

Scimitar 증후군 치험 1례

김경희 · 이해용 · 조재민* · 이종국*

= Abstract =

A Case of Scimitar Syndrome

Kyung Hee Kim, M.D., Hae Yong Lee, M.D.,
Jae Min Cho, M.D.,* Jong Gook Lee, M.D.**Department of Pediatrics & Thoracic Surgery,* Wonju College of Medicine, Yonsei University,
Wonju, Korea*

The scimitar syndrome is a rare malfomation that can be defined as a partial or complete right pulmonary venous return into inferior vena cava immediately above or below the diaphragm. This malfomation is often associated with hypoplasia of right lung, anomalous arterial supply of the lower part of the lung, and cardiac dextroversion.

We experienced a case of scimitar syndrome in a 15 years old girl and presenting the case with a brief review of the literature.

KEY WORD : Scimitar syndrome.

Scimitar 증후군 치험 1례

Scimitar 증후군 치험 1례

Scimitar 증후군 (scimitar syndrome)은 폐동맥의 일부 또는 전체가 폐동맥에서 폐동맥으로 연결되어 있는 것으로 정의된다. 이 증후군은 종종 폐의 우측 폐엽의 저형성, 폐의 하부 폐엽의 비정상적인 혈관 공급, 그리고 심장의 좌우 전위와 관련이 있다. 우리는 15세 여자 환자에게 Scimitar 증후군을 경험하였고, 이 경우를 소개하고 문헌을 간단히 검토하였다.

키워드 : Scimitar 증후군.

Scimitar 증후군 (scimitar syndrome)은 폐동맥의 일부 또는 전체가 폐동맥에서 폐동맥으로 연결되어 있는 것으로 정의된다. 이 증후군은 종종 폐의 우측 폐엽의 저형성, 폐의 하부 폐엽의 비정상적인 혈관 공급, 그리고 심장의 좌우 전위와 관련이 있다. 우리는 15세 여자 환자에게 Scimitar 증후군을 경험하였고, 이 경우를 소개하고 문헌을 간단히 검토하였다.

Scimitar 증후군 (scimitar syndrome)은 폐동맥의 일부 또는 전체가 폐동맥에서 폐동맥으로 연결되어 있는 것으로 정의된다. 이 증후군은 종종 폐의 우측 폐엽의 저형성, 폐의 하부 폐엽의 비정상적인 혈관 공급, 그리고 심장의 좌우 전위와 관련이 있다. 우리는 15세 여자 환자에게 Scimitar 증후군을 경험하였고, 이 경우를 소개하고 문헌을 간단히 검토하였다.

“scimitar” 가 (Fig. 1).
QRS 가

scimitar (Fig. 2).

(Aberrent artery)

(Fig. 3).

가

고 안

Scimitar

가

2)

(scimitar)

1960

Neill ¹⁾

Hypo -

plastic lung syndrome³⁾, Congenital pulmonary venolobar syndrome^{4,5,6)}, Epibronchial right pulmonary arterial syndrome⁷⁾, Mirror image lung syndrome⁸⁾

가

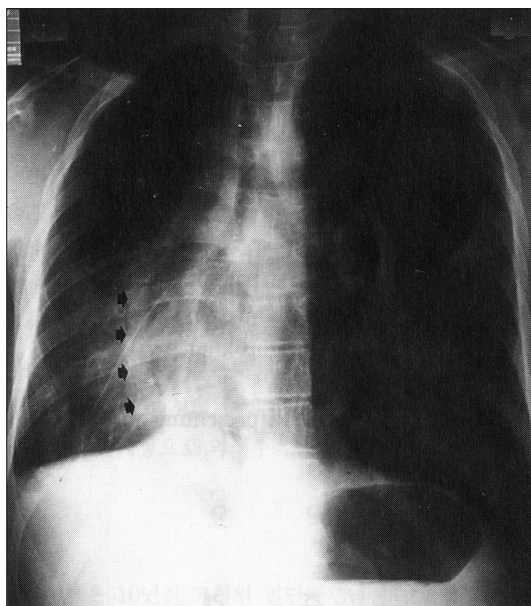


Fig. 1. Chest PA, showing right lung fields are smaller than left lung fields. the heart and mediastinum are shifted to right side. showing the classic “scimitar sign” (arrow).

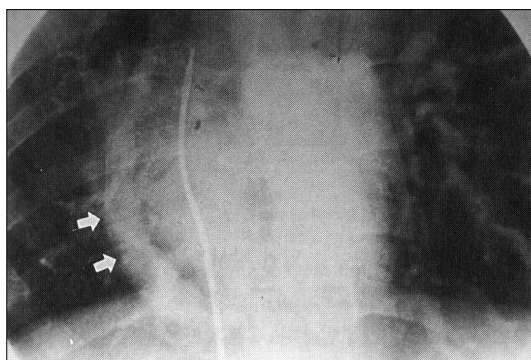


Fig. 2. Right pulmonary angiography. venous blood is drained via a large anomalous vein (arrow) into the inferior vena cava.

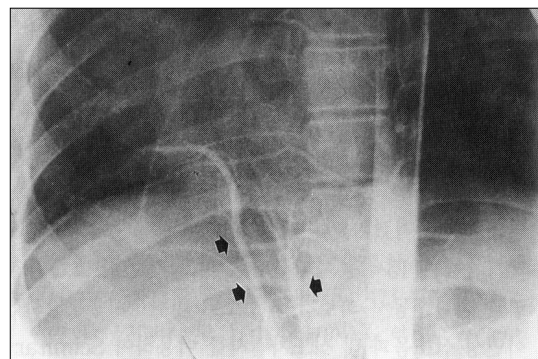


Fig. 3. Abdominal aortogram showing the aberrant artery supplied of right lower lung fields.

- arterial supply and venous drainage ; "scimitar syndrome". *Johns Hopkins Med J* 107 : 1-15, 1960
- 2) Blaysat G, Kachaner J, Villain E, Sidi D, Pedroni E, Piechaud JF, David N : Scimitar syndrome in infants : physiopathology and therapeutic implications in 12 cases. *Arch Fr Pediatr* 44 : 4, 1987
 - 3) Heitzman ER : *The lung : Radioogic-Pathologic Correlations*. 2nd Ed. St Louis, The CV Mosby Co p27-34, 1984
 - 4) Felson B : Pulmonary agenesis and related anomalies. *Semin Roentgenol* 7 : 17-30, 1972
 - 5) Landing BF : Congenital heart disease with trachibronchial anomalies. *Am J Roentgenol* 123 : 679-686, 1975
 - 6) Feldon B : *Chest Roentgenology*. Philadelphia, WB Saunders Co, p87-92, 1973
 - 7) Richter K, et al : The anomaly of the epibronchial right pulmonary artery as guiding symptom of a pulmocardio-vascular syndrome. *Fostchr Roentgenstr* 107 : 31, 1967
 - 8) Dalith F, Neufeld H : Radiological diagnosis of anomalous pulmonary venous connection ; A tomographic study. *Radiology* 74 : 1, 1996
 - 9) Clements BS, Warner JO : The crossover lung segments : congenital malformation associated with a variant of scimitar syndrome. *Thorax* 42 : 6, 1989
 - 10) Kusmotos S, Hoshi H, Koga K, Mizogushi N, Watanabe K : Scimitar syndrome associated with diaphragmatic herniation of the liver. *Radiart Med* 5 : 2, 1987
 - 11) Tomsick TA, Moesner SE, Smith WL : The congenital pulmonary venolobar syndrome in three successive generations. *J Can Asso Radio* 27 : 196-199, 1976
 - 12) Kiely B, Filler J, Stone S, Doyle EF : Syndrome of anomalous venous drainage of the right lung to the inferior vena cava : A review of 67 reported cases and 3 new cases in children. *Am J Cardiol* 20 : 102-116, 1967
 - 13) Mortensson W, Lundstrom NR : Bronchopulmonary vascular malformation syndrome causing left heart failure during infancy. *Acta Radiol* 11 : 449-458, 1971
 - 14) Folger GM : The scimitar syndrome; anatomic, physiologic, developmental, and therapeutic consideration. *Angiology* 27 : 373-407, 1976
 - 15) Hollis WJ : The scimitar anomaly with absent right pulmonary artery. *Am J Cardiol* 14 : 262-265, 1964
 - 16) Halacz NA, Halloran KH, Liebow AA : Bronchial and arterial anomalies with drainage of the right lung into the inferior vena cava. *Circulation* 14 : 826-846, 1956
 - 17) Haworth SG, Sauer U, Buhlmeier K : Pulmonary hypertension scimitar syndrome in infancy. *Br Heart J* 50 : 182-189, 1983
 - 18) Bessolo RJ, Maddison FE : Scimitar syndrome. Report of a case with unusual variations. *Am J Roentgenol* 103 : 572-576, 1968
 - 19) Salazar J : Scimitar syndrome : Five cases examined with two-dimensional and doppler echocardiography. *Pediatr Cardiol* 16 : 283-286, 1995
 - 20) 고재곤 · 김남수 · 김웅흠 · 이홍재 : Scimitar 증후군 1례. *대한소아과학회지* 33(2) : 229, 1990
 - 21) Schramel FHNH, Westermann CJJ, Knaepen PJ : The scimitar syndrome ; Clinical spectrum and surgical treatment. *Eur Respir J* 8 : 196-201, 1995