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Case Report

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Polycythernia Vera induced Chronic Thromboembolic Pulmonary Hypertension

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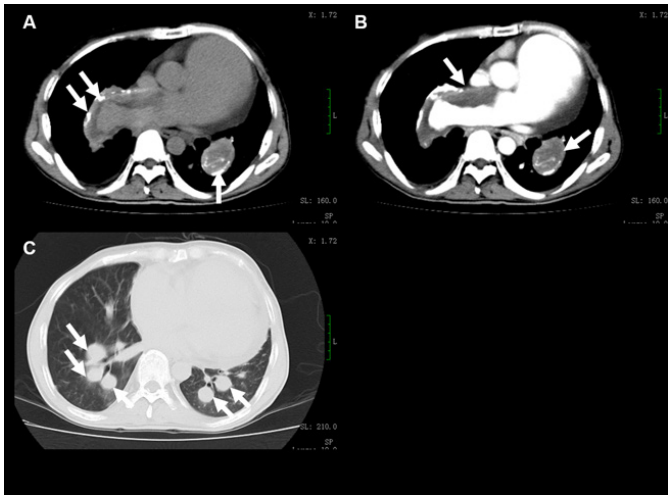
Case Report

A 47-year-old man presented with progressive dyspnea over a period of 4 years. On physical examination, central cyanosis and nail clubbing were present. Cardiac examination revealed left parasternal heave and P2 > A2 on auscultation. The hemoglobin level was 22.0g per liter. Arterial blood gas analysis showed a pH of 7.42, a PaO_2 of 44 mmHg, and a PaCO2 of 38 mmHg when he was breathing ambient air. A transthoracic echocardiogram exhibited right ventricular enlargement, severe tricuspid regurgitation and an estimated pulmonary-artery pressure of 112 mmHg. Plain (Figure1A) and contrast-enhanced (Figure1B) CT scan of the chest showed eccentric thrombotic material within the pulmonary arteries, with scattered calcification in the thrombi (arrow). The pulmonary-to-aorta diameter ratio was over 3. Enlargement of pulmonary arteries were also present (Figure1C, arrow). Bone marrow study showed erythroid hyperplasia and a mutation of JAK2V617F (mutation: wild-type 30.2). The diagnose of polycythernia vera was established. Hydroxyurea and warfarin were prescribed. Six months after discharge, his physical status worsened gradually but he refused to receive surgery.



Discussion

Chronic thromboembolic pulmonary hypertension (CTPH) is one of the most common causes of pulmonary hypertension (PH), according to the recent clinical classification of PH [1]. Myeloproliferative diseases, especially polycythaemia vera and essential thrombocytosis, have been reported to be related to the development of CTPH. Pulmonary endarterectomy could be the treatment of choice in selected patients, however, the prognosis remains poor [2].

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