinical examination revealed a lower fundal height compared to gestational age with positive and regular BCF. Vaginal examination found an erased cervix dilated to 8cm. Obstetric ultrasound shows an evolving mono-fetal pregnancy, amitotic fluid in very small quantities, with difficulty in individualizing the fetal thoraco-abdominal organs. The macroscopic examination at birth objective (figure 1) a male newborn weighing 1200g, presenting a very marked superior coelosomy (exteriorized heart, liver and intestine). The skin defect goes beyond the umbilicus. Macroscopically The apex of the heart is oriented upwards. There is also a skeletal anomaly in the left upper limb (Figure 2).

Unfortunately, the newborn died 30 minutes after birth before any surgery was performed and pathology examination for other visceral malformations could not be performed.

Corresponding Author:- A. Oubid

Discussion:-

Developmental abnormalities of the abdominal wall may involve delineation abnormalities (embryopathy) or growth abnormalities. Delimitation anomalies: CELOSOMY, following a lack of “induction” or “competence”; the delimitation can be disturbed at an early stage in the thoracic or pubic regions, around the 6th, 7th week (8SA-9SA), in the umbilical region. We can then observe high celosomies (ectopiacordis or cantrell syndrome), or low celosomies (bladder exstrophy) or completecelosomies.

Cantrell's syndrome results from midline closure defects, including lower sternum wall defect, supraumbilical abdominal wall defect, anterior diaphragmatic defect, diaphragmatic defect pericardium with free communication between the pericardium and the peritoneal cavities and congenital heart defects (1)

it occurs with varying degrees of severity, which can lead to serious and life-threatening complications. Most infants do not develop all of the potential defects, which may be referred to as Incomplete Pentalogy of Cantrell. When the five faults are present, we speak of a completepentalogy of Cantrell

Therefore and in the full spectrum this malformation includes five abnormalities: (2) supraumbilical hernia of the abdominal wall, defect of the anterior part of the diaphragm and of the diaphragmatic pericardium, abnormality of the lower part of the sternum, heart defects, cases extremes, the heart is exteriorized, giving a picturecalledcardiacectopia

The exact cause of Cantrell's Pentalogy is unknown. Most cases are thought to be sporadic.

The diagnosis of pentalogy of Cantrell can often be made prenatally with prenatal ultrasound, as early as 10 weeks of pregnancy. Echocardiography is usually done to assess the extent of damage to the heart. Prenatal fetal MRI allows optimal assessment of fetuses with CP and assess the degree of certain abnormalities such as abnormalities of the abdominal wall and associated pericardium (2)

In developing countries, whether the diagnosis is antenatal or postnatal, the prognosis is almost always fatal. A study carried out on a series of 10 patients who had benefited from antenatal diagnosis showed a uniformly fatal outcome without any treatment. The absence of a cardiovascular surgery service largely explains this prognosis (3)

The long-term prognosis for children with this anomaly depends mainly on the complexity of the associated heart defect. In most cases, Pentalogy of Cantrell is fatal withoutsurgery

The treatment strategy and prognosis depend on the size of the abdominal wall defect. Following the prenatal diagnosis, termination of pregnancy may be proposed in serious cases, after amniocentesis showing a karyotype abnormality. A multidisciplinary team should monitor the milder forms to determine the best time for delivery (2).

Surgical interventions that may be necessary soon after birth, After delivery, the repair of omphaloceleshould not bedelayed.

In severe cases, some physicians advocate a staged repair of defects associated with the Pentalogy of Cantrell. The initial operation immediately after birth separates the peritoneal and pericardial cavities, covers the midline defect, and repairs the omphalocele. After proper growth of the chest cavity and lungs, the second stage is to repair heart defects and return the heart to the chest. Eventually, usually around age 2 or 3, reconstruction of the lower sternum or epigastriummaybenecessary.

Other Pentalogy of Cantrell treatments are symptomatic and supportive.

This syndrome has also been reported occasionally in association with sirenomelia, anencephaly, and amniotic band syndrome (2)

Figure 1:- newborn with superior coelosomy with exteriorized heart, liver and intestine.

Figure 2:- Cantrell syndrome with skeletal abnormality in the left upper limb.

Conclusion:-

The occurrence of Cantrell's syndrome is extremely rare and generally fatal, hence the importance of its prenatal diagnosis, thus allowing, in some cases, termination of pregnancy, especially in fatal forms.

Référence:-

1. Sidibé N , Dembélé B , N'Diaye M , Diarra B , Ba H , Sangaré I , Sogodogo A , Sissoko A , Dakouo R , Camara H , Koné O , Tounkara H , Menta I , Diallo M. The pentalogy of Cantrell: A case report. HealthSci. Dis: Vol 22 (8) August 2021 pp 125-127
2. Suresh Chandran, Dinesh Ari. Pentalogy of Cantrell: An Extremely Rare CongenitalAnomaly. Journal of ClinicalNeonatology | Vol. 2 | Issue 2 | April-June 2013
3. Toni KasoleLubala, Augustin MulanguMutombo , Tina Katamea , Nina Lubala , Arthur NdundulaMunkana , MaguySangajiKabuya , Joséphine KalengaMonga , Oscar NumbiLuboya. Ectopiacordis thoracique sporadique: description clinique d'un cas. Pan AfricanMedical Journal. 2012; 13:62.