JCK Oral

JCK Oral 8 (III-JCKO8)
Cardiovascular Imaging

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11:05 AM - 11:55 AM

[III-JCKO8-01]Morphological and clinical spectrum of infantile scimitar syndrome

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Objective Scimitar syndrome is a rare constellation of cardio-pulmonary anomalies. We describe the morphological and clinical spectrum in a retrospective case series of infantile scimitar syndrome. **Methods** A total of 7 patients with infantile scimitar syndrome were identified in our hospital from 2009 to 2016. **Results** The mean age at diagnosis was 4.7±2.6 months with mean body weight of 5.2±1.0kg. All patients presented with symptoms, including respiratory distress (n=5), heart failure (n=4), recurrent pneumonia (n=3) and cyanosis (n=2). There were 3 patients associated with ASD, 2 with TOF and 1 with ASD and PDA. Anomalous right pulmonary scimitar vein provided drainage for the entire right lung in 5 patients, and only the lower segments in 2 patients, including drainage site to superior vena cava in 1 patient, to the hepatic vein with obstruction in 1 and to inferior vena cava in 5. All the patients underwent cardiac catheterization. Severe and moderate pulmonary arterial hypertension was demonstrated in 2 and 3 patients, respectively. Transcatheter collateral embolization was performed in 4 patients. Surgical repair of intra-cardiac lesion with and without anomalous vein correction was carried out in 2 and 3 patients, respectively. Mean follow-up time was 12.5±10.5 months. There were 2 hospital deaths and no late death. **Conclusions** The infantile form of scimitar syndrome is a very severe form of disease, usually associated with heterogeneous morphological variations. A careful anatomic study is mandatory for prompt treatment.