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Case report

NEONATAL MASSIVE PERICARDIAL EFFUSION FROM INTRAPERICARDIAL LIVER HERNIATION : A CASE REPORT

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Abstract A full term neonate presented with massive pericardial effusion without apparent clinical manifestation of cardiac tamponade. The effusion was caused by herniation of the liver into the pericardial sac, which is the rarest type of congenital diaphragmatic hernia. The postulated mechanisms of pericardial fluid accumulation are the mechanical irritation of pericardium by herniated liver or partial venous outflow obstruction of the protruded part. The clinical features, diagnostic studies, differential diagnosis, treatment and prognosis of this condition are described. **Chiang Mai Med Bull 2003;42(1):37-44.**

Keywords : pericardial effusion, intraperiocardial liver herniation, diaphragmatic hernia

Congenital diaphragmatic hernia occurs in about 1 in 2,000 to 1 in 10,000 livebirths.[1] The most common type is a defect in the posterolateral part of the diaphragm or foramen of Bochdalek. Another well known type is anterior retrosternal herniation through the foramen of Morgagni, which is far less common and not usually encountered in the infancy period. An extremely rare type is a defect in the central tendon of the diaphragm or peritoneo-pericardial hernia.[2] We report an infant who had respiratory distress soon after birth from massive pericardial effusion, caused by this latter and rarest type of hernia.

Case report

A 2-day-old female infant was referred to Chiang Mai University Hospital with a problem of bilateral pleural effusion. She was delivered vaginally at 40 weeks of gestation to a 21-year-old mother who had an uncomplicated pregnancy. Her

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birth weight was 2,980 grams and the Apgar scores were 4, 6 and 9 at 1, 5 and 10 minutes, respectively. At 2 hours of age she was noted to have respiratory distress that progressed to respiratory failure in the following 6 hours. A chest roentgenogram showed an opacity that occupied most of both thoracic cavities, and bilateral pleural effusion was suspected (Fig. 1). Left thoracocentesis was performed and revealed a scanty amount of clear yellow fluid. Antibiotics and ventilator support were given and she was transferred to Chiang Mai University Hospital the next day.

On admission, she was dyspneic and tachypneic, but not cyanose. Her heart rate was 156 beats per minute and blood pressure 87/56 Torr. She had normal quality of first and second heart sound, but her sound of breathing was diminished bilaterally. Other physical examinations were unremarkable. The pulse oximetry did not show a distinct plethysmographic wave form fluctuation between inspiration and expiration. The

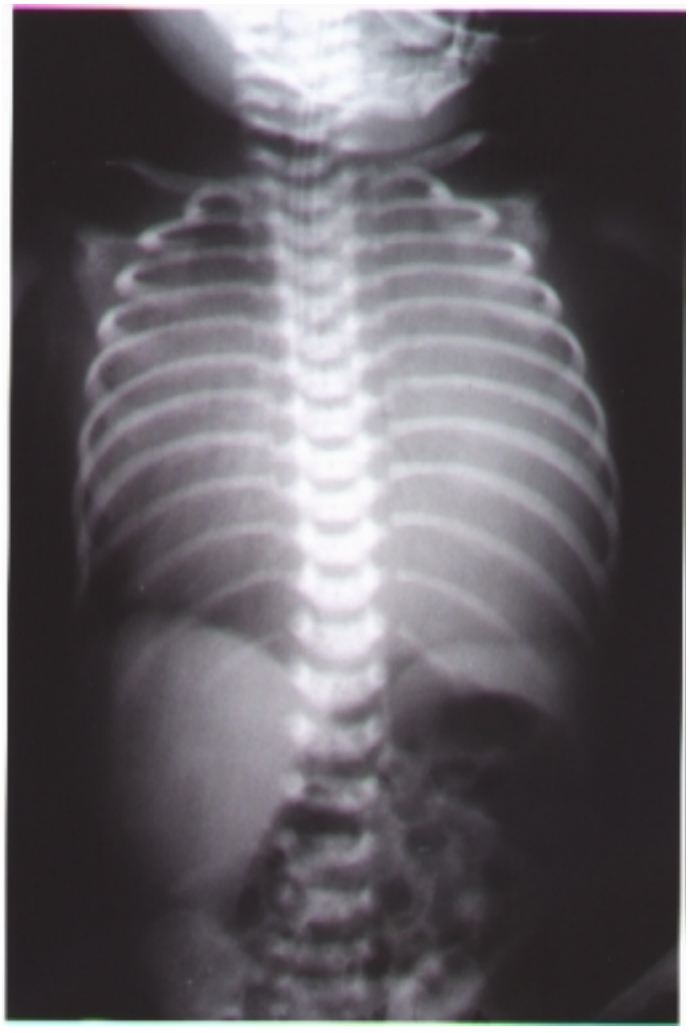

Figure 1. Chest roentgenogram on the first day of life showing an opacity that occupied most of the thoracic cavity. Note the diaphragm is clearly seen bilaterally, but the shadow of the left lobe of the liver could not be seen (liver cut off sign).

chest roentgenogram suggested a markedly enlarged cardiac silhouette with left pleural effusion (Fig. 2). An electrocardiagram showed a sinus rhythm and QRS axis of about 190 degrees, with electrical alternans suggesting pericardial effusion. Low QRS voltage was found only in lead II, III and aVF (Fig. 3). An echocardiogram was performed and it demonstrated massive pericardial effusion of about 1.5 cm in thickness. There was a mass within the pericardial sac, which was identified as being in continuity with the liver and it had liver echogenicity (Fig. 4). No venous dilatation was seen within the herniated liver. The cardiac structure appeared to be normal. There were no diastolic collapse of the right atrium and ventricle. Neither significant variation in flow between the left and right of the heart nor interventricular septum shift was found during respiration

The infant was stable when ventilated with 40% oxygen, a peak inspiratory pressure of 20 cm H_2O, positive end expiratory pressure of 5 cm H_2O and ventilator rate of 30 beats per minute. The elective surgery was carried out on the fifth day of life and part of the liver was found protruding into the pericardial cavity through a defect in the diaphragm. Straw colored fluid of about 200 mL was also found in the pericardial sac. The

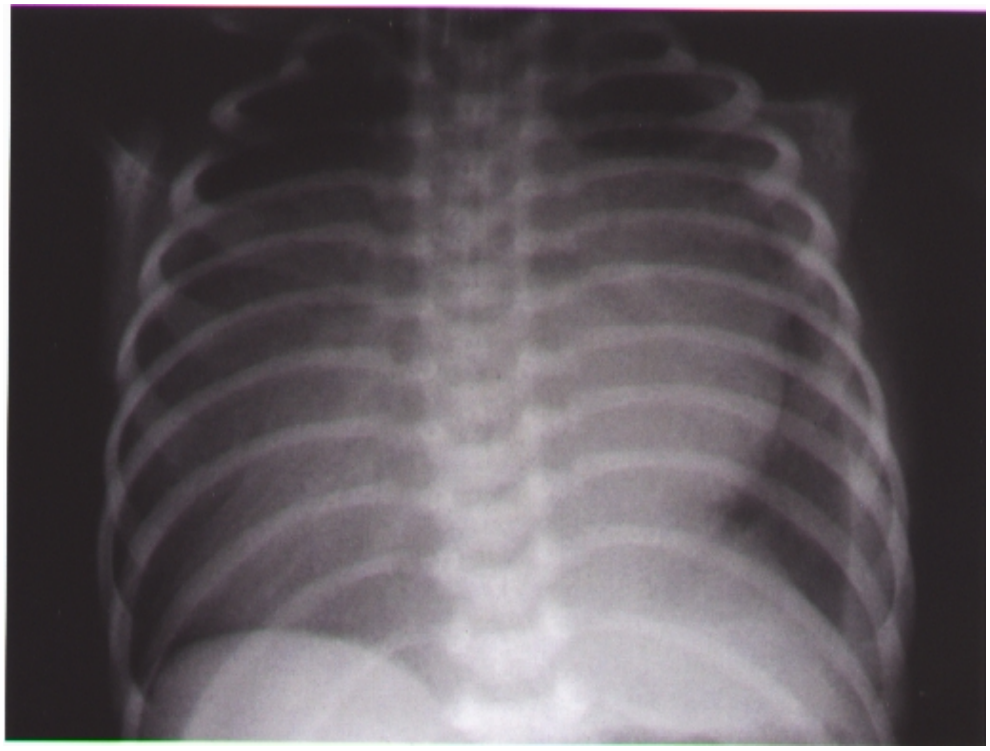

Figure 2. Chest roentgenogram on the second day of life revealing an enlarged cardiac silhouette with left pleural effusion.

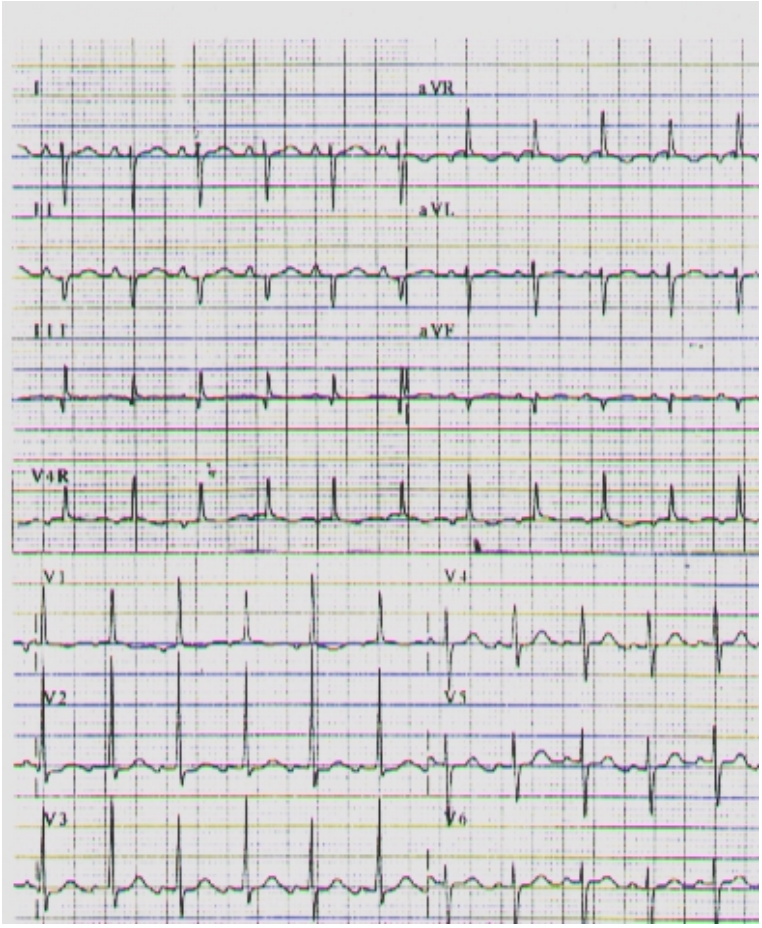

Figure 3. ECG showing electrical alternans in lead I, aVR, and V1-6 with low QRS voltage in lead II, III and aVF.

surgical procedure was to reduce the herniated liver into the peritoneal cavity and repair the diaphragmatic defect. After the operation, the infant was doing well with good lung expansion shown in the chest roentgenogram. The postopera tive course was uneventful and on the subsequent follow up several months later she was healthy with normal growth and development.

Discussion

Peritoneo–pericardial diaphragmatic hernia is a defect in the central tendon of the diaphragm. The etiology and genesis of this rare type of hernia are not clearly understood. Normally, the diaphragm firstly develops as septum transversum, the mesenchymal sheet that locates between the thoracic cavity and stalk of the yolk sac. This sheet provides cells for many structures such as the diaphragm, liver and pericardium. Septum transver-sum will grow posteriorly and become a central part of the diaphragm by fusing with the pleuropericardial fold and mesentery of the esophagus. The defect in the central tendon is thought to be caused by a secondary rupture of this septum rather than fusion failure like

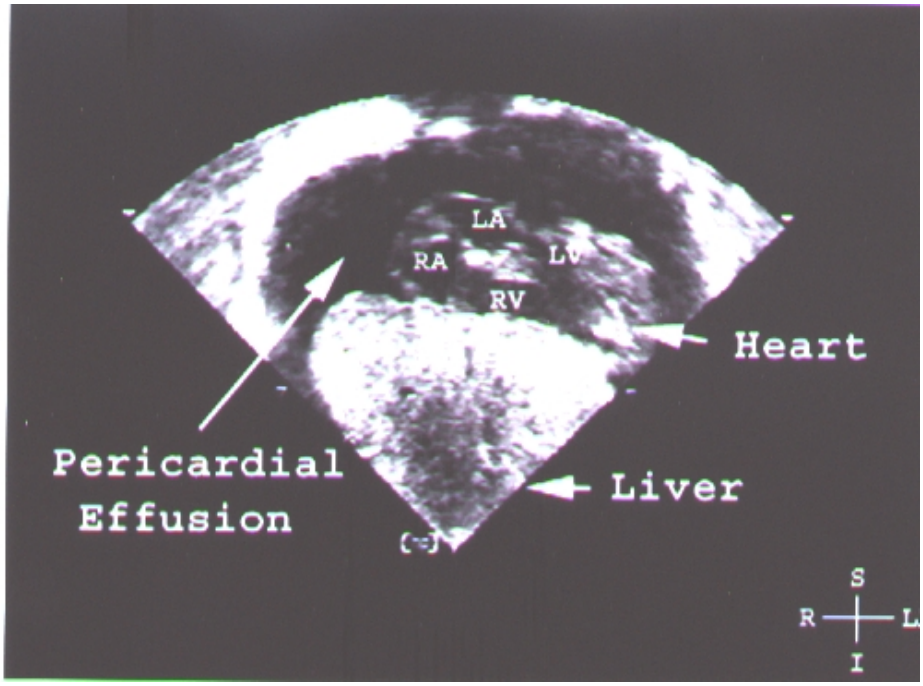

Figure 4. ECG demonstrating massive pericardial effusion with part of the liver seen in the pericardial cavity. (RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle, S: superior, I: inferior, R: right, L: left)

that which occurs in other types of diaphragmatic defect. It might occur around the fourth and fifth week of fetal life, when the liver is growing and expanding rapidly, while the septum has relatively decreased in thickness.[2]

The pericardial-peritoneal communication allows abdominal viscera to protrude into the pericardial sac. The herniated organs reported are the liver, stomach, intestine, omentum and spleen.[2-3] Herniation of the liver has been found to cause massive pericardial effusion.[3–8] The exact cause of effusion formation is still unknown, but there have been two mechanisms postulated. The first one is the effect of mechanical irritation to the pericardium by the intrapericardial mass.[4] Another proposed cause is partial hepatic venous outflow obstruction of the protruding liver, which might cause congestion of that part and lead to transudation within the pericardial sac.[5-6] However, none of the reported cases, including our patient, have shown conspicuous dilatation of the hepatic vein in herniated liver.

The presenting symptom of this condition is respiratory distress soon after birth, but this could manifest as late as one month of age.[3,6] The chest roentgenogram usually reveals a markedly enlarged cardiac silhouette. The diaphragm is clearly seen, but the left

lobe of the liver might not be seen. Da Fonseca JM and Davies MR, *et al.* described this finding as 'the liver cut off sign' and suggested that it might be the point of diagnosis.[5-6] The ECG would demonstrate signs of pericardial effusion such as low QRS voltage and electrical alternans. These findings are not specific for diagnosis and require further investigations.

In earlier reports, a definite diagnosis could be obtained by various techniques such as an injection of carbon dioxide into the pericardial sac with angiography,[4] aortogram via umbilical artery catheter[5] and computerized tomography.[5-7] However, these methods would now be considered unnecessary by the use of ultrasonography, which is noninvasive and provides a specific diagnosis rapidly.[3] The intrauterine diagnosis by ultrasonography has also been reported.[7-8]

Massive pericardial effusion is an unusual occurrence in nonhydropic infants. Apart from intrapericardial liver herniation, other causes could be intrapericardial teratoma,[9] hemangioma,[10] lymphangioma,[11] parvo virus infection[12] and complication of the central venous catheter.[13-14] In most of these cases, the accumulation of effusion is usually rapid, resulting in cardiac tamponade. In our patient, there were no overt clinical manifestations of cardiac tamponade. Measurement of the pulsus paradoxus was not feasible, since the infant did not have intra-arterial blood pressure monitoring. In addition, echocadiographic findings of cardiac tamponade were not apparent, despite the large amount of effusion. This was also found in the majority of previously reported cases, and could be due to the slow formation of fluid that allows a compensatory distension of fetal pericardium. The respiratory failure during early life in this infant could be from pulmonary compression by the expanded pericardial sac rather than cardiac tamponade or severe pulmonary hypoplasia, since it was stabilized by moderate mechanical ventilatory support without requiring pericardiocentesis. The results of pulmonary compression in other cases varied from severe and lethal[5-6] to lack of pulmonary hypoplasia.[3,7-8] In the cases without significant pulmonary hypoplasia, effusion might develop gradually and in late fetal life.

Surgical management of this condition is quite simple. The prognosis of patients depends on the severity of pulmonary hypoplasia and associated congenital anomalies, which were causes of death in some previously reported cases[3-4] but not encountered in our patient.

In conclusion, we have presented an infant with massive pericardial effusion caused by herniation of the liver into the pericardial sac. Although it is an extremely rare type of congenital diaphragmatic hernia, the diagnostic method and surgical correction are very simple and the prognosis is excellent in the absence of pulmonary hypoplasia and other malformations. This entity

should be considered in the differential diagnosis of neonates who have massive pericardial effusion with intrapericardial mass.

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รายงานผู้ป่วยทารกแรกเกิดที่มีน้ำในถุงเยื่อหุ้มหัวใจเนื่องมาจากการเลื่อนของตับผ่านกระบังลมขึ้นไปในถุงเยื่อหุ้มหัวใจ

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ภาควิชากุมารเวชศาสตร์ คณะแพทยศาสตร์ มหาวิทยาลัยเชียงใหม่

บทคัดย่อ รายงานผู้ป่วยทารกแรกเกิดที่มีน้ำจำนวนมากอยู่ในถุงเยื่อหุ้มหัวใจโดยไม่มีอาการแสดงทางคลินิกที่เด่นชัดว่ามีภาวะหัวใจถูกกด สาเหตุมาจากไส้เลื่อนกะบังลมชนิดที่พบได้น้อยมากคือการมีช่องเปิดผิดปกติในกะบังลมทำให้ตับสามารถเลื่อนจากช่องท้องขึ้นไปในถุงเยื่อหุ้มหัวใจได้ การเกิดน้ำจำนวนมากในถุงเยื่อหุ้มหัวใจเชื่อว่าอาจเป็นผลจากการเลื่อนขึ้นไปของตับก่อการระคายเคืองต่อถุงเยื่อหุ้มหัวใจจนมีการหลั่งน้ำออกมามากขึ้น หรือเนื่องจากเส้นเลือดดำในตับส่วนที่เลื่อนขึ้นไปนั้นบางส่วนถูกกดเบียดจนเกิดการคั่งของเลือดทำให้มีน้ำซึมรั่วออกมาได้ ในรายงานนี้ยังได้ทบทวนถึงอาการ วิธีการวินิจฉัย การวินิจฉัยแยกโรค การรักษาและพยากรณ์โรคในทารกที่มีภาวะดังกล่าวที่เคยมีรายงานมาแล้ว **เชียงใหม่เวชสาร 2546;42(1):37-44.**

คำสำคัญ : น้ำในถุงเยื่อหุ้มหัวใจ ไส้เลื่อนกะบังลม