DOI: 10.5152/TurkThoracJ.2019.373

[Abstract:0584] PP-289 [Accepted:Poster Presentation] [Pediatric Lung Diseases]

Hepatopulmonary Syndrome and Multiple Arteriovenous Fistulas in a Child with Niemann Pick Disease

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Introduction: Niemann Pick Disease (NPD) is a rare autosomal recessive hereditary disease and it causes abnormal storage of sphingomyelin in the tissues of reticuloendothelial system. When sphingomyelin storage affects the pulmonary system; recurrent pneumonia, dyspnea, clubbing can occur in the patients. Hepatopulmonary syndrome (HPS) and pulmonary arteriovenous fistulas (AVF) may occur in patients with NPD. In this report a 16-year-old girl who had NPD type B with HPS and multiple AVFs and coil embolization treatment was presented.

Case Presentation: A 16-year-old girl with NPD admitted to our clinic for further examination and treatment. She had cyanosis and dyspnea and in her physical examination; massive splenomegaly, mild hepatomegaly and severe digital clubbing were determined. Crepitant rale and bronchial respiratory sounds were heard in her respiratory sounds. Her oxygen saturation was 84% on room air. She was given oxygen at a rate of 10 liter per minute in order to maintain oxygen saturation at 94%. In the chest X-ray, increased reticulonodular opacity was observed. Thorax computer tomography (CT) showed there were multiple AVFs in the medial segment of the right middle lobe and laterobasal segment of the left lower lobe which were related to pulmonary arteries. There were both pleural thickening and bilaterally interlobular septal thickening and chronic empyema in the left lung. Due to her severe hypoxemia saline contrast enhanced echocardiography was performed and she was observed to have a normal ejection fraction with air bubbles after right atrial opacification consistent with intrapulmonary shunting. She was diagnosed with hepatopulmonary syndrome (HPS). Because of her multiple AVFs in contrast thorax CT she was diagnosed with type 2 HPS. Liver transplantation was contraindicated due to HPS type 2. So as to decrease her oxygen dependence pulmonary angiography and embolotheraphy were performed. After her ten AVFs were embolized her oxygen dependency reduced day by day. On her 6th day after the intervention her oxygen supplement was terminated. She was discharged from hospital without any oxygen supplement. After the embolization she was able to demonstrate communication and fulfill certain physical functions.

Conclusion: Patients with NPD should be examined for pulmonary involvement. HPS or/and AVFs may occur in those patients and coil embolotheraphy can be discussed for increasing oxygenization.

Keywords: Arteriovenous fistula, child, hepatopulmonary syndrome, Niemann-Pick disease