Unilateral Pulmonary Vein Atresia with Absent Right Superior Vena Cava

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A case of unilateral pulmonary vein atresia with absent right superior vena cava in 9 years-old boy is described with a discussion.

Clinical findings were similar to other reports and consisted of hemoptysis, dyspnea and anemia. A chest X-ray revealed irregular vascular marking on left lung field and increased interstitial marking on right upper lung field.

The diagnosis could be confirmed with radionuclide perfusion study, MRI scan of heart and great vessels and angiography which demonstrated a small right pulmonary artery, pruned its peripheral branches, stasis of contrast material and nonvisualization of draining righ pulmonary veins.

Pneumonectomy, patch angioplasty of diaphragm excision will be necessary.

KEY WORDS: Pulmonary vein atresia · Absent right superior vena cava.
증례

환자: 권○일, 9세, 남아.
주소: 반혈, 간헐적 갱렬 및 호흡곤란.
병력: 정상 만삭분만으로 범원에서 출생하였으며 출생시 체중은 2.7kg이었다. 생후 7일경 기침, 갱렬 및 반복으로 A범원에서 치료하였고, 생후 3개월경을 기준으로 청색증이 나타나고 한다. 그 후 B범원에서 2차례에 걸쳐 심도자율과 심혈관조영술 시행받아서 우측 패정맥 폐쇄와 우상대정맥 결손으로 진단되어 수술적 치료를 권유받았으나 거부하였으며, 이 후로도 잦은 패렴과 갱렬이 계속되어 정확한 진단과 치료를 위해 내원하였다.

이학적 소견: 상자의 혈압은 각각 90/60, 120/70으로 정상범위였고, 호흡수는 분당 26회, 맥박수는 분당 130회였다. 신장은 117cm, 체중 18kg으로 모두 소아 발육표준치의 3Percentile이였다. 의식은 명료하였으나 매우 창백했고 청색증은 없었으며, 호흡시 늑간의 함몰이 나타났고 전습부는 hyperactive었다. 홍부 정진상 우폐해부에서 나오(ral)등 흉골간 하부에서 Grade II/VI의 수축기 심침음이 들었으며 P2는 약간 증가되어 있었다. 간장과 비장은 만져지지 않았으며, 수지의 곤봉양 변화나 조갑의 함몰은 없었다.

검사 소견: 홍부 X-선 사진: 좌측 패의 혈관

![Fig. 1. Chest X-ray film shows irregular vascular marking on left lung field and increased interstitial marking on right upper lung field.](http://example.com/fig1)

음영이 퍼들어나게 보여 패정맥 고혈압이 의심되었고, 우측 패의 상부는 간질 음영이 증가되어 있었다(Fig. 1).

심전도 소견: 박동수는 100회/분이었고, QRS축은 100°, P축은 0°, PR간격은 0.11초, QRS간격은 0.08초였다(Fig. 2).

99mTc폐관류추사검사(99mTc MAA lung perfusion scan): 우측 패의 완전혈류결손이 나타났다(Fig. 3).

심혈관계 자기공명 영상: 우상대정맥은 보이지 않았고 좌상대정맥이 높은 관상정맥동을 통해 우심방으로 연결되어 있고(Fig. 4A), 좌측 패에서 두개의 패정맥이 좌심방으로 연결되어 있으나.

![Fig. 2. Standard EKG of the patient.](http://example.com/fig2)
Fig. 3. 99m Tc MAA lung perfusion scan shows total perfusion defect of right lung.

우측에서는 명백한 폐정맥이 보이지 않았고 우측 하부에서 좌심방으로 이어지는 작은 혈관의 존재가 의심되었다(Fig. 4B).

심도자술 및 심혈관조영술: 심도자술상 폐동맥 암이 중등도로 증가되어 있었고 좌우단락은 없었으며(Table 1), 심혈관 조영술상 우측 무명정맥이 좌상대정맥을 통해 우심방으로 연결되었고(Fig. 5), 우폐동맥 조영술상 우폐동맥의 크기가 좌폐동맥에 비해 작았고 말초로 갈수록 직접이 급격히 감소되어 고사목(dead tree)모양을 보였으며 조영제가 좌폐동맥으로 역류하였다. 좌폐동맥의 모양은 비정형적이었고 맥혈관 응영은 좌측 배격에서만 관찰되었으며, levophase에서 두개의 좌폐정맥이 좌심방으로 연결되었으나 우측에서 환관 응영이 인지되지 않았다(Fig. 6).

혈액 및 기타 검사 소견: 말초혈액검사 소견은 심한 정구성 정색소성 변형, 백혈구 감소, 및 혈소판의 증가를 보이고 있었으며(Table 2) 광수검사상 M:E ratio는 10:1로 적혈구 전구세포들이 상당히 감소되어 있었고 광수구체(myeloid series)는 경한 좌반성정을 보이고 있었으며 megakaryocytic series는 정상이었다. 혈청 ferritin은 106 ng/ml, plasma Hb는 21mg%, SGOT 21μL, SGPT 7μL, 혈청총bilirubin치는 0.5mg%로 모두 정상

Fig. 4. MRI scan of the patient
A) left superior vena cava (arrow) drains to right atrium through dilated cornary sinus (arrow head) and right superior vena cava is not visible.
B) two left pulmonary veins are clearly delineated but on right side, a small vessel from lower lung field connected to left atrium but its patency is not clear.
Table 1. Findings on cardiac catheterization

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure(mmHg)</th>
<th>Oxygen saturation(%)</th>
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<tr>
<td></td>
<td>systolic</td>
<td>diastolic</td>
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<tr>
<td>LPA</td>
<td>50</td>
<td>25</td>
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<tr>
<td>MPA</td>
<td>52</td>
<td>28</td>
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<tr>
<td>RV</td>
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<td>0</td>
</tr>
<tr>
<td>RA</td>
<td>a=14</td>
<td>v=9</td>
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<tr>
<td>SVC</td>
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<td>Lt PV</td>
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<tr>
<td>LA</td>
<td>a=22</td>
<td>v=26</td>
</tr>
<tr>
<td>LV</td>
<td>108</td>
<td>5</td>
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<tr>
<td>Aorta</td>
<td>100</td>
<td>76</td>
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고 찰

폐동맥의 환류이상을 동반하지 않은 폐동맥 폐쇄 또는 혈착은 폐동맥고혈압의 드문 원인으로 1951년 Reye가 11) 8세여아를 부검하여 처음 언급한 이후 수십례가 보고되어 있다.

Shone 등 12)은 폐동맥 폐쇄 또는 혈착이 혈착성 심낭염, 종격동염, 폐결핵 또는 종양침습과 같은 후천성 질환에 의해서도 야기될 수 있음을 지적하였으나, 대부분의 보고들은 1〜6, 11〜17) 소천성에 의한 것으로 생각하였다. 훈축성 폐동맥 폐쇄는 발생후기에 측부혈행정맥이 떨어진 후 측력동맥이 좌심방으로 분한전하게 함방되므로서 발생한다고 생각되어진다 13).

폐동맥의 폐쇄로 폐동맥압이 증가되면 폐모세혈관층의 압력에 영향을 미쳐 페임과의 혈류가 증가하고, 기관지 정맥계를 통한 측부혈행이 일어나며, 폐모세혈관의 투과성을 변화시키고, 세동맥 저항의 증가로 환측으로의 혈류량이 감소하게 되며 환측으로부터 건측으로 폐동맥혈의 역류가 일어나 건측으로의 혈류량이 증가하게 된다 1, 16). Ferenz 14)가 정맥폐쇄가 훈축성인 경우에도 폐동맥의 병리학적 변화는 양측성으로 발생한다는 것을 처음 언급하였는데 그 정확한 기전은 알려져 있지 않으나, 환측폐가 굽어져 Compliance가 감소함으로써 폐포 저산소증이 유발되며 이에 대한 반응으로 폐동맥의 수축이 일어나고, 때로 건측 폐의 준임상적 병변도 작용하여 폐동맥의 변화가

Fig. 5. Right innominate venography shows absent right superior vena cava and persistent left superior vena cava which drains into right atrium through coronary sinus.

Fig. 6. Right pulmonary arteriography.
A) Early phase, showing a small right pulmonary artery with dead tree appearance, pulmonary capillary can be seen only on the left.
B) Late phase, showing two left pulmonary veins emptying into a normal left atrium but not visible on right.

<table>
<thead>
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<th>Table 2. CBC finding</th>
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<tbody>
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 pulse high, colored blood, colorless blood, blood pressure, blood cell count, blood test, blood chemistry, blood flow, blood vessel, blood vessel wall, blood vessel lumen, blood vessel dilatation, blood vessel contraction, blood vessel inflammation, blood vessel damage, blood vessel intima, blood vessel media, blood vessel adventitia, blood vessel lumen obstruction, blood vessel lumen enlargement, blood vessel lumen narrowing, blood vessel lumen occlusion, blood vessel lumen stenosis, blood vessel lumen dilation, blood vessel lumen reflux, blood vessel lumen inversion, blood vessel lumen dislocation, blood vessel lumen displacement, blood vessel lumen impaction, blood vessel lumen occlusion partial, blood vessel lumen occlusion total, blood vessel lumen occlusion partial with flow, blood vessel lumen occlusion total with flow, blood vessel lumen occlusion partial with flow obstruction, blood vessel lumen occlusion total with flow obstruction, blood vessel lumen occlusion partial with 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타나는 것으로 보고되어 있다[5,16].

hone 부 X-선 소견은 만성적 정맥정체, 간질성 부종, 폐경색, 반복적 폐렴 등이 복합적인 영향을 받아 나타나며 다양한 전방 및 혈관 음영이 특징적이고, 심장은 정상이나 약간의 즙대를 보이고 폐기능 추가 현저하게[3,4,16].

심전도 검사는 간별진단에 도움이 되지 못하며 정상이거나 우심실 비대의 소견을 보인다[2,7]. 2D 심도초음파와 Doppler 검사는 혈착된 폐정맥을 적절 확인하거나 좌심방으로의 jet를 Doppler로 찾아내는 경우에만 진단에 도움이 될 수 있다[4,7].

방사선 동위원소를 이용한 혈관투사법으로 환자를 응용한 관을 감소하거나 전혀 없다는 것을 확인할 수 있으며, 폐혈기주검사상 환족폐의 환기가 감소되어 있는 경우로도 있다[3,4]. 심혈관계 자기공명영상은 비관찰적 방법으로 진단에 도움이 될 수 있으나 보고된 없다.

전경적인 진단은 심도사진과 선택적 심혈관조영술에 의하는데 대개 여러 정도의 폐정맥 고혈압을 보이고, 환족폐 wedge암의 증가가 나타나며 기 관지동맥과의 축방환 형성으로 환측 폐장막의 산소포화도가 증가될 수 있다[2,4,16]. 폐정맥조영술 상 환족 폐정맥 경지가 감소되어 있고 조영제가 정체를 보이며 건축으로 혈류하고 환측 말초폐장 맥지들이 나무가지처럼 만들어저서 고사막혈당을 보이게 되며 폐정맥 음영은 나타나지 않는다[3,6,16].

저자들의 증례는 흉부 X-선사진상 우측폐 상부의 간질 음영이 증가되어 있었고 좌측 폐혈관 음영이 증가되고 팽창하게 폐동맥압의 증가가 의심되었으며 심전도상 우심실비대의 소견은 없었다. 폐혈관투사상 우측 폐의 음영이 거의 없었고 자기공명영상상 좌측 폐로부터 2개의 폐정맥이 보였고 우측 폐의 음영은 하부에서 하나의 작은 혈관 음영이 나타났으나 개방 가능성이 의심하였다. 심도사진상 중동도 폐동맥압 증가를 보이고 있었으며 폐 wedge암은 측정되지 않았다. 우측폐 조영술상 우측폐막이 고사막 모양을 보이는 등 문헌의 보고들과 일치하였다.

치료는 병리적 양상에 따라 격막결제술(diaphragm excision) [21], patch협착정맥술 [13], 일차적 문합술 [16], 또는 경정맥 풍선정맥술 [22]등이 시행되었으며, 수복이 불가능한 경우는 정맥결제술 또는 폐결재술이 행해졌다[2,3]. 수술적 치료수 익후는 양호한 것으로 알려져 있다[3,13,16,21].

우상대정맥 결손증은 태생기에 precardinal vein들로 연결하는 혈관이 좌하행으로 향하여 우측 anterior cardinal vein이 되행하는 경우에 발생되며 대개 좌상대정맥이 개조하여 간장정맥을 통해 우심방으로 연결된다[23].


좌상대정맥이 지속적으로 혈류적 이상은 아니하지 않으나 신경증세를 사용하는 경우 삼판지 손상 후 부정맥을 피하기 위해 간장정맥동에 손상을 주지 않도록 조심해야 하고, Glenn, Mustard수술을 행하는 경우 역시 주의를 요해야하고, 또한 인공박동기 삼판지 문제를 일으키기도 한다[29].

본 증례의 경우 심전도상 rhythm은 규칙적이었으며 심신등의 병력도 없었고 frontal plane에서 P파의 측은 0°였다.
결론

저자들은 비정상, 간헐적 혼란 및 호흡곤란 등의 증상으로 내원한 9세 남아를 통부 X-선 사진, 삼도, 심도자율 및 혈관조영술로 관찰상 폐장벽 폐쇄와 우상폐경맥 결손으로 진단하였고, 시험적 개통술을 권하였다. 개통술상 병리적 양상에 따라 적절한 술식의 치료가 원활한 증상을 개선시키는 데 도움이 되리라 생각한다.

References

6) 岡本均, 神崎義雄, 馬場清 : 一侧性肺静脈閉鎖を 合併した高血圧症お伴心室中隔欠損症の1 治療例. 胸郭外科. 42 : 965-968, 1989
23) Tavbi H, Kurlander GJ, Lurie PR and Campbell JA : Anomalous systemic venous connection to the left atrium or to a pulmonary vein. AJR 94 : 62-77, 1965
24) Choi JY, Anderson RH and Macartney FJ : Ab-
sent right superior caval vein (vena cava) with normal atrial arrangement. Br Heart J 57: 474–478, 1987


28) Hancock EW: Coronary sinus rhythm in sinus venosus defect and persistent left superior vena cava. Am J Cardiol 14: 608–615, 1964