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Scimitar syndrome - a late onset associated with asthma clinic

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Background: Scimitar syndrome is a rare congenital anomalous pulmonary venous drainage of the right or left lung into vena cava inferior.Cases diagnosed in early childhood have significant clinics of pulmonary findings but late onset group may have subtle clinics and also be diagnosed accidently.
Method/Result: A thirteen years old girl was brought to the hospital with the complaints of recurring respiratory distress, grunting and cough which was started in her babyhood and continued periodically.She had lower respiratory infections after when she was 7 months old and then repeated every 2-3 months.She had these infections every month last 3 years.During this period, she had the diagnoses of bronchitis, bronchiolitis, asthma and pneumonia and was given antibiotics and bronchodilator therapies for many times.When she was 3 years old, adenoidectomy was performed because of the complaints of grunting and respiratory distress.Lung sounds were equal but coarsening and expiratory sounds were prolonged in her physical findings.Her pulmonary function tests are low, FEV1 was 72%,FVC was 69%,FEV1/ FVC was 100%.in PA radiographics there was an atypical view of asymmetrical fullness in the left hilar area.There was a huge venous structure at the base of the right lung that drained into vena cava inferior and in the neighborhood there were foci of focal emphysema in thorax computed tomography.in three dimensional thorax angiographic CT there was an aberrant pulmonary vein at the base of right lung and the largest dimension of pulmonary vein through costodiaphragmatic recess was 11 mm and it was draining into vena cava inferior adjacent to liver.This view was compatible with scimitar syndrome.Drainage of 3 of the pulmonary veins into left atrium and drainage of aberrant pulmonary vein into hepatic vein were observed in her echocardiography.
Conclusion: Scimitar syndrome should be evaluated in children with recurrent pulmonary infections, asthma clinic and respiratory distress.