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Case Report

RIGHT SIDED AORTIC ARCH CAUSING CHRONIC OBSTRUCTIVE AIRWAY DISEASE AND COR PULMONALE

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INTRODUCTION

A large number of vascular malformations can exist in the superior mediastinum, but all of them are not of clinical importance. They gain significance when they disturb the surrounding anatomy of mediastinum due to their abnormal anatomical characteristics. Such abnormal or displaced arterial malformations can lead to the compression of trachea or esophagus leading to obstruction of these vital passages. Various vascular anomalies that lead to respiratory distress and need interventions include double aortic arch, right aortic arch associated with left ligamentum arteriosum or patent ductus arteriosus, aberrant subclavian artery, aberrant left pulmonary artery, abnormally placed innominate artery, anomalous left common carotid artery. Right sided aortic arch is found in approximately 0.1% of population (Ahmadi and Sonbolestan, 2013). It is mostly asymptomatic, but when symptomatic, it generally presents with exertional dyspnea and chronic cough. It may be more common in the community than estimated (Ozkaya *et al.*, 2012). We are presenting an elderly male, being managed as a case of COPD, after thorough investigations, was revealed to have right sided aortic arch causing respiratory distress by causing compressive symptoms.

CASES

Clinical Presentation

A 57- years- old male, HIV negative, carpenter, ex-smoker presented to our institute with progressive exertional dyspnea for 12 years associated with orthopnea and swelling of feet and intermittent productive cough. There was no history of wheezing, fever, chest pain, repeated colds, haemoptysis, loss of appetite, loss of weight or any other complaint. He was prescribed inhaled steroids and bronchodilators with chronic obstructive pulmonary disease (COPD) and cor pulmonale as the diagnosis in the subsequent years. Subsequently he developed hypertension and was started on antihypertensives. 4 years later he was admitted to intensive care unit (ICU) for NIMV (Non Invasive Mechanical Ventiliation) support. Subsequently he was also diagnosed to have diabetes mellitus. He has since required frequent hospital admissions for intermittent NIMV support. General physical examination at presentation revealed blood pressure of 124/70 mm Hg, respiratory rate of 22 per minute, saturation at room air was 94%. There was no pallor, icterus, cyanosis or clubbing. On auscultation, bilateral vesicular breath sounds with bilateral rhonchi were audible.

Investigations

Complete blood count, urine analysis and renal and hepatic functions were within normal limits. ECG showed p-pulmonale suggestive of cor pulmonale. Pulmonary function testing revealed severe airflow obstruction with evidence of intrathoracic tracheal compression. Sputum examination for acid fast bacilli, other aerobic organisms and fungi was negative. Echocardiography was within normal limits. HbA1C was 8.3% suggestive of poorly controlled diabetes. Chest radiograph showed hyperinflation with widening of right side of mediastinum. Contrast enhanced computed tomography of chest revealed hyperinflation of bilateral lung fields. CT angiography showed right sided aortic arch compressing the trachea along with normal placed descending aorta. On fiberoptic bronchoscopy, large bulge with pulsations was seen in posterior part of trachea about 5 cm above carina causing tracheal narrowing.

DISCUSSION

Congenital vascular anomalies causing significant tracheobronchial compression are uncommon. Most of the anomalies are due to faulty development of six primitive aortic arches. Right sided aortic arch is found

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in approximately 0.1% of population and aberrant left subclavian artery is the most common anomaly associated with it (Ahmadi and Sonbolestan, 2013). Other anomalies associated with right sided aortic arch include left ligamentum arteriosum, patent ductus arteriosus, ventricular septal defect. It is very rare to have right sided aortic arch without any associated congenital heart anomalies as is present in our case (McElhinney *et al.*, 2001; Mart *et al.*, 2001). During the anomalous development of right sided aortic arch, the first part of ascending aorta is in normal position, but it takes the anomalous position by ascending to the right of trachea and esophagus as compared to normal anatomical development where it is directed upwards and to the left of trachea and then continuing as descending aorta (Gross *et al.*, 1955). Right sided aortic arch alone can be asymptomatic or it can compress the bronchus or trachea leading to respiratory difficulties or when accompanied with ligamentum arteriosum or patent ductus arteriosus, it can form the constriction ring encompassing trachea and esophagus causing symptoms. Patients with right sided aortic arch are usually asymptomatic.

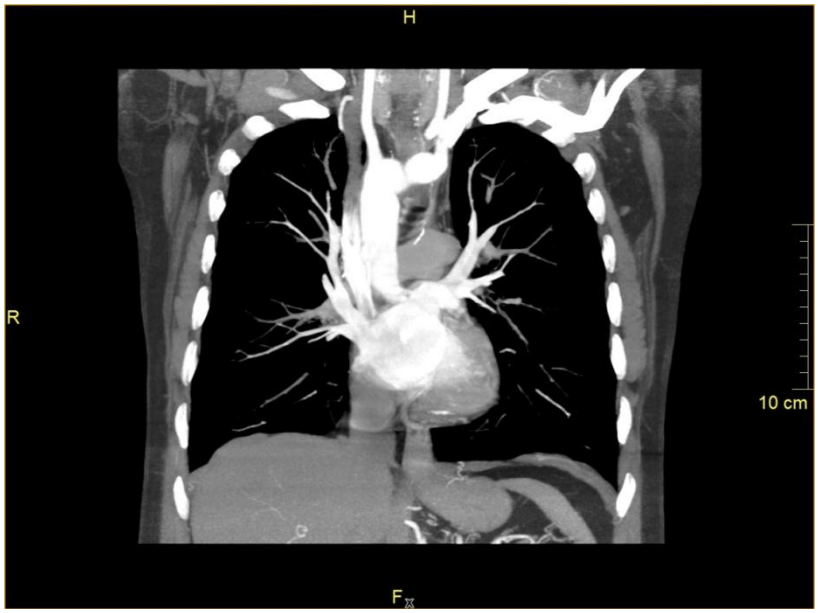

Figure 1: Chest X ray PA View showing bilateral hyperinflation with widened right mediastinum

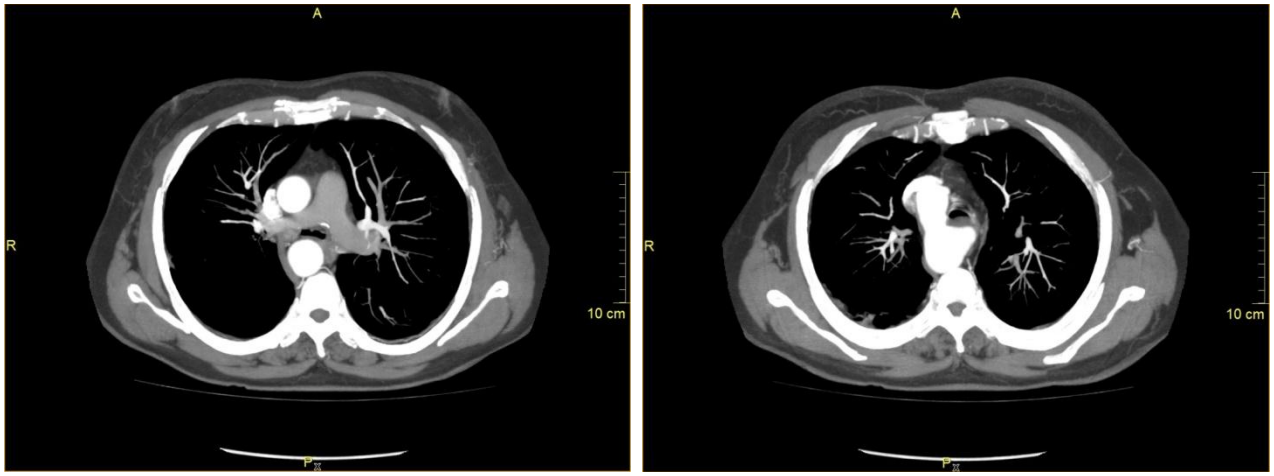

Figure 2: CT scan axial images showing right sided aortic arch (Figure 2a) with normally pleced ascending and descending aorta (Figure 2b)

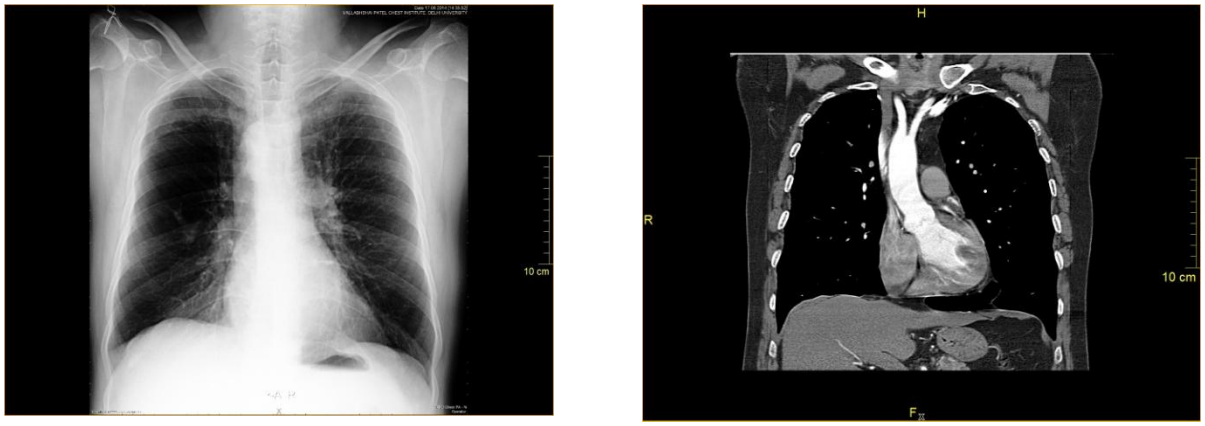

Figure 3: CT angiogram showing right sided aortic arch (Figure 3a) with normally placed pulmonary and systemic vasculature (Figure 3b)

In our study, the chief complains of the subject are progressive exertional dyspnea and intermittent productive cough. In one of the recent study (Ozkaya *et al.*, 2012) clinical, radiological and spirometric features were evaluated in 13 patients with ages range of 17 to 86 years and males to females ratio of 11:2, over a period of 4 years, and it was found that 7 patients (54%) were symptomatic due to tracheal compression by right sided aortic arch and it was also found that hypertension was found in 2 cases which increased the exertional dyspnoea, 5 patients showed intrathoracic tracheal obstruction on pulmonary function test, as is the scenario in our case; and it was suggested that right sided arcus aorta to be included in differential diagnosis in patients presenting with exertional dyspnoea and chronic cough. In our study, chest radiograph, contrast enhanced computed tomography, CT angiography, pulmonary function test, fiberoptic bronchoscopy and echocardiography helped us to reach the diagnosis. Other tests which could be useful include magnetic resonance imaging and barium swallow to detect esophageal compression. In one study (Vučurević *et al.*, 2013) on anatomy and radiology of variations of aortic arch branches in 1266 patients, 946(74.72%) showed normal vascular patterns whereas 320(25.28%) presented with variations of the supraaortic vessels and the aortic arch and out of those right sided aortic arch was present in 4 patients(0.32%). Patients who are diagnosed as cases of asthma, chronic obstructive pulmonary disease and who are unresponsive to conventional treatment should be thoroughly investigated to search for the cause of persistent breathlessness. In one case series (Payne *et al.*, 2000), three children, two of them were diagnosed cases of asthma and one was interstitial lung disease, were evaluated for persistent breathlessness, and were found to have right sided aortic arch as the cause of their non resolving symptoms inspite of the best medications.

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