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Cantrell Pentalogy-Arare Congenital Disorder

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Abstract: ***Cantrell pentalogy is a rare congenital thoraco-abdominal disruption with five characteristic: Ectopia cordis and intracardiac anomalies, lower sternal defect, Midline supraumbilical thoraco-abdominal wall defect, anterior diaphragmatic defect, defect of diaphragmatic part of pericardium that results in relation between pericardial cavity and peritoneum. A diagnosis of pentalogy of Cantrell can often be made prenatally using a fetal ultrasound. Detail evaluation requires CT scan with contrast study. Long term prognosis is poor even in super speciality hospital and even after multiple corrective surgeries, morbity and mortalities are high.***

Keywords: Cantrellpentalogy, fetal ultrasound, MDCT

1. Introduction

Cantrellpentalogy is a rare congenital thoraco-abdominal disruption, first described by Cantrell etal with five characteristic:

1) Ectopiacordis and intra cardiac anomalies;
2) Lower sternal defect;
3) Midline supraumbilical thoraco-abdominal wall defect;
4) Anterior diaphragmatic defect;
5) Defect of diaphragmatic part of pericardium that results in relation between pericardial cavity and peritoneum.

Most infants donot develop all of the potential defects, which may be referred to as in complete pentalogy of Cantrell. When all five defects are present, this is referred to as complete pentalogy of Cantrell. The variability of the disorder from one individual to another can be significant. The exact cause of pentalogy of Cantrell is unknown. Most cases are believed to occur sporadically.

Prognosis of pentalogy of Cantrell depends on severity of intra and extra cardiac defects, pulmonary hypoplasia, extent of abdominal wall defect, cerebral anomalies and diaphragmatic herniation.

2. Case Report

3 year female child came to our department as a referral from cardiology department at U. N. Mehtainstituteof cardiology and research centre, Ahmedabad, Gujarat for congenital heart disease evaluation. Initially patient presented with complains of failure to gain weight, fatigability.

2D echo cardiography had showed double outlet of right ventricle. In our department patient underwent MDCT pulmonary angiography after 15 ml intravenous injection of non ionic contrast through anti cubital vein. These images were reviewed with MIP, SSD and Volume Rendering.

On CT pulmonary angiography, following findings were obtained in presented case. Double outlet right ventricle with D-posedaorta and dilated right ventricle, Severe RVOT/ in fundibular pulmonary stenosis, severe stenosis in proximal and mid part of left LPA, large ASD amounting to commonatrium, peri membranous VSD, bilateral SVC. CTalsoshowedLeftventricularapicaldiverticulum, herniating through diaphragmatic defect in abdominal cavity. Defects in anterior abdominal wall, pericardial layer, diaphragm and lower sternum noted along with congenital heart disease. Elevated left dome of diaphragm.

3. Discussion

A diagnosis of pentalogy of Cantrell can often be made Prenatally using a fetal ultrasound. Detail evaluation requires CT scan with contrast study. Complete clinical examination from head to toe is needed to check associated anomalies. The most severe expression of pentalogy of Cantrell presents at birth with ectopiacordis and omphalocele. Ectopiacordisisa severe condition in which the heart is completely or partially displaced outside of the thoracic avity and therefore not protected by the chest wall. Associated congenital heart defects including ASD, VSD, Dextrocardia may occur.

Abnormalities affecting the sternum can range from complete absence of the cartilage prominence at the end of the sternum (xiphoid) to complete absence of the sternum. In some cases, the sternum may be cleft or abnormally short.
Defects of pericardiumoccur in pentalogy of Cantrell, specifically in the lower portion where it meets the diaphragm. Affected infants may also have a hole in the diaphragm allowing the contents of the abdomen to protrude into the chest with resultant congenital diaphragmatic hernia.
Additional anomalies associated with pentalogy of Cantrell include cleft lip, cleft palate, dysplasia of the kidneys, cystichygroma, limb defects (club feet, absent bones in the arms or legs) and neural tube defects (encephalocoele) and trisomy 13 and 18. Studies showed that even with care monitoring in professional centers and multiple corrective surgeries, patient have high morbidity and mortality rate and long time prognosis is poor.

4. Conclusion

The presented case has all portions of pentalogy of Cantrell except midline supraumbilical wall defect and ectopiacordis, The various defects potentially associated with pentalogy of Cantrell can cause a serious issues including under development of the lungs, breathing difficulties, embolism, heart failure. Infants with pentalogy of Cantrell are at risk of

developing widespread internal infection of the abdominal cavity.

The treatment of pentalogy of Cantrell is directed toward the specific symptoms that are apparent in each individual. Surgical intervention for cardiac, diaphragmatic and other associated defects is necessary. Affected infants will require complex medical care and may require surgical intervention. In most cases, pentalogy of Cantrell is fatal without surgical intervention. However, in some cases, the defects are so severe that the individual dies regardless of the medical or surgical interventions received.

References

[1] Peixoto-filho FM, Docima LC, Nakamura-pereiraM. Prenatal diagnosis of Pentalogy of Cantrellin the first trimester: is3-dimensional sonography needed? J Clin Ultra sound. 2009;37 (2):112-4.
[2] McmahonCJ,Taylor MD, CassadyCI et-al. Diagnosis of pentalogyof cantrellinthefetususing magneticresonance imaging and ultrasound. Pediatr Cardiol. 28 (3):172-5.
[3] VanHoornJH,MoonenRM, HuysentruytCJ et-al. Pentalogyof Cantrell:twopatientsand a review to determine prognosticfactorsforoptimalapproach. Eur. J. Pediatr.2007;167 (1):29-35. 0578-9 -Freetextatpubmed-Pubmedcitation
[4] DiBernardoS, SekarskiN, Meijboom E. Left ventricular diver ticulum in aneonate with Cantrell syndrome. Heart. 2004;90 (11):1320.

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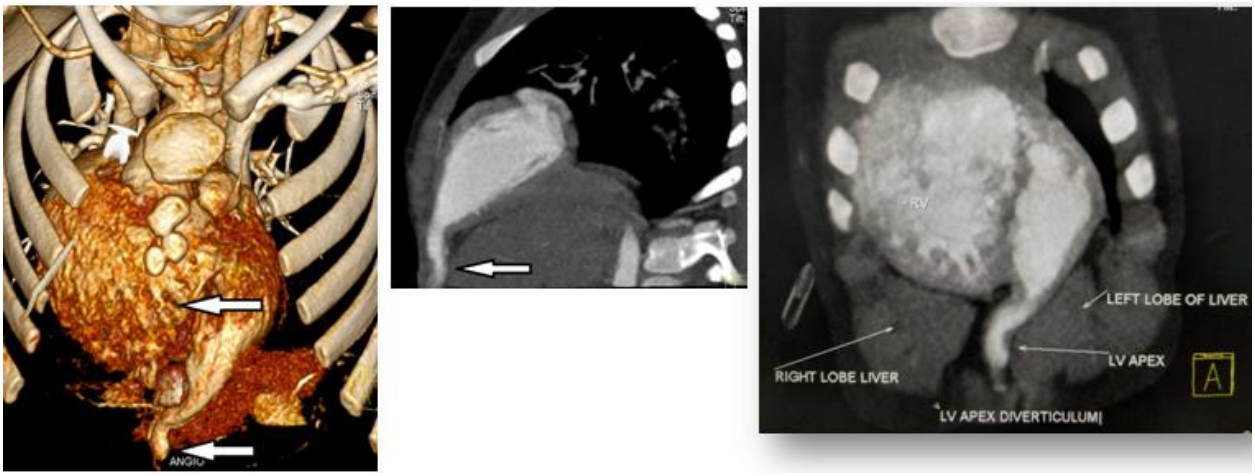
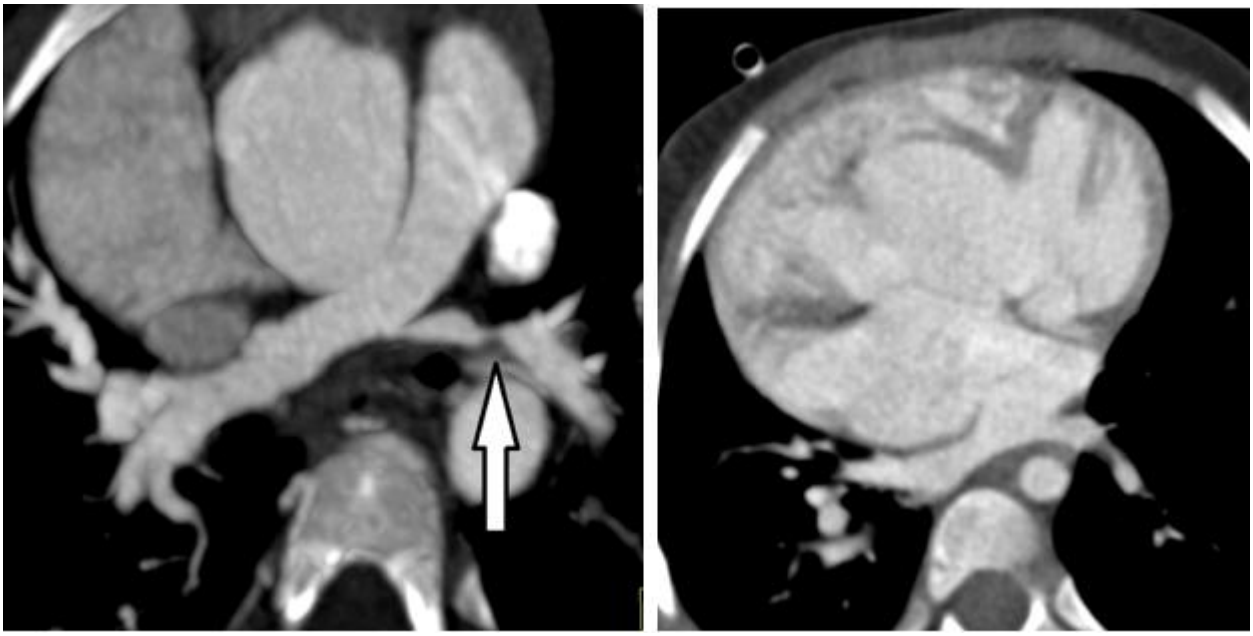


Arrow in image demonstrate the sternal defect and left ventricular apical diverticulum herniating into abdomen between right and left lobe of liver through pericardial and diaphragmatic defect.



Severe stenosis in proximal and mid part of left LPA is seen in image. Large ASD amounting to common atrium and perimembranous VSD is also noted in presented case as a part of intracardiac anomaly.