Case Report

Scimitar Syndrome: Plain Radiographic and CT Scan Appearances

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Abstract

Scimitar Syndrome, Hypogenic Lung Syndrome or Halasz Syndrome [1] is a rare congenital anomaly. The eponym "Scimitar Syndrome" is a term which has principally been used to describe the appearance of one specific type of anomalous connection of the right pulmonary vein to the systemic venous system, at a point near the level of the right leaf of the diaphragm.

The term has been derived from the scimitar shaped configuration of the right pulmonary in its course through the right hemithorax [2] which can be visible roentgenologically as in our case where we present the plain radiographic and CT appearances of a single case of a right inferior pulmonary vein draining into the inferior vena cava (IVC) which represents the most common presentation of scimitar syndrome.

Keywords: Scimitar, partial anomalous venous return, computerized tomography, pulmonary venolobar syndrome, hypogenetic lung syndrome.

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Introduction

Scimitar syndrome is a rare congenital anomaly consisting of a partial anomalous venous return from the right lung to the IVC, hypoplasia of the right lung with the dextraposition of the heart and an anomalous systemic supply from the abdominal aorta to a portion of the right lung (usually the lower lobe). Not all patients with scimitar syndrome will have all the above mentioned findings, but the most common is the anomalous pulmonary venous return os all or the most of the right lung to the IVC creating the image of a Turkish sword on a chest radiograph. The variability and pathological spectrum of this disorder has been well described by other investigators. [1-3]

Other cardiac anomalies may also coexist. The clinical presentation of these patients is quite variable– from asymptomatic to a significant heart failure– and because of that, the age at presentation can differ significantly, from infancy to adulthood. For most cases, radiographic investigation has provided the best available anatomic assessment. Plain Radiography, computerized tomography, pulmonary angiography and bronchography have all been revealing.

Case Report

A 30 year old female patient presented to the E/R with hemoptysis. A plain film was done and revealed a linear opacity in the right lower lung fields which (Fig 1, 2) was proved by a spiral CT scan (Fig 3) with IV contrast to be an aberrant right pulmonary vein connected to IVC (Fig 4).

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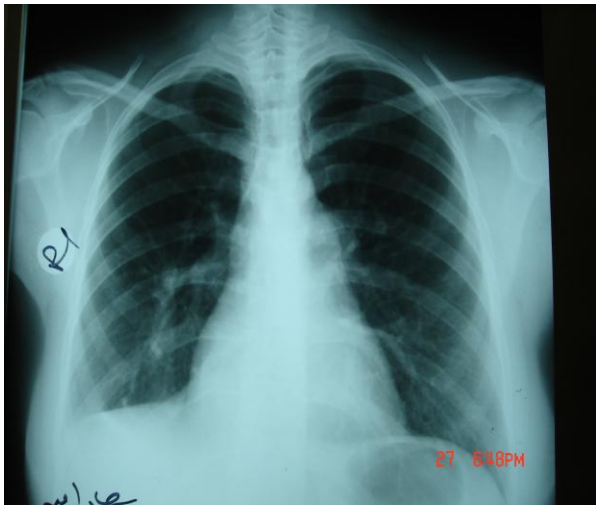

Figure (1): Chest X-ray P.A view demonstrates the curved linear shadow of an anomalous vein.

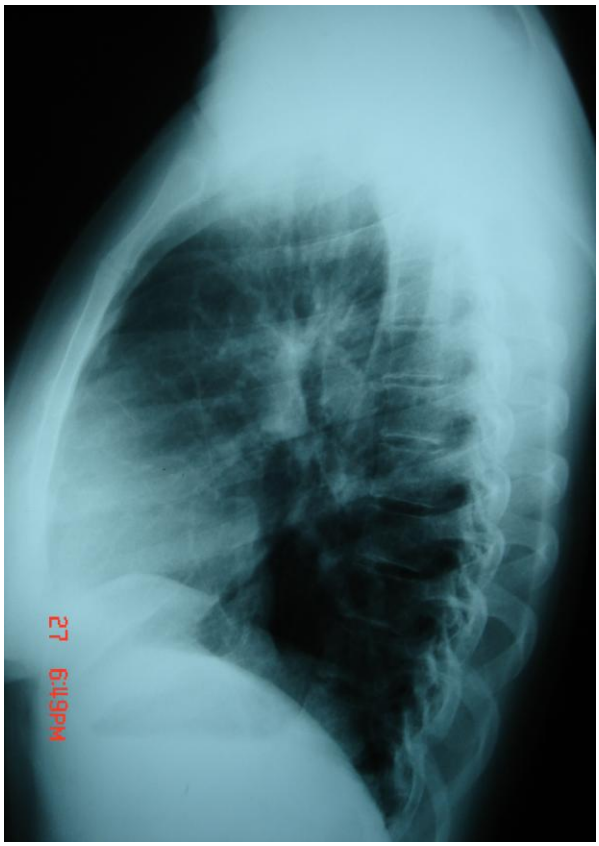

Figure (2): Chest X-ray lateral view showing a linear shadow overlying the heart shadow.

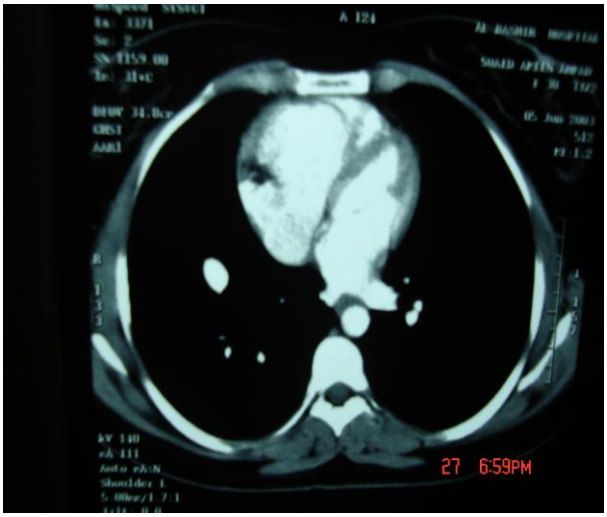

Figure (3): Chest CT scan axial view showing the right anomalous pulmonary vein as a round density in the lower lung fields.

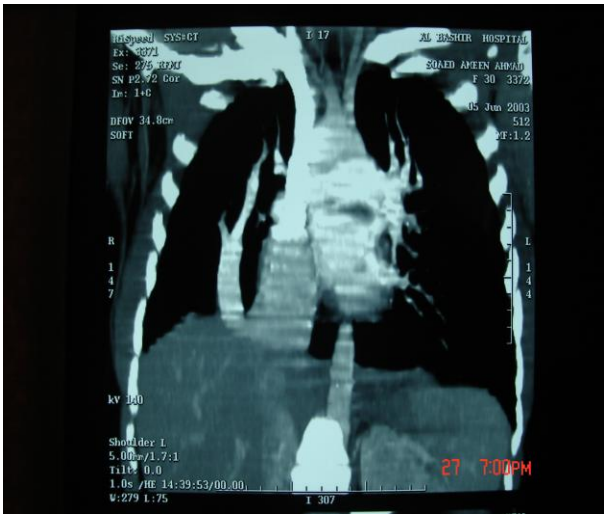

Figure (4): Chest CT scan coronal reconstruction showing a linear curved density connected to IVC which represent an anomalous right pulmonary vein.

Discussion

The first case report of pulmonary venous drainage into the inferior vena cava was published in 1836 by Cooper.[4] The scimitar syndrome was named by Neil et al. in 1960 describing a syndrome of partial anomalous pulmonary venous drainage of the right lung into the inferior vena cava.[1] Of the 100 cases, 98 cases were right sided and there were more females than males involved with a ratio of 1.4/1.[5] There

were only 2 cases which proved to be familial.[1,2]

Manifestations

Scimitar Vein: The most constant abnormality is the anomalous vein draining a variable amount of blood into the inferior vena cava. The vein is usually visible on plain radiographs but may be obscured behind the heart; the vein is not always wide and curved like a scimitar but may be thin, straight or multiple. Drainage is usually to the IVC below the diaphragm, less often above the diaphragm, but the drainage can also be to the right atrium or even to the left atrium.[6] The drainage of the right lung to the inferior vena cava constitutes a left – to – right shunt, which is usually small, less than half the systemic flow.[7,8]

Malformation of the Right Lung: A degree of hypoplasia is found in almost all cases. This causes a shift of the mediastinum and heart to the right with errors of segmentation and lobulation. When one fissure is absent, usually the minor fissure, the lung is bilobed mimicking the left lung. If both fissures are absent, the lung is unilobed.[7, 9]

Abnormal Arterial Supply: The degree of reduction in the size of the right pulmonary artery reflects the degree of lung hypoplasia.There is also a systemic arterial supply to the lung and it could be either from the abdominal aorta which gives a branch that penetrates the diaphragm and supplies the right inferior pulmonary lobe or less often from the descending aorta. These arteries are not bronchial arteries since they adopt the pulmonary artery distribution once they enter the lung.[8]

Cardiovascular Malformations: Dextrocardia of the lung reflects hypoplasia of the right lower lung lobe. About 25% of the cases are associated with cardiovascular malformations. This congenital heart disease is more prevalent in cases diagnosed in childhood rather than adult diagnosed cases.[10] The most common congenital heart disease is atrial septal defect (ASD) followed by teratology of Fallot, and less commonly there is patent ductus arteriosus (PDA), coarctation of the aorta, ventricular septal defect (VSD), and double chambered right heart.

Noncardiovascular Malformations:
Abnormalities of the right hemidiaphragm (eventration, bochdalek hernia, and accessory leaf of hemidiaphragm) are mostly involved with noncardiovascular malformations. Less commonly hemivertebra have been reported in a few cases.

Role of CT in Diagnosis: A CT scan helps in clarifying anatomy by showing the abnormal vein that inserts into the inferior vena cava even if it is masked by the heart on a plain radiography. It also shows the reduced size with hyperlucency and distorted vessels of the involved hypogenetic lung. A mediastinal shift and abnormalities of the diaphragm are also noticeable on computerized tomography.

Differential Diagnosis

When the scimitar vein is shown on a plain radiography, the diagnosis is straight forward, but when it is masked by the heart, the diagnosis of dextrocardia, hypoplastic lung or Macleod syndrome may be mistakenly made. However, in patients with Macleod syndrome at full inspiration, ipsilateral air-trapping due to bronchial obstruction is observed by roentgenography.[11] We did not observe this sign in our patient. Also, reduction in both ventilation and perfusion are found in perfusion-ventilation lung scans of Macleod's patients.

Conclusion

To our knowledge this is the first reported case of adult Scimitar syndrome in Jordan. The diagnosis can be suspected on a plain radiography, but confirmation needs a CT scan with IV contrast to delineate the anomalous pulmonary vein drainage into the inferior vena cava.

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متلازمة سجميتار: مظاهر الصورة الشعاعية العادية والصورة الطبقية المحورية

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الملخص

متلازمة سجميتار هي تشوه خلقي نادر يتميز بشكل رئيس بتشوه في العود الوريدي الرئوي وبشكل خاص الوريد السفلي الرئوي الأيمن؛ حيث يصب في الوريد الأجوف السفلي مع نفرات نادرة في الأوعية الرئوية الأخرى، والتي تصب بشكل شاذ. في هذه الحالة لدينا صورة شعاعية عادية وصورة طبقية محورية لهذه الحالة المنفردة لوجود وريد سفلي رئوي أيمن يصب في الوريد الأجوف السفلي، وهذا يظهر أهم دلائل متلازمة سجميتار.

الكلمات الدالة: سجميتار، تشوه جزئي في العود الوريدي الرئوي، التصوير الطبقي المحوري، متلازمة الوريد الفصي الرئوي.