Clinical Case: Right pulmonary artery agenesis

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Background: Among arterio-venous malformations, unilateral agenesis of a pulmonary artery (UPAA) is definitely considered one of the rarest, with few cases described in the literature, and its number decreasing in pediatric cases. First described by Fraentzel at 1868, it is frequently associated with cardiac or genetic defects, and those not related to these associations are extremely rare. The case of a female infant with a history of wet cough and episodes of hemoptysis is presented, which leads to performing a contrast-enhanced HRCT (high - resolution computed tomography) and diagnosing agenesis of the pulmonary artery.

Description: 1-year old female infant, mother´s first pregnancy, 40 weeks gestational age, c- section delivery due to prolonged labor, APGAR score 8-9, birth weight 7.7 pounds, birth height 19.6 inches, no abnormalities detected. Medical record: 1-week hospitalization at 3 months of life due to bronchiolitis, discharged with inhaled steroid and long action beta agonist. After that, occasional wet cough was referred and 5 months later comes up to E.R. suffering wet cough and hemoptysis and coffee ground vomitus. Therefore, endoscopy and bronchoscopy were performed, with no relevant findings. After that, the patient presents hematemesis, so that contrast tomography of the thorax was performed, showing contrast medium into the left upper lobe, which makes the diagnosis of a suggestive pulmonary arterio-venous malformation, likewise the hypoplasia / agenesis of the right pulmonary artery is also observed. A right lateral thoracotomy and pneumonectomy was performed, no right pulmonary artery was identified.

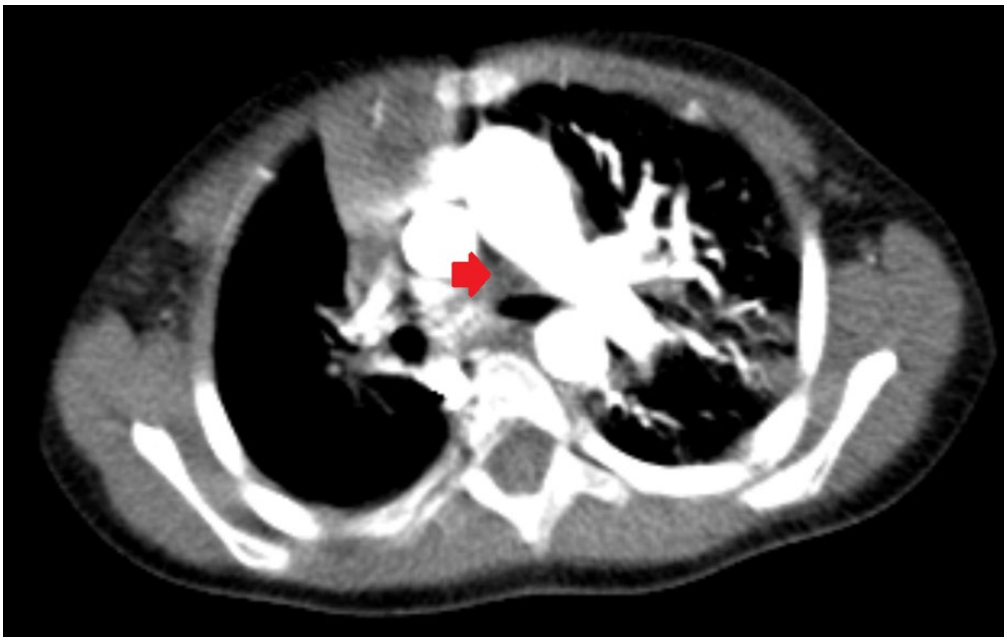

Figure 1.- Axial contrast - enhanced CT, demonstrates agenesis of the right pulmonary artery. (red arrow)