

간 부엽에 의해 심장압박이 동반된 선천성 횡격막 거상증

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= Abstract =

A Central Diaphragmatic Eventration with Accessory Hepatic Lobe Causing Cardiac Compression.

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A case of congenital diaphragmatic eventration on the right and central tendinous portion with accessory hepatic lobe causing direct compression of the right heart is presented. We have performed the video assisted thoracoscopic plication of the right hemidiaphragm and eliminated the mass effect of the accessory hepatic lobe.

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Key words: 1. Diaphragmatic eventration
2. Cardiac compression.
3. Thoracoscopy

Introduction

Congenital diaphragmatic eventration may involve either the right or left diaphragm, or it may be bilateral^{1~3)}. Eventration of the diaphragm on the central tendinous portion with accessory hepatic lobe causing external compression of the heart has not been reported in the other literature^{4~7)}. Herein we describe a first case of the combined right hemidiaphragmatic eventration, plicated by video assisted thoracoscopy and eventration of the diaphragm in the central tendinous portion, namely central diaphragmatic eventration with accessory hepatic lobe that cause

direct compression of the heart and result in deterioration of the hemodynamics(Fig. 1).

Case Report

A 3-day- old boy who weighed 2,885g was referred to our department with cyanosis on crying or feeding. The patient was a full-term baby from the second gestation of a 43-year-old mother. On physical examination, the chest was expanded symmetrically and the cardiac murmur was not auscultated. Blood examination and arterial blood gas analysis on room

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본 논문의 저작권 및 전자매체의 지적소유권은 대한흉부외과학회에 있다.

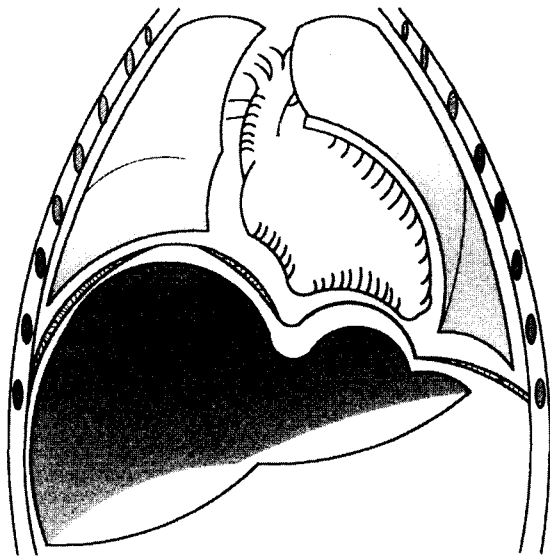


Fig. 1. Schematic presentation of summarized surgical findings. The elevated right and central membranous diaphragm impacted with accessory hepatic lobe were noted

air showed as following: hemoglobin 17.7g/dL, hematocrit 53.5%, serum calcium 6.9 mg/dL, PaO₂ 38.5 mmHg, PaCO₂ 34.2 mmHg and oxygen saturation 75.3%. The chest x-ray performed on admission showed the elevation of the right hemidiaphragm and the presence of a mass like density enlarging the right cardiac profile. Computed tomography of the thorax and transthoracic echocardiogram identified that the right atrium and ventricle were considerably reduced in diameter and compressed on its antero-lateral wall by sonographically homogenous mass(Fig. 2). These findings were considered to be consistent with diaphragmatic eventration caused by accessory hepatic lobe. The respiratory distress progressed, accompanied by progressing hypercarbia and hypoxia.

On fifth day of life, right diaphragm was plicated by video assisted thoracoscopic surgery with previous reported our sliding loop ligation method⁸⁾. Only a small window and one port of access were necessary to plicate the diaphragm and to apply the extracorporeally created sliding knot.

Although his respiratory status was improved gradually allowing discontinuation of mechanical ventilation after operation, cyanosis on the lip was persisted and protracted low oxygen support was required. Serial chest radiographs obtained over the next week and postoperative echocardiogram displayed the remained mass effect to the

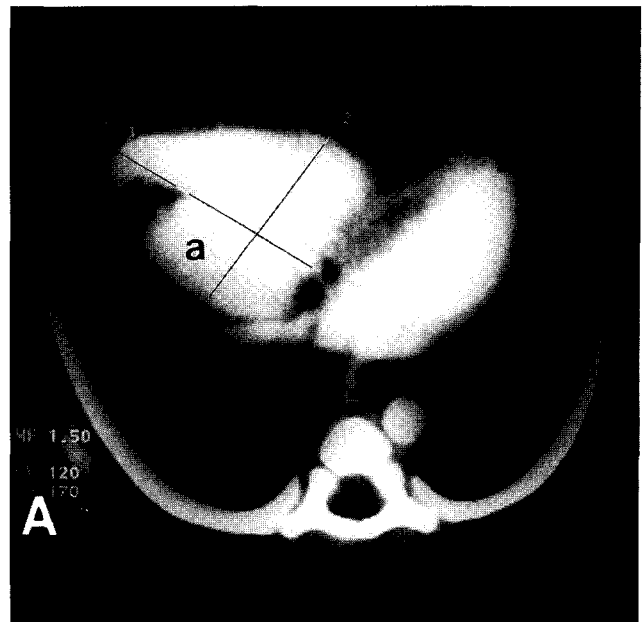


Fig. 2. (A) Computed tomography of the chest shows right and accessory hepatic lobe(a) compressing the right ventricle of the heart. (B) Echocardiography displays external compression of the right ventricle(arrow) by a accessory hepatic lobe and the subsequent reduction of the right ventricle.

right ventricle.

On day-of-life 13, central diaphragmatic plication was performed via a laparotomy. Accessory hepatic lobe shpaed as like as a head of snowman in the right side of the falciform ligament was impacted on the membranous central

portion of the diaphragm. After the falciform ligament was divided, plication was performed using traction sutures on the both muscular margins to eliminate the mass effect of the accessory hepatic lobe to the heart.

Postoperative recovery was uneventful, and the patient was sent home on day-of-life 19. He is well 12 months after operation.

Discussion

Eventration of the diaphragm of the infant has been reported but congenital eventrations on right and central leaflet with accessory hepatic lobe has not reported.

It is now generally accepted that surgical treatment is indicated in symptomatic diaphragmatic eventration. Diaphragmatic plication is technically simple, easy and straightforward with a very low complication rate when carefully performed and a thoracic approach is mandatory for optimal access⁹⁾. Recently, some authors have reported a video-assisted thoracoscopic plication of the diaphragmatic eventration in adult, but not in infancy¹⁰⁾. Narrow intercostal space and difficult one lung ventilation during the operation make it difficult to introduce the video assisted thoracoscopic surgery in neonate. Applying the extracorporeally created sliding knot with the small window and one port of access for thoracoscopy, we have performed right hemidiaphragmatic plication with easy by video-assisted thoracoscopic surgery and concluded that the plication of central diaphragmatic eventration should be strategized via abdominal approach. We believe this is the first report of thoracoscopic plication in the pediatric age group.

The dome of the right hemidiaphragmatic leaf is normally at a higher level than is that of the left. It has been generally thought that this is the result of the hepatic mass beneath the right leaf. However, this view has been brought into question by the studies of Reddy and colleagues¹¹⁾. They insisted that the cardiac mass determined the caudad displacement and lower position of the related hemidiaphragm from the review of sixty-five cases of congenital heart disease. We thought that their analysis based on a supine posteroanterior chest roentgenograms took a little account of gravity of the liver and the development of the

diaphragm. In a neutral upright position, the gravity of the liver is transmitted to the left diaphragm through the falciform ligament and convexed right lobe of the liver may affect the elevation of the right hemidiaphragm. Our case may be one of the evidence of lifting effect of the liver to the diaphragm. We thought what dictates the position of the diaphragm is the liver, not a heart.

We have described a case of combined central and right hemidiaphragmatic eventration causing direct compression of the heart and presented with deterioration of the hemodynamics.

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=국문초록=

선천성 횡격막 거상증은 우측이나 좌측 또는 양측으로 발생할 수 있다고 알려져 있다. 횡격막의 중앙부위와 우측 중앙 전삭부위에서 간 부엽에 의해 심장압박이 동반된 선천성 횡격막 거상증 환자가 있어 비디오 흉강경을 통한 횡격막 주름 성형술을 시행하였던 증례를 보고하고자 한다.

- 중심 단어:** 1. 횡격막 거상증
2. 심장압박
3. 비디오 흉강경