Horseshoe Lung and Scimitar Syndrome Associated with Congenital Hydrocephalus in an Infant: Case Report

At Nalı Akciğer ve Scimitar Sendromuna Eşlik Eden Konjenital Hidrosefalili Bir İnfant

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ABSTRACT Horseshoe lung is a rare congenital anomaly characterized by the presence of unilateral pulmonary hypoplasia and a midline isthmus bridging the right and the left lung. In this anomaly, the lower lobe of the right lung is hypoplastic and supplied with arterial blood from the descending aorta. Its pulmonary venous return is connected to the inferior caval vein. Left sided scimitar syndrome and bilateral forms are rarely reported in medical literature. Almost all cases of horseshoe lung are associated with the scimitar syndrome. Cranial abnormalities with these malformations are extremely uncommon. We describe a symptomatic infant with a combination of typical scimitar syndrome and horseshoe lung associated with congenital hydrocephalus. Diagnosis was established by echocardiography, angiocardiography and computerized tomography. To the best of our knowledge, this case with these malformations seems to be the first which is reported in the literature.

Key Words: Hydrocephalus; scimitar syndrome

ÖZET At nalı akciğer tek taraflı pulmoner hipoplazi varlığı, sağ ve sol akciğeri orta hatta birleştiren istmus ile karakterize olup nadir görülen bir konjenital anomalidir. Bu anomalide sağ akciğerin alt lobu hipoplaziktir ve arteryel kanlanması inen aorta tarafından sağlanmaktadır. Pulmoner venöz dönüşü vena kava inferiora olmaktadır. Bilateral ve sol taraf Scimitar sendromu formları literatürde nadiren bildirilmiştir. Hemen hemen tüm at nalı akciğer vakaları Scimitar sendromu ile ilişkilidir. Bu malformasyonla kranial anormallik birlikteliği ise son derece nadir görülür. Bu yazıda, tipik Scimitar sendromu ve at nalı akciğer kombinasyonuna eşlik eden konjenital hidrosefalili semptomatik bir olgu sunuldu. Tanı ekokardiyografi, anjiyokardiyografi ve beyin tomografisiyle konuldu. Bilgilerimize göre literatürde bu malformasyonların birlikte görüldüğü bir vaka bildirilmemiştir.

Anahtar Kelimeler: Hidrosefali; scimitar sendromu

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Horseshoe lung (HL) is a rare congenital anomaly characterized by the presence of unilateral pulmonary hypoplasia and a midline isthmus bridging the right and left lung. It was first described in 1962 by Spencer.[1] This anomaly is almost always associated with Scimitar syndrome or hypogenetic lung syndrome.[2]

In this lesion, the lower lobe of the right lung is hypoplastic and supplied with arterial blood from the descending aorta. Its pulmonary venous return is connected to the inferior caval vein. Left sided Scimitar syndrome

and bilateral forms are rarely reported in medical literature.[3-6] Radiologically, this anomalous vein appears as a Scimitar-like shadow (Turkish Swords) on chest X-Ray, and runs from the middle of the right lung to the cardiophrenic angle.[7]

Heart malformations are the most common forms of birth defects but the association with central nervous system anomalies (CNS) are unusual.[8,9] We described the first reported case of HL which occured in conjunction with Scimitar syndrome and congenital hydrocephalus, to our knowledge.

CASE REPORT

A 6–month-old boy was referred to our clinic for evaluation of recurrent pulmonary infections and congenital heart disease. The physical examination demonstrated an enlarged head and fontanels, bilateral diffuse rhonchi and systolic murmus at the left sternal border. The frontal plain chest radiography showed dextroposition of the heart, hypoplasia of the right lung and a nonhomogenous opacity in the right lung base (Figure 1). Echocardiography demonstrated large high venosum atrial septal defect (ASD), pulmonary hypertension (PH) but right pulmonary veins couldn't be detected. Cardiac catheterization with angiography revealed suprasystemic pulmonary artery pressure, ASD with a large left to right shunt, hypoplastic right pulmonary arteries and branches which course to the lower lobe of the left lung. All the left pulmonary veins and upper right pulmonary vein appear to drain normally. The right lower pulmonary vein was seen to drain to the inferior caval vein (Figure 2 A,B). Non-invasive angiogram using computed tomography (CT) demonstrated that large systemic collateral artery from the abdominal aorta supplied the lower half of the right lung (Figure 2C). To investigate further cardiopulmonary and also cranial abnormalities, we performed a contrast enhanced computerized tomographic scan of the chest and cranium. CT of the thorax showed fusion of the posterobasal portions of both lungs behind the heart consistent with HL and rightward shift of the heart were observed (Figure 3). CT of the cranium demonstrated normal fourth ventricle,

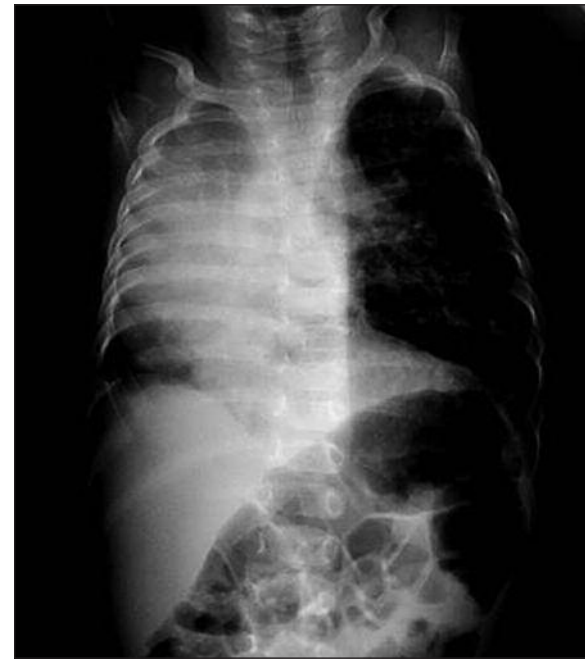

FIGURE 1: The frontal chest radiography shows hypoplasia of the right lung with the heart displaced to the right.

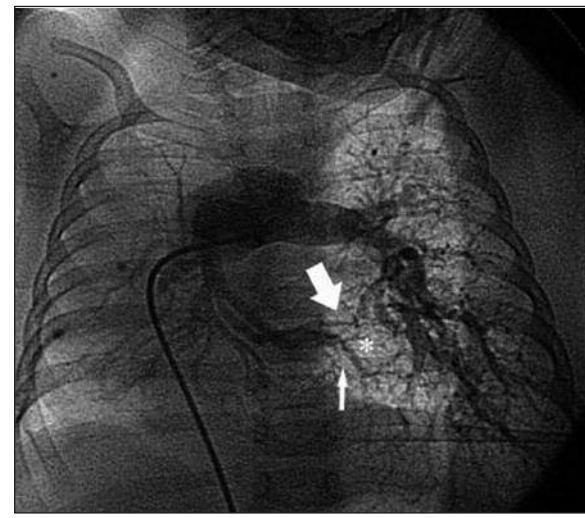

FIGURE 2A: Pulmonary angiogram showing hypoplasia of right pulmonary artery. Abnormal branch from right pulmonary artery extends accross midline to lower lobe of left lung (arrows and asterics *).

and enlarged third and lateral ventricles (Figure 4). Congenital hydrocephalus due to the stenosis of the aquaductus sylvii was diagnosed. An abdominal ultrasound demonstrated normal liver, spleen and kidneys. Digoxin, furosemide and inhaled iloprost was started for his cardiac failure and pulmonary

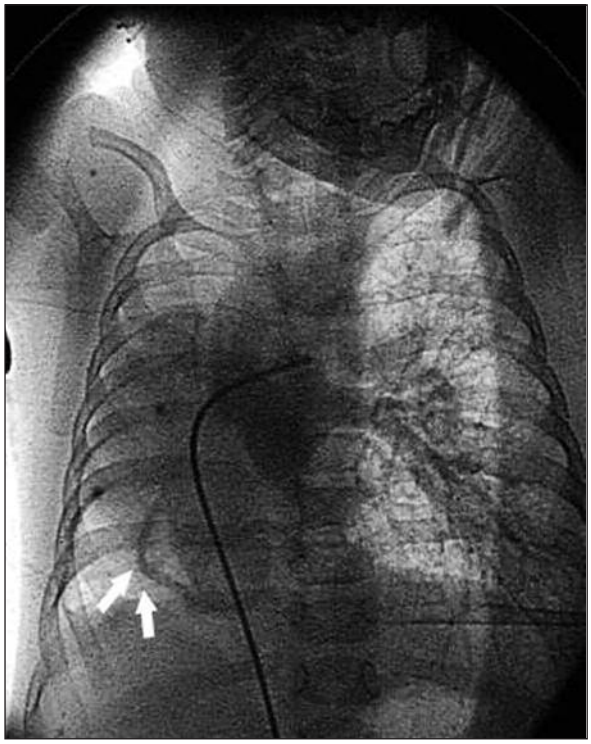

FIGURE 2B: Levophase of pulmonary artery injection. Right pulmonary venous return to inferior vena cava via Scimitar veins (arrows).

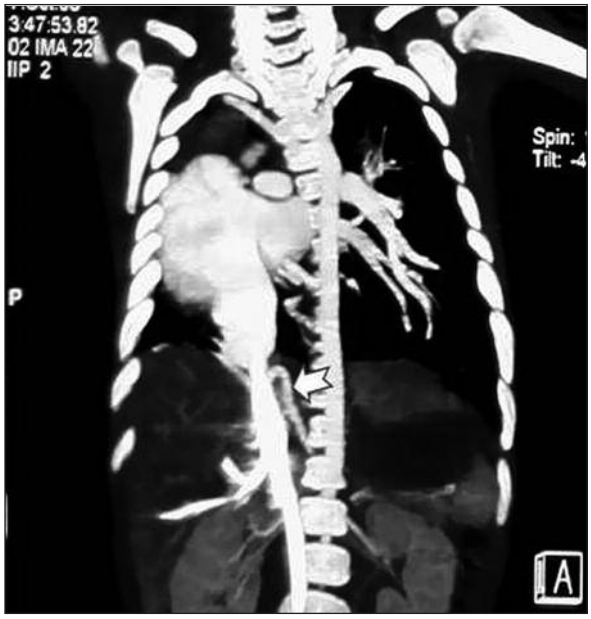

FIGURE 2C: CT Aortogram shows a systemic collateral artery (arrow) supplying a segment of the right lower lobe.

hypertension. Because of the severity of the cardiopulmonary lesions, the associated cranial anomaly, the young age and his low weight, corrective surgery has presently been judged inadvisable in view of the life threating risks. He referred to neurosurgery department for his hydrocephaly but the parents of the patient declined any surgical options. The patient was discharged home in stable condition to be followed as an outpatient.

DISCUSSION

HL deformity is a rare congenital pulmonary anomaly of childhood characterized by fusion of the posterobasal portions of the right and left lung through a narrow isthmus between the heart and aorta.[1]

Pulmonary hypoplasia on the right side is a constant feature of HL. Left sided lung hypoplasia is reported rarely.[3] The majority of reported cases of HL in conjunction with Scimitar syndrome which is an anomaly characterized by hypoplasia of the right lung and its artery, anomalous drainage of the right pulmonary veins, systemic arterial supply to portion of the lung, anomalies of bronchial trees and cardiac dextrorotation or dextroversion. This anomaly is usually located unilaterally, often on the right side. Left sided or bilaterally Scimitar

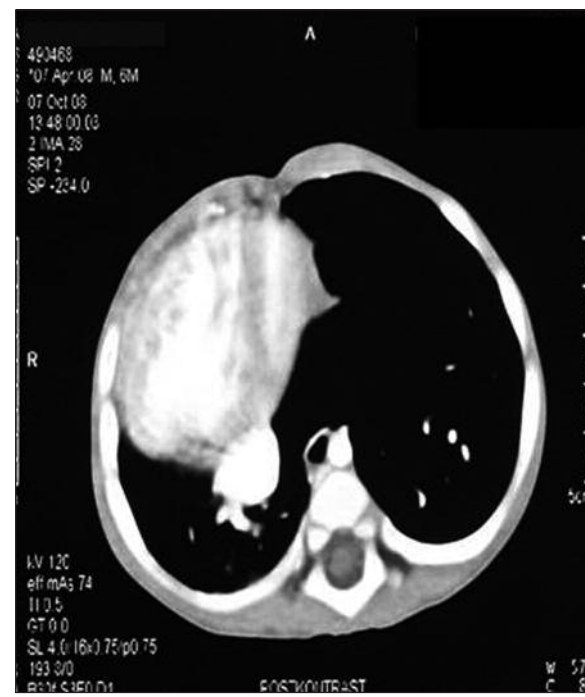

FIGURE 3: Thoracic computed tomography image at the level of the lung bases demonstrating horseshoe lung deformity.

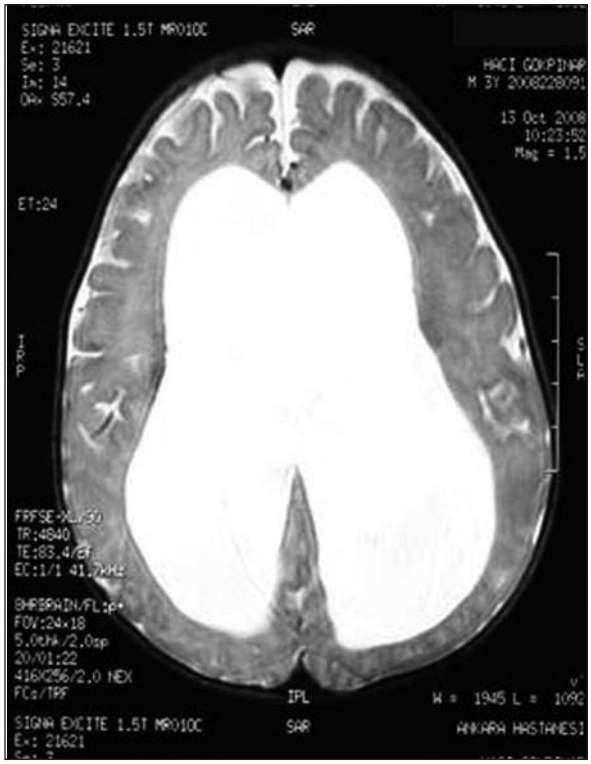

FIGURE 4: Cranial computed tomography shows hydrocephalus.

syndrome has been reported very rarely.[3-6] Its incidence is estimated to be from 1 to 3 per 100.000 live births.[10]

The embryological explanation for HL is unclear. The development of HL is thought to be associated with an arrest in development of the lung tissue differentiation early in embriogenesis, arround the 3rd week of gestation. Pulmonary agenesis is thought to be related to a vascular disruption and may also result in abnormal venous return. The timing of this disturbance parallels septation of the cardiac tube and suggests that a coincident early disturbance leads to the associated cardiac anomalies.[11-13] Congenital heart disease is present about 25% of the cases and at least half of the lesions is ASD.[12] Other cardiac malformations reported include ventricular septal defect (VSD), patent ductus arteriosus (PDA), tetralogy of Fallot, aortic coarctation, pulmonary sling, hypoplastic left ventricle.[11,14-19]

Although craniofacial malformations and CNS abnormalities has not been mentioned with HL, these are rarely reported with Scimitar syndrome. Ruggieri et al. reported Scimitar vein anomaly with multiple cardiac malformations, craniofacial and CNS abnormalities in a brother and sister.[8] D'Allessandro et al. reported a case of HL lung in association with facio-auriculo-vertebral sequence.[9]

The present case had HL, partial anomalous venous return of the Scimitar vein type, ASD, systemic to pulmonary collateral vessels, pulmonary hypertension and cardiac dextroposition. He had also congenital hydrocephaly. To our knowledge, the association of HL, Scimitar syndrome and cranial malformations have not been reported before. This is the first description of this incidence.

Most patients with Scimitar syndrome are relatively asymptomatic. The patients with a coexistent HL diagnosed in infancy often has pulmonary problems and cardiac failure. The degree of pulmonary hypoplasia, associated cardiac malformations and pulmonary hypertension contributes to the high mortality rates observed.[19-21]

Our patient had been admitted to our unit for evaluation of recurrent pulmonary infections and the suspicion of a congenital heart disease. After chest radiography and echocardiography we thought right lung hypoplasia and right anomalous partial pulmonary venous return. We couldn't exactly assess the return of right pulmonary veins by CT. Cardiac angiography was performed for the definition of abnormal venous return and associated cardiac malformation.

CT and angiocardiography confirmed the diagnosis of Scimitar syndrome with HL, ASD and pulmonary hypertension. Digoxin, furosemide, inhaled iloprost and antibiotherapy were started for relief of cardiac failure, pulmonary hypertension and pulmonary infection. We didn't plan any surgical repair for his cardiopulmonary malformations because of high risk. He was referred to of neurosurgery department for his hydrocephaly but the parents of the patient declined any surgical options.

In conclusion, the association of HL, Scimitar syndrome and congenital hydrocephaly have not been described before, to our knowledge. Echocar-

diography, CT and/or MRI are the best noninvasive diagnostic techniques for complex combination of vascular and parenchymal anomalies.[6,22-24] Cardiac catheterization and angiography can be required for confirming diagnosis and associated cardiac malformations.[5]

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