INTRODUCTION

Congenital bronchial atresia is a rare disorder. It was first reported by Ramsay and Byron in 1953.\(^1\) Obstruction in the bronchial origin results in enlargement of the peripheral bronchus, mucous accumulation, and emphysematous changes in the peripheral lung.

The lung buds develop from the foregut and drain to the splanchnic venous system of the embryo. A vascular insult occurring after the 15th week and before birth could cause a focal obliteration of a segmental bronchus with normal remaining distal lung.

Anomalous pulmonary venous return signifies a total or partial draining of pulmonary venous blood into the systemic veins or directly into the heart. The common pulmonary vein develops as an out pouching of the primitive left atrium and then fuses with the primitive lung vasculature followed by obliteration of the connections of the lung buds to the splanchnic veins. If the common pulmonary vein becomes atretic, either totally or partially, while pulmonary to systemic venous connections are still present, total or partial anomalous pulmonary venous connection results.\(^2\)

Most patients are diagnosed accidentally during a screening chest radiograph. Only one third of patients are symptomatic and present with cough, dyspnea and infection.

CASE REPORT

A 21 years old male patient visited PNS Shifa Hospital for routine medical examination. His CXR demonstrated branching opacity in the left upper lung (Figure 1a, b).

CT chest showed a branching opacity in the left upper zone surrounded by characteristic low attenuation lung parenchyma consistent with hyperinflated lung (Figure 2a, b).

**Figure 1 (a, b):** CXR showing branching opacity in left upper zone.

**Figure 2 (a, b, c, d):** Axial and sagittal slice CT chest showing branching opacity in the left upper lobe surrounded by characteristic low attenuation lung parenchyma consistent with hyperinflated lung (a, b). Coronal reformate, showing aberrant opening of the left superior pulmonary vein into the left brachiocephalic vein (c, d).

ABSTRACT

Bronchial atresia and anomalous superior pulmonary venous return into the left brachiocephalic vein are unusual anomalies. We present a young asymptomatic man in whom these two distinct anomalies were co-existingly found on routine medical examination. Patient was diagnosed as a case of congenital bronchial atresia with partial anomalous pulmonary venous return based on the evidence of special features on X-ray chest film, characteristic contrast enhanced 16-slice CT chest findings, lack of symptoms and no change on follow-up X-ray chest films.

**Key words:** Bronchial atresia. Bronchocele. Partial anomalous pulmonary venous return.

Department of Radiology, PNS Shifa (Naval Hospital), Karachi.

Correspondence: Maj. Tariq Saeed Siddiqui, Head of Radiology Department, Sheikh Khalifa Bin Zayed Hospital (CMH MZD), Azad Kashmir.

E-mail: tariqssr@gmail.com

Received January 19, 2010; accepted March 29, 2011.
Contrast enhanced axial sections with coronal and sagittal reformats revealed aberrant opening of the left superior pulmonary vein into the left brachiocephalic vein (Figure 2c, d). Arterial supply was from left pulmonary artery. No systemic supply was seen from aorta. The diagnosis was bronchial atresia with partial anomalous pulmonary venous return and followed in the OPD. Follow-up X-ray chest after 6 months revealed no change.

**DISCUSSION**

Congenital bronchial atresia (CBA) is an unusual pathology. CBA belongs to the category of broncho-pulmonary anomalies. Obstruction in the bronchial origin results in dilatation of the peripheral bronchus filled with mucous and surrounded by emphysematous changes in the adjacent lung. Emphysematous changes result from collateral ventilation through the pores of Kohn and canals of Lambert in the surrounding lung and bronchi, and check valve mechanism.

Emphysematous changes can result in bulla formation and pneumothorax from rupture of the bulla.3 However, hyperinflation of the pulmonary parenchyma is not an essential feature in lobar bronchial atresia. Bronchial atresia without hyperinflation is reported by many.4,5 Bronchial atresia generally produces no symptoms. It can present with pulmonary symptoms such as fever, cough, or shortness of breath due to recurrent pulmonary infection or over inflation of the involved lung parenchyma.6 The common sites of CBA have been reported to be the left upper pulmonary lobe (64%), followed by the left lower lobe (14%) and the right upper lobe (12%). The most common site is the left upper lobe, particularly of the apical-posterior segment.6

The cause of bronchial atresia is controversial. Bronchial tree distal to atresia branches normally, it has been postulated that the insult may have occurred after completion of the process of airway development at about 16 weeks. Co-existence of other congenital defects that occur early in embryological development (as in this case) supports the hypothesis that an insult occurs during the stage of lung budding resulting in a localized abnormality followed by normal development of the distal bronchial tree. Bronchial atresia can result from compression by the pulmonary artery when superior transposition of the superior lobar bronchus is present on the left side; however, superior transposition, tracheal bronchus, is more common on the right side.7

Partial anomalous pulmonary venous return (PAPVR) involves the right lung more frequently than the left.8 The overall incidence of PAPVR is approximately 0.5%.9 Anomalous veins of the left lung most often drain the left upper lobe. After entering the mediastinum, these pulmonary veins continue cephalad and lateral to the aortic arch in a vertical vein that then joins the left brachiocephalic vein, as in this case. Anomalous right lung veins can drain into the superior vena cava (SVC), azygous vein, right atrium, coronary sinus, or inferior vena cava (IVC). Most frequent is an isolated minor PAPVR of the right upper lobe draining to the SVC.9

Bronchial atresia needs to be differentiated from pulmonary sequestration. Pulmonary sequestration is defined as segment of lung parenchyma separated from tracheobronchial tree and receiving its blood supply from systemic artery rather than pulmonary artery. Arterial supply is usually from aorta. It is a rare congenital malformation that can present as a solid or cystic mass. Two types are recognized. Intralobar sequestration, in which the sequestrated lung lies within normal pulmonary visceral pleura, and extralobar sequestration in which abnormal lung tissue is completely separate and enclosed in its own pleural covering. In intralobar disease the venous drainage is usually through pulmonary vein, and in extralobar disease it drains through pulmonary vein or a systemic vein.10 In this case bronchocele was supplied by a branch of pulmonary artery and was drained into left brachiocephalic vein. There was no systemic arterial supply. In this way it was also differentiated from pulmonary sequestration.

**REFERENCES**