Asymptomatic congenital Bronchial Pulmonary Arterial vascular malformation in an Adult.

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Consent:
Written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy.

Abstract:
This case report describes a 41-year-old male patient whose screening chest X-ray revealed irregular tubular radio dense opacities in the right hilar region, suggestive of a vascular lesion. Subsequently, CT angiogram of the chest revealed a congenital bronchial artery-pulmonary artery fistula. Despite being informed about the diagnosis and available treatment options, the patient refused treatment and wanted to be on regular follow-up. This case highlights the importance of thorough imaging evaluation in diagnosing rare vascular abnormalities and explores the existing gap in literates regarding treatment in asymptomatic patients.

Key words
Bronchial-pulmonary artery, Arteriovenous malformation, CT angiography, Asymptomatic.

Introduction:
Abnormal communication between a systemic artery and pulmonary artery is a rare anomaly which may be congenital or acquired. The acquired causes of such malformations are inflammatory or infective lung disease, penetrating trauma, or tumor (2). Here we describe a primary anomalous communication between a dilated convoluted bronchial artery and the branch of the pulmonary artery incidentally found on a routine health checkup radiograph. Such communications have been given various names in the literature, one commonly used name being the racemose hemangioma of the bronchial artery (7).

Case Report.
A 41-year-old male patient came for a screening chest X-ray. He had no complaints. No history of fever, vomiting, and cough. No previous medical and surgical history was noted. Physical and clinical examination revealed no gross abnormalities. Chest X-ray PA view (Figure 1) demonstrated irregular tubular radiodense opacities on the right hilar region. Both lung fields were normal with normal broncho-vascular markings. A diagnosis of some form of vascular lesion was made and the patient was advised a CT angiogram chest.

**Figure 1:** Chest X-ray PA view shows an irregular tubular radiodense lesion on the right hilar region (arrow).

CT angiogram of the chest demonstrated a cluster of dilated and tortuous vessels (Figures 2 and 3) in the right paraspinal region, 5th and 6th intercostal areas, and the superior segment of the right lower lobe with 3 feeding vessels arising from the aorta (Figure 4) and draining into the superior segmental branch of the right descending pulmonary artery (Figures 5 and 6). No abnormality of the lung was noted. A diagnosis of congenital bronchial artery-pulmonary artery fistula was made.

Differential diagnosis for the case was pulmonary arteriovenous malformation, vascular malformations, coarctation of the aorta with collaterals however, CT angiogram clearly demonstrated normal aorta and the presence of feeding vessels from the aorta draining into the superior segmental branch of the right descending pulmonary artery which ruled out these differentials. Patient was explained about the diagnosis and treatment options. He refused treatment and opted to be on regular follow-up.
Discussion:

Bronchopulmonary arterial malformation (BPAM) is a rare congenital anomaly that can present with a wide range of respiratory symptoms. The exact incidence of congenital BPAM has not been reported in literature. Some cases of acquired BPAM have also been reported in the literature with causes ranging from infection to tuberculosis, pneumonia, actinomycoses, tumor, and trauma (9). It is typically diagnosed in childhood or adolescence, but an asymptomatic case presenting in adulthood, like the one discussed here, is extremely rare. There were no lung findings or history in our patient to suggest an acquired cause. In this case, we discuss the diagnosis and pathophysiology of BPAM and the dilemma in the treatment of an asymptomatic patient.

Bronchial-pulmonary arterial malformation (BPAM) or racemose hemangioma of the bronchial artery is a rare congenital malformation of the bronchial artery which is characterized by a dilated and convoluted bronchial artery with vascular hyperplasia and anastomosis with adjacent systemic vessels and fistulous communication with pulmonary artery (7). Such a convoluted bronchial artery with pulmonary anastomosis has been described in the literature since the late 1970s and termed angiomatoid vascular convolution of the bronchial artery, racemose angioma of the bronchial artery, bronchial angiomas (6,8). Since then, case
reports of malformation between systemic to pulmonary arteries has been reported in the literature by various names like bronchial arteriovenous malformation, bronchopulmonary arterial anastomosis, bronchopulmonary shunt, bronchial artery-pulmonary artery malformation and fistula, and racemose hemangioma of bronchial artery (1, 2).

The lungs have two separate vascular systems consisting of the pulmonary and bronchial arteries. Pulmonary artery arises from the right heart and is a low-pressure system whereas bronchial artery arises from the aorta (systemic circulation) and has approximately 6 times higher pressure than the pulmonary circulation. Pulmonary arteries participate in gas exchange at the alveolar level, while bronchial arteries provide nourishment to the supporting structures of the lungs, including the pulmonary arteries (2). Bronchial arteries are connected to the pulmonary arteries through several microvascular anastomoses at the level of the alveoli and respiratory bronchioles. This preexisting right to left shunt is nonfunctional in the physiological state, however significant flow can be noted in the disease state like pulmonary embolism. This is one of the anatomical basis of bronchial to pulmonary arterial shunt. (1).

Most of the patients with systemic to pulmonary AV malformations are asymptomatic. Case reports of life-threatening hemoptysis due to the condition have however been described. The condition is more commonly seen in young male patients (3,4). Chest radiographs are often abnormal, but lesions are usually nonspecific and minimal. Definitive diagnosis is possible by CT angiography or conventional angiography.

Broncho-pulmonary arterial malformation is an extremely rare vascular malformation, so treatment guidelines are less established. Treatment of life-threatening hemoptysis have been reported with modalities like lobectomy, trans-arterial embolization. However, the natural course of the disease and the need for treatment in an asymptomatic patient is not well established (5). Our patient was counseled about the incidentally identified abnormality and given the option of treatment by embolization and follow-up. He opted to be on routine clinical and radiological follow-up.

Conclusion:

Broncho-pulmonary arterial vascular malformation is a rare entity and has been reported in the literature with various names. The cases are usually asymptomatic; however, they have the potential for life-threatening hemoptysis.

Conflict of interest:

None

References:

