Silent Shunt, Loud Consequences: Chronic Thromboembolic Pulmonary Hypertension

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ABSTRACT

Chronic thromboembolic pulmonary hypertension (CTEPH) is a type 4 pulmonary hypertension (PH) that results from chronic or repeated episodes of pulmonary emboli, leading to the formation of fibrous lesions from residual emboli. There is a potential risk of developing CTEPH after ventriculoatrial shunt (VAS) surgery in patients with hydrocephalus, even in the absence of typical disease symptoms. Herein, we present a rare case of a 31-year woman who developed CTEPH after undergoing VAS surgery. This report examines how CTEPH can mimic the symptoms of other diseases, emphasising the importance of physicians considering this condition during diagnosis. Early detection of CTEPH is crucial for preventing cardiac deterioration.

Key Words: *Ventriculoatrial shunt*, *Chronic thromboembolic pulmonary hypertension*, *Hydrocephalus*, *Ventriculoatrial shunt thrombosis*.

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INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH) is a type of pulmonary hypertension (PH) in which most cases develop through abnormal resorption of acute pulmonary embolism. CTEPH incidence varies from 3 to 30 cases per million populations. However, it can be stated that the precise incidence of CTEPH is often underestimated.[1,2] Ventriculoatrial shunt (VAS) is frequently employed in the management of all forms of hydrocephalus, including idiopathic normal-pressure hydrocephalus and secondary post-traumatic, post-haemorrhagic, and post-infectious hydrocephalus.[3] We present a rare case of a 31-year female who developed CTEPH after VAS. As far as we know, this is the second such study of CTEPH related to VAS since the year 2000.

CASE REPORT

A 31-year female patient consulted the clinic in January 2024 with complaints of exertional dyspnoea (NYHA Class III to IV) for 3-4 months, generalised weakness, and bilateral pedal oedema for four months. Upon reviewing the patient's medical and surgical history, it was discovered that she had a ventriculoperitoneal (VP) shunt placed in 2017 to address hydrocephalus secondary to tuberculous meningitis.

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Due to dysfunction, this VP shunt was subsequently replaced with a right VAS. Despite the intervention, her symptoms recurred in August 2020, requiring the revision of the lower end of the shunt.

Upon examination, the patient was afebrile with a SpO2 of 96%, with a pulse rate of 98 beats/min and a blood pressure (BP) of 110/70 mmHg without postural drop. However, she had pedal oedema to her ankles, and her jugular venous pressure (JVP) was elevated. Cardiac examination revealed a loud S2 and a grade IV/VI pan-systolic murmur over the left sternal border. Electrocardiography (ECG) revealed sinus rhythm at a heart rate of 80 beats/min, normal axis, and a prominent R wave in leads V1-V3 with ST-T wave changes. Echocardiography showed severe tricuspid regurgitation, a dilated right atrium and ventricle with reduced right ventricular function, and raised pulmonary artery systolic pressure (PASP) of 120 mm Hg (Figure 1). Given the patient's history of chronic central nervous system infections and numerous neurosurgical procedures, CT pulmonary angiography (CTPA) was performed, which showed a large eccentric filling defect in both pulmonary arteries, partially occluding the lumen of the 1st-order branch of the right superior pulmonary artery and the 1st- and 2nd-order branches of the right inferior pulmonary artery (Figure 2). Thus, we concluded that the patient was suffering from CTEPH secondary to a VAS causing multiple thromboembolic episodes.

The patient was started on Digoxin 0.25 mg, Lasoride 40 mg, and Apixaban, resulting in the alleviation of her symptoms. Her dyspnoea improved, and her NYHA class was enhanced from IV to II.

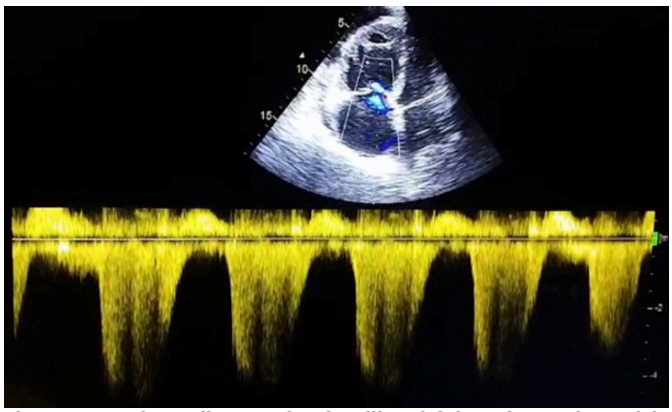

Figure 1: An echocardiogram showing dilated right atrium and ventricle with severe tricuspid regurgitation and a pulmonary artery systolic pressure (PASP) of 120 mmHg.

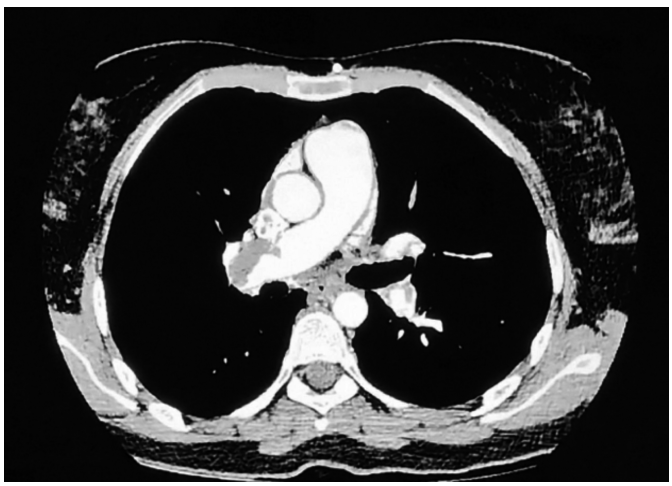

Figure 2: A CT pulmonary angiography showing occlusion of the first-order branch of the right superior pulmonary artery, first- and second-order branches of the right inferior pulmonary artery, and eccentric filling defect.

DISCUSSION

VPS is the preferred option for treating hydrocephalus. Still, some conditions cannot tolerate a VPS. The most frequently employed alternative standard shunt is the VAS to the right atrium. VAS became the gold standard in treating hydrocephalus in 1952. This, however, was soon followed by the realisation that VAS was not without flaws. Rather, it carried the risk of potentially fatal outcomes due to specific cardiac placement and systemic drainage leading to complications such as infections, shunt obstruction, and subdural haemorrhage.[4] Although VAS are linked to greater risks, including cardiopulmonary and renal diseases such as PH and corpulmonale, CTEPH is a relatively rare consideration for patients with VAS.[3] It is a rare sequel of pulmonary embolism. Potential risk, factors for it include a VAS, which may not be given due attention.[5] CTEPH is caused by pulmonary vascular obstruction by non-occluding thromboembolic.[6] SARC is an acronym for suspect, assess risk, and confirm and has been posed as a diagnostic approach to CTEPH. Published evidence has also shown that delays in diagnosis affect the prognosis. The patient presents with typical cardiopulmonary symptoms such as exertional dyspnoea and progressive exercise intolerance. As right ventricular (RV) dysfunction progresses, other symptoms of right heart failure, including abdominal swelling, oedema of the legs, shortness of breath, congestive chest pain, dizziness, and fainting on exertion, also develop. Pulmonary flow murmurs are auscultatory findings in about 30% of patients. However, while V/Q scintigraphy is the imaging modality of choice in excluding CTEPH, CTPA is beneficial in that the lung vasculature and parenchyma can be evaluated, as well as ruling out conditions that may mimic CTEPH. Although, such findings can sometimes go unnoticed by physicians who are not well acquainted with CTEPH.[6,7] Pulmonary endarterectomy is the treatment of choice for patients with CTEPH. Despite this, patients who are not suitable candidates for surgery such as those with advanced NYHA class, significant pulmonary dysfunction, signs of heart failure, or inconsistent imaging findings are typically managed by medical therapy.[8] The cornerstone of medical treatment for CTEPH is lifelong anticoagulation, such as direct oral anticoagulants (DOACs) for the prevention of thrombus formation. Research and clinical trials in medical therapy for CTEPH are ongoing. In practice, the treatment of CTEPH frequently involves a diversified approach.[7]

In conclusion, it is imperative to consider CTEPH as a possibility after VAS surgery in patients with hydrocephalus even when they are not presenting with typical signs of the disease. We believe that mitigating the adverse outcomes associated with CTEPH secondary to VAS requires close monitoring of individuals undergoing the VAS procedure by ensuring regular follow-up and the use of V/Q scintigraphy if necessary.

PATIENT'S CONSENT:

Informed consent was taken from the patient to publish this case.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

MH: Finalising of the topic, designing of the work, and drafting of the manuscript.
KF: Drafting the article, literature search, review of the manuscript.
ESM: Literature search, interpretation of data, and critical review.
MTF: Revision and critical analysis of the manuscript.
All authors approved the final version of the manuscript to be published.

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