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Pulmonary capillary haemangiomatosis: an unusual histological presentation of vascular lesion in lung biopsies from patients with congenital cardiac shunts and pulmonary hypertension

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Introduction: The typical pulmonary arterial occlusive lesions occurring in patients with congenital cardiac shunts are characterized by medial layer hypertrophy and intimal proliferation. On the other hand, pulmonary microvasculopathy (former capillary hemangiomatosis) is a rare form of histological lesion of pulmonary hypertension (PH), usually described in the idiopathic or “primary” form of the disease, being sometimes associated to veno-occlusive lesions but not described in other etiologies. We report two cases of microvasculopathy in a large series of lung biopsies obtained from patients with congenital cardiac shunts.
Patients and methods: Among 297 lung biopsies analyzed over an 11year period, two from children with PH were found to have microvasculopathy. First patient is a 4year-old girl with Down’s syndrome, an 8mm secundum atrial septal defect and stenosis of the right pulmonary artery. The other is a 2year-old girl with a 13mm perimembranous ventricular septal defect. The mean pulmonary arterial pressure was above 30mmHg in both cases. No signs of left ventricular obstructive lesions were observed on echocardiography. Lung biopsies were performed to evaluate the severity of the arterial lesions and collected inflated in order to maintain the overall lung architecture. The first patient underwent closure of the atrial defect and enlargement of the right pulmonary branch and the second undewent pulmonary artery banding.
Results: Both biopsies showed severe hypertrophy of the walls of pre and intra-acinar arteries. Intimal proliferative lesions were present in pre and intra-acinar arteries from the first case and only in pre-acinar arteries from the second one (Heath-Edwards grade II). Both cases showed thin-walled capillary vessels infiltrating the perivascular and peribronchial connective tissue and enlarging focally the alveolar walls. Hemossiderin laden macrophages were found in alveolar lumens. Such histological features are typical of pulmonary microvasculopathy. The cases reported here correspond to 1.5% of all lung biopsies in the series.
Conclusion: To the best of our knowledge, the occurrence of pulmonary microvasculopathy in association to the classical histological occlusive arterial lesions in congenital cardiac shunts has not been described. Its possible role in increasing the severity of pulmonary hypertension in this context and its pathogenesis need to be elucidated.