Ectopiacordis - A Rare Congenital Anomaly

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Summary:

Ectopiacordisis a rare congenital malformation in which the heart is located partially or totally outside the thoracic cavity. It may occur as an isolated malformation or it may be associated with a larger category of ventral body wall defects that affect the thorax, abdomen or both. We present a case of ectopiacordis associated with complete ventral body wall defect.

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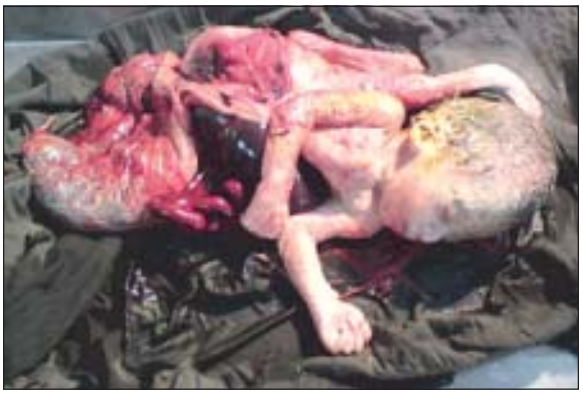
Case Report:

A 28-year- G3P0 +2 (abortion) patient was admitted at Faridpur Medical College Hospital at her 38weeks of pregnancy with labour pain. The patient had regular antenatal checkup. The patient did not give any history of medical or surgical illness and no exposure to drugs and toxins. There was no history of congenital anomaly in her family or consanguinealmarriage. She had three ultrasonography during pregnancy at her 20weeks, 28weeks and at 34weeks. Her ultrasonogram showing major congenital anomaly of the fetus. The sonologists figured it out to be a probable case of huge sacrococcygealteratoma. However each time the patient had antenatal checkup and ultrasonography, she was counseled and advised to get admitted into hospital for termination of pregnancy but she ignored and continued her pregnancy. She came at labour and on examination symphysiofundal height was about 36 weeks pregnant uterus size, presentation could not be definitely identified due to uterine contraction, fetal heart sound was not audible. On per vaginal examination - cervical os was full dilated, membrane ruptured, few intestinal loops were presenting through the os. A dead female baby was delivered spontaneously per vagina by breech. Placenta, cord and membrane were expelled out by controlled cord traction. On examination of the baby, there was complete absence of anterior abdominal wall with evisceration of all the abdominal organs (small and large intestines, liver, spleen, kidneys) along with the heart. There was also abnormal position of the lower limbs with hypoplasia. Other parts of the body (head, face, upper limbs) appeared to be normal. The umbilical cord and placenta were normal in appearance and location. A diagnosis of ectopiacordisassociated with ventral body wall defect was made.



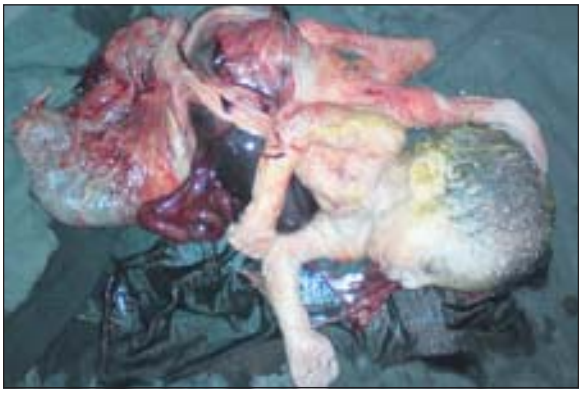


Fig: *Ectopiacordis with anterior abdominal defect with evisceration of all viscera.*

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Discussion:

Ectopiacordis is a very rare anomaly with an estimated prevalence of 0.079/10,000 births and may occur more frequently in females[1].It is a congenital malformation which was observed thousands years ago [2].Clinically, ectopiacordis has been classified into four types: cervical, thoracic, abdominal and thoracoabdominal whether the heart is respectively in the neck, anterior to the sternum, within the abdomen or between the thorax and the abdomen. Most common are thoracic and thoracoabdominal[3,4,]Embryologically, in about the third week, early disturbance in the formation of the cephalic fold will result in defective formation of the thoracic and epigastricwalls, finally resulting in ectopiacordis with anterior defect of the sternum and diaphragm and an omphalocele[5,6]

To date, the cause of ectopiacordis is still unknown.There have been a number of reports linking it to chromosomal abnormalities. Reported karyotypic abnormalities include trisomy [7,] Turner syndromeand 46,XX,17q+20.[8] various type of congenital anomaly such as central nervous system, cardiac, skeletal, gastrointestinal and other malformation is associated with ectopia cordis.[6] with the advent of ultrasonography it can be diagnose in early pregnancy.

The prognosis of ectopiacordis is generally poor, death commonly occur before or immediately after birth, usually due to associated malformations.

Conclusion:

Ectopiacordis is a rare congenital malformation with a poor prognosis. Ultrasonography is of great value in the prenatal diagnosis. Obstetrical management should include a careful search for associated anomalies, especially cardiac, and assessment of fetal karyotype. Pregnancy termination prior to viability should be considered.

References:

1. Khoury MJ, Cordero JF, Rasmussen S: Ectopiacordis, midline defects and chromosome abnormalities: An epidemiologic perspective. Am J Med Genet 30:811-817, 1988.
2. Taussing HB. World survey of the common cardiac malformation: developmental error of genetic variant . Am J Cardiol1982 ; 50 : 544-59.
3. Byron F. Ectopiacordis: report of a case with attempted operative correction.JThoracSurg1949 ; 17:717-722.
4. Leca F, Thibert N, Khoury W, Fermont L, Laborde F, Dumez Y. Extrathoracic heart (ectopiacordis) : report of two cases and review of literature . Int J Cardiol1989 ; 22 : 221-8.
5. Achron R, Schimmel M, Farber B, Glaser J. Prenatal sonographic diagnosis and perinatal management of ectopiacordis. Ultrasound ObstetGynecol1991 ; 1 :431-434.
6. Firmin RK., Fragomeni LS ,Lennox SC. Complete cleft sternum. Thorax 1980 ; 35 : 303-306.
7. Bick D, Markowitz RI, Horwich A: Trisomy 18 associated with ectopiacordis and occipital meningocele. Am J Med Genet 30:805-810, 1988.
8. Fox JE, Gloster ES, Mirchandani R: Trisomy 18 with Cantrell Pentalogy in a stillborn infant. Am J Med 31:391-394, 1988.