

# 112 年奇美醫院胸腔內科臨床病例討論會

日期：中華民國 112 年 09 月 05 日(星期二)

時間及地點：16:00-17:00

課程活動題目：**Scimitar syndrome in CXR**

主講人:謝俊民主任

主辦單位：奇美醫院胸腔內科

課程地點:10 樓討論室

教育積分：台灣胸腔暨重症加護醫學會

參加對象：主辦單位所屬院內醫師

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摘要:

Scimitar syndrome, or congenital pulmonary venolobar syndrome, is a rare congenital anomaly that results in hypoplasia or aplasia of one or more lobes of the right lung. Also, there is anomalous pulmonary venous connection on the right to the inferior vena cava. The name scimitar syndrome comes from the appearance of the anomalous vein, which resembles a Turkish sword on the frontal chest radiograph. An absent or small pulmonary artery is also seen, and occasionally rib and vertebral anomalies are seen.

The syndrome may be autosomal dominant in inheritance with variable penetrance. It can be associated with the tetrad of Fallot, or truncus arteriosus. An atrial septal defect can be found in up to 25% of these children.

Radiographically, the right hemithorax appears small with dextroposition of the heart. The anomalous vein may be seen draining medially and inferiorly.

The workup of this patient's symptoms suggested the diagnosis of scimitar syndrome based on the chest PA roentgenogram signs, namely the anomalous right vein and right lung hypoplasia. However a superimposed infection cannot be excluded due to the lack of comparison films.

Although rare, scimitar syndrome is very well described in the literature. Just over 100 cases have been described and all except for two involved the right lung. More women are involved than men (1.4:1). There are two forms: infantile and adult, the former is often more severe than the later.

Radiographic findings on the chest reontgenogram suggest the diagnosis, namely the anomalous vein that travels downward and parallel to the right atrium and then enters the inferior vena cava. The arc formed usually enlarges as it courses downward. Drainage into the inferior vena cava is usually infradiaphragmatic, but in this patient the anomalous vein entered above the diaphragm. The arc-like shadow made by the anomalous vein resembles the blade of a Turkish sword, also called a scimitar.

The constellation of findings found in scimitar syndrome include: hypoplasia of the right lung, associated dextroposition of the heart, systemic arterial supply to the right lung, and the most constant feature an anomalous right pulmonary venous drainage to the inferior vena cava. It is also associated with hypoplasia or absence of the right upper lung segment, atrial septal defects and other cardiac malformations, vertebral anomalies or an abnormal right hemidiaphragm.

Other radiographic techniques can be used to evaluate the anomalous drainage, to include computed tomography, magnetic resonance imaging, angiography, nuclear medicine (V/Q scan to evaluate the hypoplastic lung) and ultrasound (transthoracic echocardiography and transesophageal echocardiography, TEE). Evaluation of the arterial and venous supply to the right lung via angiography provides the definitive diagnosis. One case report suggests TEE to be a promising way to evaluate the anomalous vein, especially if surgery is considered because it is one of the best ways to evaluate the precise location of the anomalous vein draining into the inferior vena cava.

The optimal therapeutic strategy is unknown. Surgery is usually reserved for those with severe symptoms, and signs such as pulmonary artery hypertension and left to right shunts greater than 50%. Surgical techniques include creating an atrial septal defect and baffling the anomalous venous drainage to the left atrium. Pneumonectomy is another option. There are two reports of an interventional radiology approach embolizing vessels with the transcatheter coil occlusion method. As stated earlier, anomalous systemic arteries, usually arising from the descending aorta, can be found feeding the anomalous right lung. These arteries were embolized by Perry et al in 1989 that resulted in complete obliteration of the shunt. For patients who are of older child and adult age, medical therapy such as aggressive management of pulmonary infections seem to be at least as effective as surgical management. Bronchiectasis can be a long-term complication. A ten-day-old male baby was admitted to intensive care unit with respiratory distress. He was a full term baby born at home to a primiparous mother. Immediate postnatal period was uneventful. On fifth day of life he developed respiratory distress and was admitted to the hospital. The baby was cyanotic, had severe tachypnoea with respiratory rate of 110-120/minute and chest retractions. He had tachycardia with heart rate of 160–180/minute, and the heart sounds were audible on the right side of the chest.

There was no evidence of any external congenital anomalies. Crackles on left side of chest were present on systemic examination. Investigations revealed normal haemogram, the X-ray chest showed increased density in the right hemi thorax, absent cardio- mediastinal structures on the left side and hyperinflation of the left lung. The ultrasound of the thorax revealed dextroposition of heart and a large atrial secundum defect. A large anomalous pulmonary vein crossing the diaphragm and joining the hepatic inferior vena cava was noted. Thus the diagnosis of "Scimitar syndrome" was considered in view of partial anomalous pulmonary venous drainage, hypoplastic right lung and dextroposition of the heart. The baby was managed with decongestive measures, antibiotics and ventilator support.

X-ray chest done on seventh day of admission revealed the classic "Scimitar sign" as a result of clearing of right lung of fluid. The baby died on tenth day because of worsening of hypoxia and sepsis.

Case:

The patient is a 20 year-old active duty enlisted female who was evaluated because of a gradual decrease in exercise tolerance over a one-year time span. She reports having frequent respiratory infections as a child and

was slower to recover from them compared to her siblings. She denied any chest or abdominal pain, dyspnea, orthopnea, syncope, nausea, vomiting, fevers or chills, or easy bruisability. Image Findings

Chest PA reontgenogram without comparison films demonstrates right-sided volume loss and an asymmetric opacity in the right chest. There is a dominant vertical vein in the medial right chest and is prominent on the lateral view. The right hemidiaphragm is elevated. There is scoliosis of the thoracic spine on the frontal view and loss of thoracic kyphosis on the lateral view. Differential Diagnosis include Swyer-James syndrome , Effects of radiation therapy.