

47 Year Old Patient with Pseudo-Asthma

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Received: 20 Oct 2010 / Accepted: 26 Dec 2010
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A 47- year- old female patient presented in out-patient department in Afyon Chest Disease Hospital with dyspnea, cough and purulent sputum. She has been receiving regular inhaler β -agonist and corticosteroids with the diagnosis of asthma bronchial since childhood. However, the patient had a history of very often upper and lower respiratory tract infections. The patient did not smoke, drink alcohol, or use illicit drugs intravenously. She was currently on asthma medications.

On physical examination, the patient has central cyanosis, pretibial edema and her lung sounds were decreased to auscultation bilaterally with deep inspiration. In cardiac auscultation, the heart sounds were heard at the right of sternum and there was splitting in second heart sound (S^2). ECG demonstrated sinus tachycardia, inversion of the P and T wave in lead 1 with loss of R wave progression and p pulmonale.

In routine complete blood test, the patient had a mild leukocytosis 12000 / μ L with % 81 neutrophil, C- reactive protein level was 20 mg/dl, erythrocyte sedimentation rate was 56 mm/h. Other parameters were in normal ranges.

Question

What is your diagnosis from the figures?

- Cystic fibrosis
- Allergic bronchopulmonary aspergillosis
- Young syndrome
- Kartagener syndrome
- Churg-strauss syndrome

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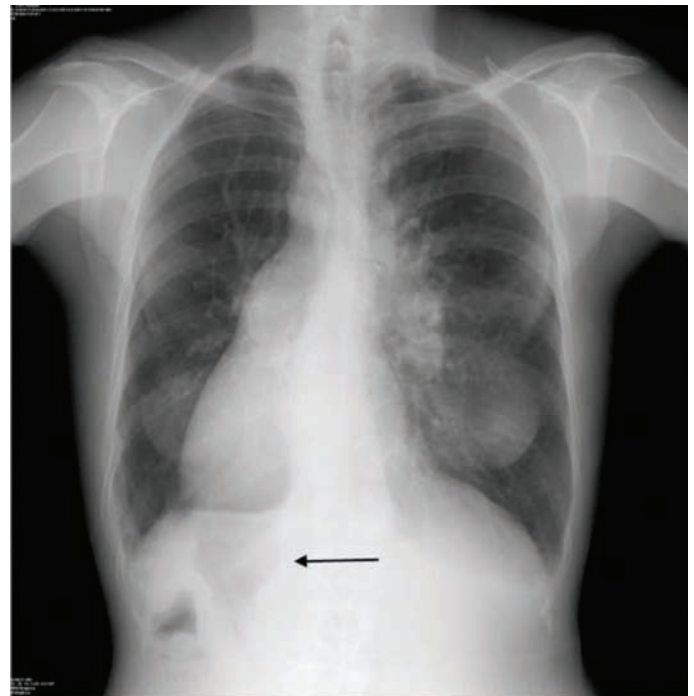


Figure 1: Chest x-ray revealed dextrocardia and a right-sided stomach bubble (white arrow).

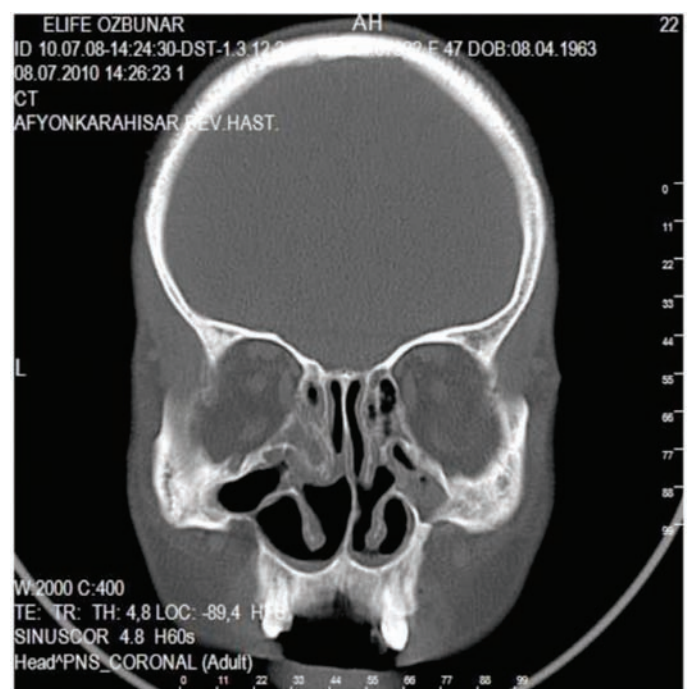


Figure 2: CT of the paranasal sinuses showing the sinusitis.

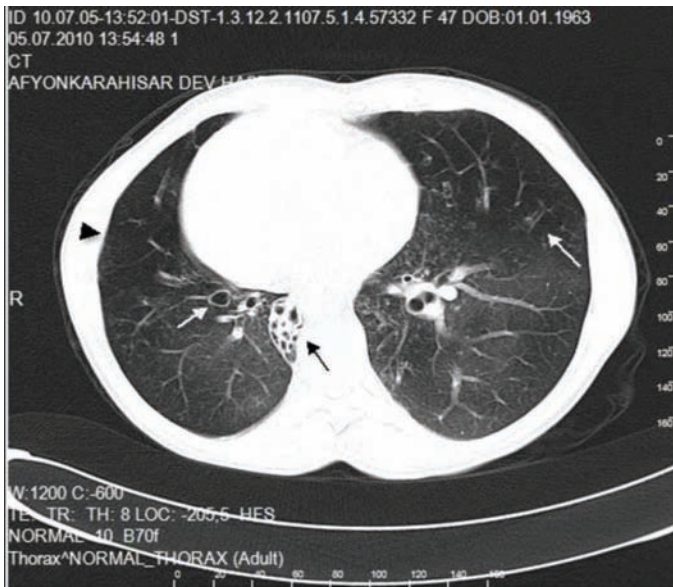


Figure 3: Chest Ct demonstrated; right middle lobe atelectasis (black arrow), tubular (long white arrow) and cylindrical (short white arrow) bronchiectasis, air trapping (arrowhead) and bilateral tree in bud and ground glass attenuation.

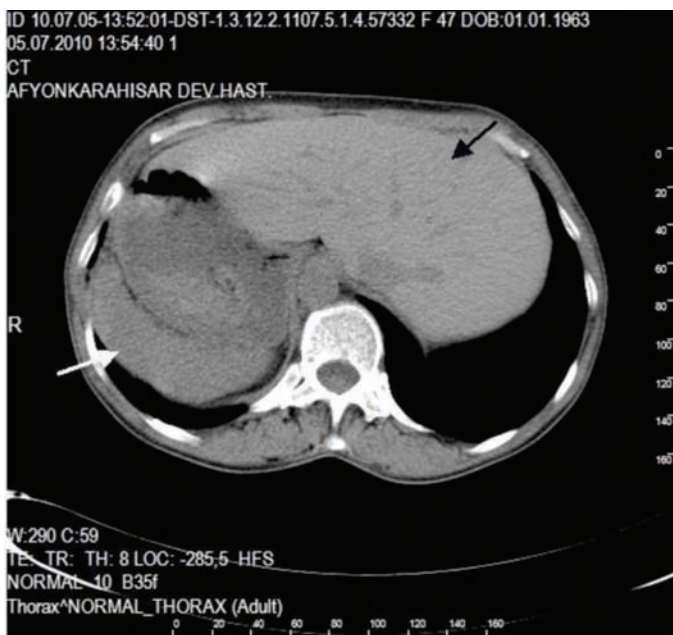


Figure 4: Spleen is right sided (white arrow) whereas liver is left sided (black arrow) in lower Chest CT cross-section.

Answer

Diagnosis: d. Kartagener syndrome

Chest x-ray revealed dextrocardia and right-sided stomach bubble. (Fig. 1). In pulmonary function test there was severe obstructive pattern with no reversibility [FEV1 0.71 liters (30%) and FEV1/FVC=49%]. In arterial blood gas analysis, the patient has hypoxemic and normocapnic respiratory deficiency with PaO₂: 55mmHG, PCO₂: 46mmHg, SaO₂: 88%. Paranasal sinus CT showed bilateral maxillary sinusitis (Fig. 2). Thorax computed tomography (CT) revealed, dextrocardia, right sided

lung volume loss, right middle lobe atelectasis, tubular and saccular bronchiectasis in bilateral lower lobes, ground glass attenuations in both lung with posterior preponderance, multifocal tree-in bud pattern and air trapping areas in bilateral lung fields. Spleen was at right sided whereas liver was at left sided relevant with situs inversus in lower cross-sections of thorax CT. (Fig. 3 and Fig. 4). Pneumococcus was isolated from sputum culture. While pulmonary artery pressure was found 40 mmHg no cardiac anomaly was found in echocardiographic investigation. Eustachian Tube dysfunction was determined in right ear during ENT consultation.

Total immunoglobulin levels and class analysis was normal. PR3-ANCA, MPO-ANCA and sweat testing were negative. Bronchodilator and antibiotherapy was done, since she had obstructive lung disease and pulmonary infection. Saccharine test time was found 1 hour 9 minute (<30 minute normal) after medical therapy. The patient was diagnosed as Kartagener syndrome with classical triad of bronchiectasis, sinusitis, situs inversus and with mucociliary dysfunction.

Discussion

Kartagener syndrome is part of the larger group of disorders called primary ciliary dyskinesias. Approximately one half of patients with primary ciliary dyskinesia have situs inversus and, thus, are classified as having Kartagener syndrome.¹ This syndrome is characterized by the clinical triad of chronic sinusitis, bronchiectasis, and situs inversus. The condition can be diagnosed as clinical and radiological and it can corroborate with abnormal mucociliary dysfunction. A more definitive diagnosis of primary ciliary dyskinesia is made by light and electron microscopy of intact nasal or bronchial cilia. Symptoms result from defective cilia motility. The main consequence of impaired ciliary function is reduced or absent mucus clearance from the lungs and susceptibility to chronic recurrent respiratory infections, including sinusitis, bronchitis, pneumonia and otitis media. Progressive damage to the respiratory system cause bronchiectasis which has more predilection for middle and lower lobes in comparison with cystic fibrosis. As probably results from elevated levels of local inflammatory mediators in a chronically irritated airways. In these patients obstructive lung disease progress in course of time which results in some complications such as secondary pulmonary hypertension and respiratory failure.

Despite being an inherited defect, it is sometimes only diagnosed late in life.² Patients were generally misdiagnosed with asthma bronchiale.³ Our patient was diagnosed as asthma bronchiale until 47 year old. Early diagnosis and management of chest infections can prevent irreversible damage to lungs and prevent potential lifelong complications.

Middle lobe syndrome (MLS) is an uncommon lung disorder, usually refers to any cause of recurrent or persistent atelectasis of the right middle lobe. Originally thought to be due only to bronchial obstruction, but it is more often present with non-

obstructive lesions. The right middle lobe is more often involved because of its isolation and poor collateral flow from the upper and lower lobes.⁴ We believe in that chronic inflammation and bronchiectasis led to MLS in our patient.

Conclusion

Hereditary diseases is not appear only in childhood. The differential diagnosis of asthma is important. The many other causes of wheezing, dsypnea and cough which must be kept in mind.

Acknowledgements

The authors reported no conflict of interest and no funding has been received on this work.

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