

Pulmonary Capillary Hemangiomas (PCH) (A Rare Cause of Pulmonary Hypertension)

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History and presenting complaints



A 26-year-old nonsmoking male presented with progressive exertional shortness of breath for the past 2 months that significantly worsened over the last 2 weeks

requiring 2 to 4 L of oxygen, depending on his level of exertion.

No history of fever, cough, sputum, or hemoptysis.

Physical examination:

Mild central cyanosis with mottled skin lesions, tachycardia, accentuated P2, mild ascites, and edema of the lower extremities, Otherwise unremarkable.

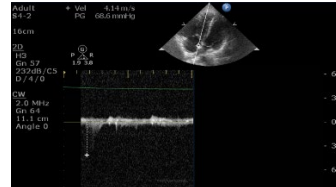
ECG: RAD, RVH + ST- T changes.

CXR: showed a slightly enlarged cardiac silhouette with dilatation of the main and central pulmonary arteries and abrupt peripheral tapering. There were also mild, nonspecific interstitial markings. Pulmonary hypertension was suspected.

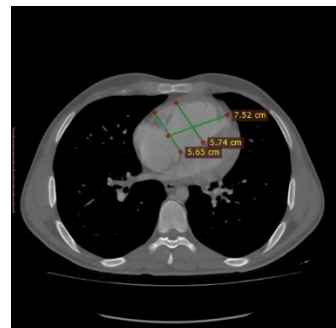
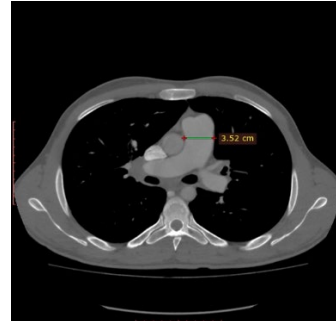
So, Pulmonary function test in the form of spirometry reflected mild fixed obstruction, borderline restriction.

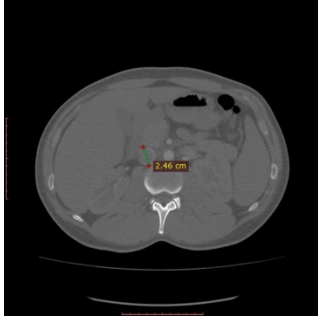


TTE:



MSCT Chest:





Discussion:

First recognized and described by Wagenvoort et al in 1978, PCH is a vascular proliferation occurring with a frequency of 4 cases per million individuals. It affects young adults aged 20 to 40 years (mean age, 29 years), although cases have been previously reported in adolescents, children, premature infants, and even in newborns.

Pulmonary capillary hemangiomatosis occurs in a sporadic fashion, although familial patterns have been described. It manifests as pulmonary hypertension with an indolent onset of symptoms and signs slowly progressing to cor pulmonale with normal pulmonary wedge pressures.

A clinical presentation of postcapillary pulmonary hypertension manifesting as dyspnea, cough, and hemoptysis with fatigue and weight loss is often described. Fever,

respiratory tract infection, digital clubbing, thrombocytopenia (especially in the pediatric age group), and hemorrhagic complications, including secondary hemothorax, may occur.

Conclusion:

The differentiation of rare causes of pulmonary hypertension (such as PCH and PVOD) from isolated pulmonary arterial hypertension is of clinical relevance because of the problem of severe, life-threatening, vasodilator-induced pulmonary edema. Differentiation is difficult on clinical grounds alone.

Imaging studies suggest that high-resolution CT findings of centrilobular nodules, GGA, and septal thickening should raise suspicion for PCH or PVOD and provoke further investigations and, possibly, open lung biopsy.

The cause of PCH remains unknown.

The natural history of the disease is not well defined.

It is a very rare disease, and generally poor prognosis.

Most recently, evidence of increased expression of vascular endothelial growth factor and platelet-derived growth factor activity in patient with PCH has been reported.

Lung transplantation is the only curative therapeutic option available.