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Pentalogy of Cantrell in a Neonate : A Case Report[1]

Min Sun Kim, M.D., Kyu Soon Kim, M.D.,
Seoung-Oh Yang, M.D., Ph.D., Keum Won Kim, M.D.[2]

The pentalogy of Cantrell is a rare thoraco-abdominal disruption with a variable combination of defects involving the abdominal wall, diaphragm, pericardium, sternum, and heart. Herein, we report the multi-detector computed tomography (MDCT) findings of Cantrell's pentalogy in a neonate. A MDCT proved to be useful and complementary for the diagnosis of Cantrell's pentalogy and evaluation of its associated anomalies.

Index words : Pentalogy of cantrell
Ectopia cordis
Omphalocele
Multi-detector computed tomography (MDCT)

The pentalogy of Cantrell is a rare congenital malformation characterized by midline defects resulting from the defective development in the septum transversum. The constellation of findings includes a deficiency of the diaphragmatic pericardium, lower sternum, anterior diaphragm, supraumbilical abdominal wall (often allowing an omphalocele as well as an intrapericardial diaphragmatic hernia), and a cardiac lesion (most often a ventricular septal defect) (1). In severe cases, the heart herniates through the diaphragmatic defect, causing thoracoabdominal ectopia cordis (2). Other associated congenital cardiac lesions may include an atrial septal defect, pulmonary valve stenosis, tetralogy of Fallot, dextrocardia, anomalous pulmonary venous connection, tricuspid atresia, and truncus arteriosus. An associated left ventricular apical aneurysm has been reported in several cases (3, 4).

[1]Department of Radiology, Eulji University Hospital,
[2]Department of Radiology, Konyang University Hospital
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Address reprint requests to : Kyu Soon Kim, M.D., Department of Radiology, Eulji University Hospital, Dun-san 2 Dong, Seo-gu, Daejeon 302-799, Korea.
Tel. 82-42-611-3581 Fax. 82-42-611-3590 E-mail: kskim@eulji.ac.kr

Herein, we describe the radiologic findings of the pentalogy of Cantrell in a neonate.

Case Report

A 3820 g male neonate was born at 39 weeks gestation by spontaneous normal vaginal delivery to a 34-year-old woman with no history of systemic illness. The parturient had no obstetric examinations, including a sonogram, after the first trimester. At birth, the infant's Apgar scores were 6 and 9 at 5 and 10 min, respectively. Upon physical examination, the infant had a 12 × 8 cm protruding midline mass at the lower thorax and upper abdomen (Fig. 1). The mass was slightly lobulated, with an upper pulsatile mass and a lower softer mass connected to the umbilicus. The umbilicus was located at the lower midline portion of the mass. The overlying skin was nearly intact. No other associated anomalies were detected. An echocardiography showed a functional single ventricle (right ventricular type) with a rudimentary left ventricle.

A simple radiography showed two types of abnormal densities: a well-defined solid mass in the upper ab-

domen and a well-defined, gas-containing round lesion in the middle and lower abdomen (Fig. 2). A contrast-enhanced computed tomography (CT) showed a defect involving the lower chest wall, anterior abdominal wall, and shortened sternum with herniation of the heart and intestines (Fig. 3). The midline defect extended from the lower chest to the umbilicus. The sternum was shortened and there were defects of the anterior diaphragm, through which the heart was protruding for 4–5 cm, and the apex was directed anteriorly. A surface shade display (SSD) and volume-rendering technique (VRT) of multi-detector computed tomography (MDCT) clearly showed an inferior sternal cleft with the ectopia cordis (Fig. 4). A coronal reformatted image shows that a single ventricle connects with the aorta and pulmonary artery (Fig. 5). Based of the MDCT findings, the pentalogy of Cantrell was identified.

It was recommended that the neonate undergo surgical correction, but the parents declined.

Discussion

Cantrell's pentalogy, first described in 1958, consists of lower sternal anterior diaphragmatic and parietal pericardial defects, an omphalocele, and herniation of the heart. Ectopia cordis describes the herniation or extrusion of the heart, which may occur through either the thorax or the thoracoabdominal region(1).

This case is consistent with the thoracoabdominal type. The following characteristics were observed in our patient: a lower sternal defect, an anterior extrathoracic heart, absence of the anterior diaphragm, and an omphalocelce with a supraumbilical abdominal wall defect. As such, our patient had all the classic symptoms of Cantrell pentalogy. The hypothesis underlying this condition is abnormal development of the mesoderm at a very early stage in embryonic life, occurring prior to or immediately after differentiation of the primitive intraembryonic mesoderm into its splanchnic and somatic layers, since derivatives of both these layers are involved. To date, the etiology of this anomaly is unclear (5). In our case, no family history for fetal malformations was documented, and the parturient was denied exposure to drugs or medications during the pregnancy period. The overall objectives of ectopia cordis management are closure of the chest wall defect, repair of the assocciated omphalocelce, placement of the heart into the thorax, and repair of the intracardiac defect (6, 7). In our case, the parents declined surgery. The prenatal diagnosis of pentalogy of Cantrell is usually performed by ultrasound (8, 9); however, the CT findings of the pentalogy of Cantrell in a neonate have rarely been reported (10). We have described a case of pentalogy of Cantrell diagnosed in a neonate. The MDCT findings clearly demonstrated the thoracic and abdominal defects. MDCT facilitates the spatial assessment of the malformation; specifically, the SSD images showed ectopia cordis and an omphalocele, and the VRT images showed a shortened sternum with a cleft and ectopia cordis.

In conclusion, a MDCT is a useful and complementary imaging modality in the evaluation of complex

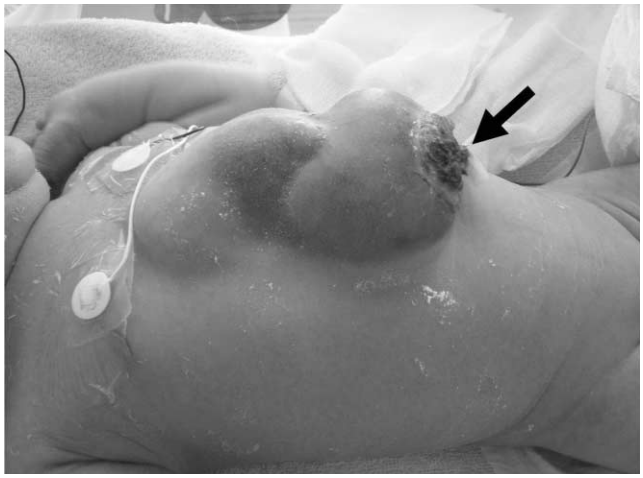

Fig. 1. A 1-day-old male infant with a large mass protruding from the lower chest and upper abdomen. The umbilical clamp is seen at the lower midline portion of mass (arrow).

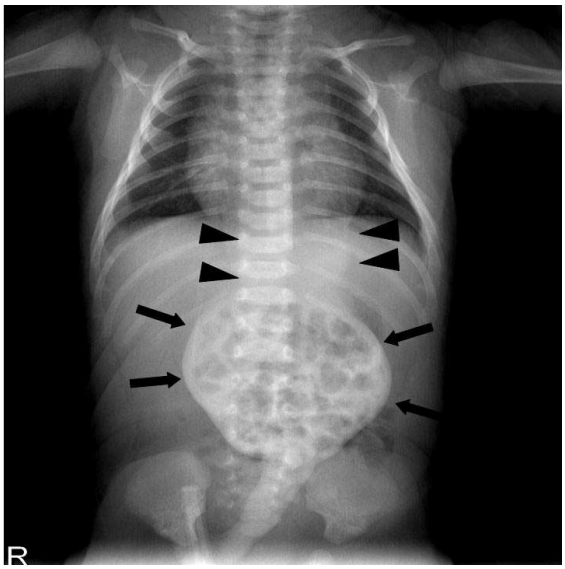

Fig. 2. A simple radiograph shows two types of abnormal densities: a well-defined solid mass in the upper abdomen (arrowheads), and a well-defined gas-containing round lesion in the middle and lower abdomen (arrows).

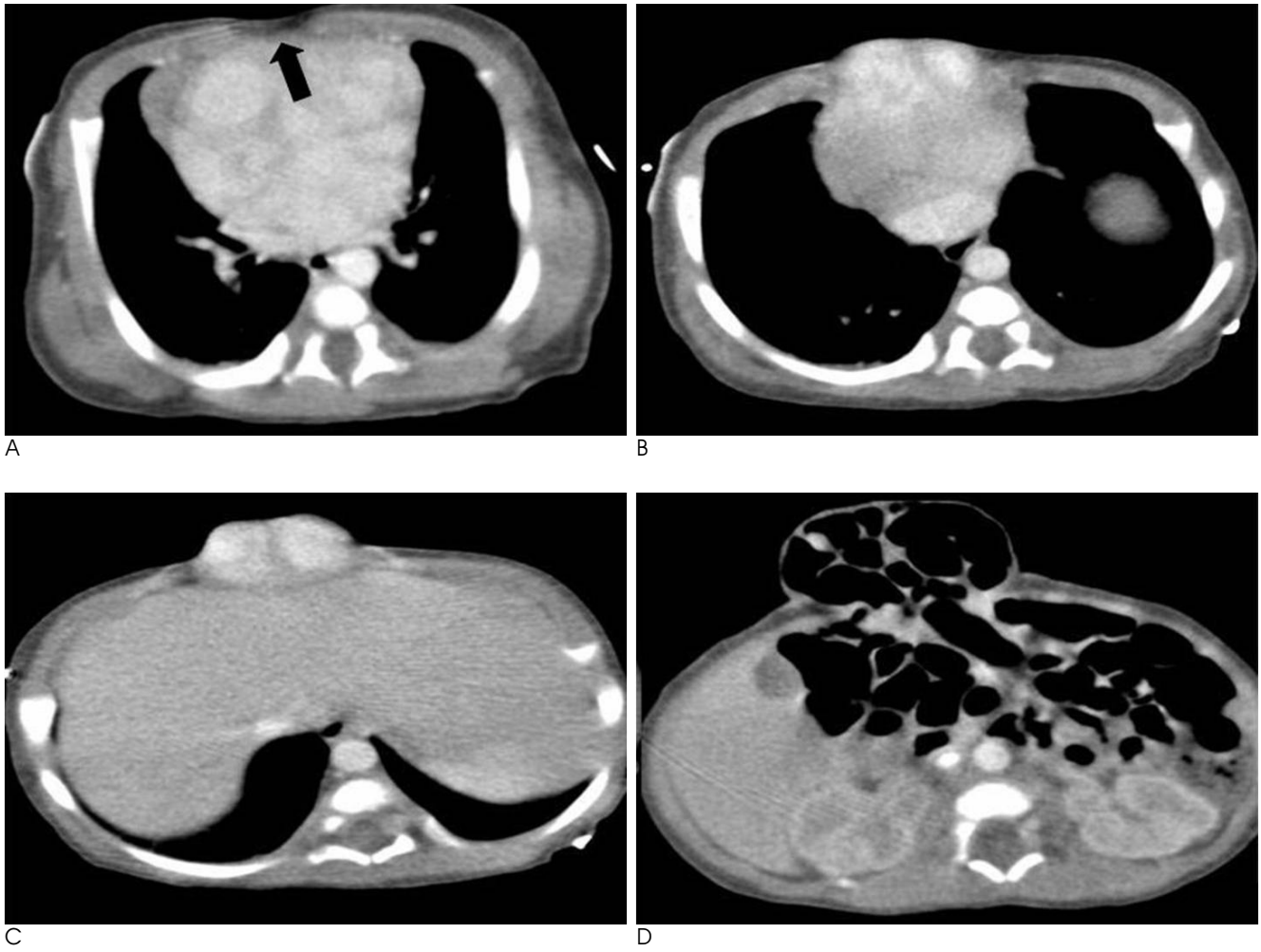

Fig. 3. The contrast-enhanced axial CT shows a defect of the inferior sternum (arrow) (A), anterior diaphragm, and anterior abdominal wall with ectopia cordis (B, C) and an omphalocele (D).

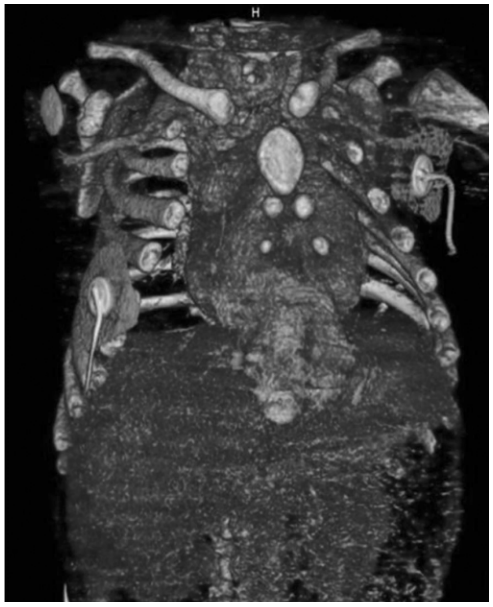

Fig. 4. Volume rendering image shows the defect of the sternum.

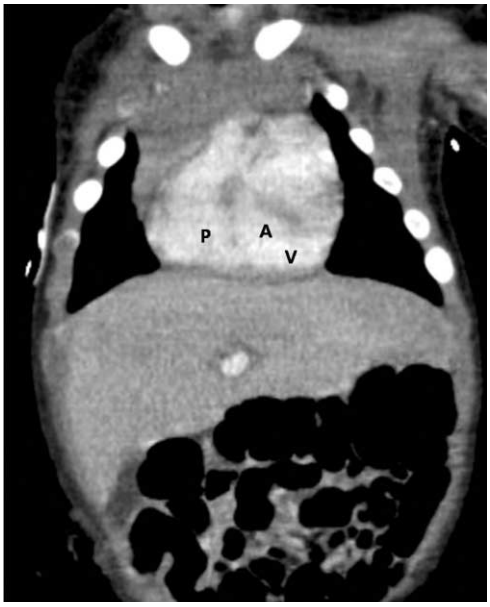

Fig. 5. The contrast-enhanced coronal reformatted image shows the single ventricle (V) connecting with the aorta (A) and pulmonary artery (P).

anomalies associated with sternal defects.

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신생아의 Cantrell씨 증후군: 증례 보고[1]

[1]을지대학병원 영상의학과, [2]건양대학병원 영상의학과

김민선 · 김규순 · 양승오 · 김금원[2]

Cantrell씨 다섯 징후는 흉복벽의 융합이 이루어지지 않은 드문 선천성 기형으로 다양한 정도의 복벽, 횡경막, 심낭, 흉골 및 심장기형과 동반된다. 저자들은 Cantrell씨 다섯 징후를 보인 신생아의 다중검출기 CT 소견을 보고하고자 한다. 다중검출기 CT는 Cantrell씨 증후군을 진단하고 동반된 기형의 평가에 매우 유용하다.