PNEUMONECTOMY IN CONGENITAL LOBAR EMPHYSEMA

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ABSTRACT

Background: Congenital lobar emphysema is a rare congenital disease which caused by anomaly on bronchial cartilage development. Usually, it is limited to a single lobe. It could be treated conservatively or surgically depend on the clinical condition. In this case report we would like to discuss the management strategy for congenital lobar emphysema that affected the whole lung in 17 days old baby.

Case presentation: The patient was admitted to NICU due to respiratory distress. The patient had a difficulty to wean from ventilator. Radiologic examination revealed the whole left hemithorax is filled by multiple bullae which push mediastinal organ contralaterally. Intraoperatively, we found the whole left lung is emphysematous and there is PDA which is not detected preoperatively. Surgeon decided to do pneumonectomy and PDA ligation. After operation, patient recovered on NICU slowly and discharged from hospital 34 days post operation.

Discussion : our case of congenital lobar emphysema which affect one side of lung is a rare case. Our decision to do pneumonectomy was challenging post operatively.

Conclusion: Multidisciplinary team should be involved in treatment of congenital lobar emphysema. Team should be prepared for prolonged intensive care if decision to do lung resection is chosen.

Keywords: Congenital lobar emphysema, pneumonectomy, neonates

Abstrak

Latar belakang: Congenital lobar emphysema adalah penyakit bawaan yang disebabkan oleh anomali pada pertumbuhan kartilago bronkial. Biasanya, penyakit ini terbatas pada satu lobus saja. Penyakit ini dapat ditatalaksana secara konservatif atau secara operatif tergantung kondisi klinis. Pada laporan kasus ini kami ingin mendiskusikan strategi manajemen dari congenital lobar emphysema yang terjadi pada seluruh paru kiri pasien bayi berusia 17 hari.

Presentasi kasus: Pasien dirawat di NICU karena distre nafas. Pasien sulit dilepas dari ventilator. Pemeriksaan radiologis memperlihatkan banyak bula pada hemitoraks kiri yang mendorong organ mediastinal ke kontralateral. Temuan intraoperasi menunjukkan seluruh paru kiri yang emfisema dan terdapat PDA yang tidak terdiagnosis saat pre-operasi. Diputuskan dilakukan pneumonektomi dan ligasi PDA. Setelah operasi, pasien pulih dengan lambat di NICU dan baru dapat pulang dari rumah sakit pada hari ke 34 setelah operasi.

Diskusi: kasus kami dimana congenital lobar emphysema mempengaruhi satu sisi paru adalah kasus yang langka. Keputusan kami untuk melakukan pneumonektomi cukup sulit pada perawatan setelah operasi.

Kesimpulan: Terapi congenital lobar emphysema sebaiknya melibatkan tim berbagai disiplin. Tim harus siap untuk perawatan intensif yang lama jika diputuskan reseksi paru.

Kata kunci: congenital lobar emphysema, pneumonektomi, neonatus

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PNEUMONECTOMY IN CONGENITAL LOBAR EMPHYSEMA : A CASE REPORT

Introduction

Congenital anomaly is a group of disease which arise from abnormality on organ development. It can affect any organ including lung. One of them is congenital lobar emphysema (CLE). CLE is a rare congenital disease which caused by anomaly on bronchial cartilage development. Usually, it is limited to a single lobe. It could be treated conservatively or surgically depend on the clinical condition.
In this case report we would like to discuss the management strategy for congenital lobar emphysema that affected the whole lung in 17 days old baby.

Case presentation

A 12 A 12-day-old baby was consulted to our cardiothoracic surgery due to respiratory distress and difficulty with ventilator weaning.
The baby was born by C-section due to impending eclampsia at 37 weeks gestation.
On the second-day patient gets intubated due to respiratory distress. On 11th day patient was extubated but then reintubated due to desaturation and respiratory distress. Post reintubation patient's plain chest x-ray showed left hemithorax was full of bullae and the heart was pushed to right side. Further investigation with CT-scan confirm multiple bullae on left hemithorax (figure 1).
The patient had increased septic marker procalcitonin by 2.41 and decrease kidney function. So although septic sign was clear, antibiotic choice was limited due to decreased kidney function.

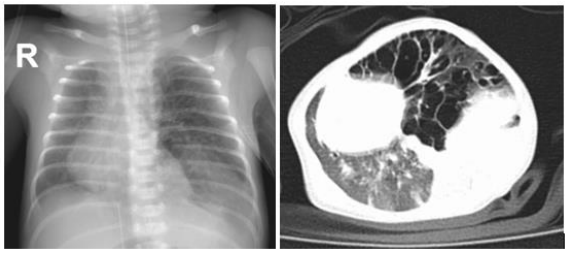

Figure 1. Chest Xray shows hyperaeration on left upper hemithorax which push the mediastinal organ to the right hemithorax **(left).** CT scan shows multiple bullae on left hemithorax (**right**).

Multidisciplinary Team discussion was held shortly and decided to operate on patient as soon as possible with diagnosis of congenital lobar emphysematous with secondary infection. We plan on doing lobectomy with possibility of pneumonectomy since we can't sure wether both left lobe was affected or just one them.
We did left posterolateral thoracotomy when neonate was 17 days old. Intraoperative finding shows whole left lung was emphysematous and there is small PDA of 1 mm (figure 2). We perform left pneumonectomy and PDA ligation.

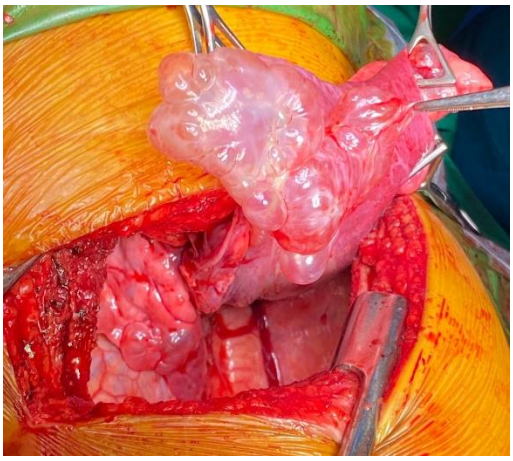

Figure 2. Left lung of a 17 days old neonate notice both upper (blue arrow) and lower lobe (yellow arrow) were emphysematous.

We admit the patient back to NICU post operatively. Patient recover slowly and finally discharged 34 days post-operation (post-operative care is summarized on figure 3).

Figure 3. post-operative timeline.

Discussion

CLE is a rare disease with an incidence of [1] out of 20000-30000 life birth found the most on the left upper lobe (57%), right middle

lobe (30%), then right upper lobe (27%).[1] On this report, 25% cases have two lobes involved.[2] It rarely affects the lower lobe but it can affect both sides of the lungs or whole side of the lung.

Treatment of CLE could be surgically or conservatively. Surgical treatment is usually chosen for patients with a severe cases. In our case, the patient had severe respiratory distress and get intubated. The weaning process was difficult so we decide to operate on the patient.[3]

On preoperative evaluation, we couldn't sure whether the affected was one lobe or a whole lung. From preoperative plain radiography and CT scan evaluation, we suspect only the left upper lobe was emphysematous and the left lower lobe was atelectasis.

About 12-20% of CLE case is concomitant with congenital cardiac disease.[4] Cardiac evaluation to detect congenital cardiac disease must be done on all patients with CLE. We had done echocardiography but we missed PDA on our echocardiographic evaluation. This might be caused by the anatomical disturbance which causes the mediastinal organ to shift to the right and air trapped on the affected lung to disturb the echocardiography imaging. Kylat RI recommends evaluation by CT scan or MRI to evaluate cardiac anomaly on CLE patient.[4]

We found that exposing the hilum intraoperatively was difficult in this case since the left upper lobe was overinflated. To overcome this problem we pull the overinflated lobe out. We could expose the hilum better and inspect another lobe this was.

Pneumonectomy is rarely done for this disease since it's rarely found to affect the whole lung. Tempe et al (2010) described their experience in treating congenital lobe emphysema which affects the whole lung and then underwent pneumonectomy on one patient. In their case, the patient recovers faster post-operatively. The patient was only intubated for six hours and discharged from the hospital after eight days post-operative.[5]

In our case, the patient recovers slowly after the operation. It might be caused by an infection that is difficult to handle and laboratory result which shows bad renal function so we can't aggressively administer antibiotic which also affects renal function. But after the surgery, this case was resolved so we could administer antibiotics more aggressively.

Conclusion

CLE is a rare disease that usually affects only one lobe. Patient with this disease which affects the whole lung is even rarer.

CLE could be managed surgically or conservatively depending on the clinical condition of the patient.

All patients with CLE should undergo a cardiac evaluation to detect congenital cardiac anomalies.

CLE patients whose been undergone pneumonectomy could recover fast if there is no complication which causes patients difficult to wean from respiratory support.

Disclosure:

This patient data has been presented as scientific poster on SEATS 1st inaugural meeting Vietnam.

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