Constrictive pericarditis-induced shunting through a PFO: Persistence despite pericardiectomy

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A B S T R A C T
A patent foramen ovale (PFO) is found in around 25–30% of patients. The discovery is often made only on autopsy, as most PFOs are clinically silent and any inter-atrial blood exchange typically shunts from the left to right heart [1]. Thus, when a patient presents with hypoxic respiratory failure, concern for presence of a PFO is rarely at the top of the differential. However, in the setting of elevated right heart pressures, PFOs can become of great hemodynamic importance and can lead to deadly complications, including right to left shunting and refractory hypoxic respiratory failure. We present an unusual case of constrictive pericarditis leading to significant shunting through a PFO, and resultant hypoxic respiratory failure which only resolved with PFO closure.

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1. Introduction

PFOs are a necessity during fetal development, as they allow for oxygenated blood to flow from the right to left atrium. The closure process begins in the fourth week of pregnancy, as the septum primum and secundum overlap and fuse, eventually progressing to flap closure at birth. The flap is completely fused by age two in 70–75% of children, with the remaining children having a PFO. It is not entirely clear why PFOs may fail to close, with possible contributions from genetics and familial factors [1]. Most patients with PFOs remain asymptomatic throughout their lifetime. However, situations can occur which lead to the development of symptomatology.

Constrictive pericarditis occurs due to scarring and pericardial damage, usually as a result of prior episodes of acute pericarditis. The pericardium becomes thickened and calcified, leading to impaired diastolic filling of the ventricles, with elevated right heart pressures and signs and symptoms of right heart failure. On cardiac catheterization, an equalization of diastolic pressures as well as a characteristic dip-and-plateau sign can be seen on hemodynamic tracings. In the setting of a PFO, the elevated right heart pressures can lead to potential for right-to-left shunting. The mainstay of therapy for constrictive pericarditis is surgery with pericardiectomy, which is a procedure that holds up to a 6–12% mortality [2]. Pericardiectomy is often done with the goal of improving symptoms of heart failure and dyspnea and preventing pulmonary hypertension [3,4]. It is uncommonly employed to improve dyspnea due to right-to-left shunting from a PFO. In theory, it should improve those symptoms and decrease shunting due to de-escalation of right atrial pressures; studies have shown that elevated preoperative cardiac pressures have declined after pericardiectomy. Yet, sometimes the elevated pressures may remain this way for weeks-months after the procedure [5,8], suggesting that no immediate benefit may be gained from pericardiectomy for hypoxic respiratory failure.

2. Case

A 75-year-old man with no significant past medical history presented with progressive dyspnea for one month. Upon presentation, he was found to be hypoxic with an oxygen saturation of 80%. His cardiac and pulmonary examinations were unremarkable. An arterial blood gas (ABG) showed pure hypoxia without hypercarbia. A chest x-ray was completely unremarkable. Due to concern for possible pulmonary embolism (PE), a CT scan with IV contrast was performed and was negative for PE; it also did not show any clear lung pathology. However, it did show a small pericardial effusion and pericardial calcifications.

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Trans-thoracic echocardiography with agitated saline was then done to evaluate for possible shunting, and confirmed moderate shunting through a PFO. Right heart catheterization (RHC) was then performed and showed an equalization of diastolic pressures, as well as the characteristic “square root sign,” consistent with constrictive physiology (Fig. 1).

Cardiac MRI was done for further classification, and did show a thickened pericardium with a small circumferential effusion and ventricular interdependence, confirming constrictive pericarditis. The workup for the etiology of his pericarditis was unremarkable, including negative autoimmune and tuberculosis testing.

The patient continued to be hypoxic, requiring oxygen by high-flow nasal cannula to maintain an adequate oxygen saturation. It was thought that the pericardial constriction was leading to elevated right heart pressures, and therefore the shunting and respiratory failure. The decision was made to undergo pericardiectomy, for which he was electively intubated. He underwent successful pericardiectomy with complete liberation of the bilateral anterior ventricles and 80% pericardial resection from the right atrium; certain areas of the ventricles (primarily the lateral and basilar walls of the left ventricle), the pericardium could not be completely removed and so was scored in a crosshatch pattern. After the procedure, he was unable to be weaned from the ventilator, and continued to require 100% FiO2. This indicated that the shunting was still ongoing through the PFO and causing hypoxia. The following day he underwent trans-esophageal echocardiography which again showed a moderate PFO with significant right-to-left shunting (Fig. 2).

The patient underwent another RHC which did show mildly elevated right sided filling pressures, continued equalization of right and left heart filling pressures with a restrictive right ventricular filling pattern, and significant right-to-left shunting (QP:QS = 0.86). He had successful trans-catheter placement of a PFO closure device. He was weaned from the ventilator and successfully extubated three days post-procedure, and discharged home several days later on room air. Repeat trans-thoracic echocardiography four months later did not demonstrate any shunting.

3. Discussion

This case demonstrates both the potential clinical significance of PFOs, as well as the treatment dilemma when there is shunting through a PFO due to constrictive pericarditis. PFOs, despite their high incidence, are typically not problematic for patients. When patients do become symptomatic, there can be various manifestations of their symptomatology. While treating physicians are frequently aware of PFO complications such as cryptogenic stroke, it is less frequently recognized as the culprit in hypoxic respiratory failure. However, it is imperative to recognize that PFOs need to be a part of the differential in the setting of hypoxic respiratory failure, especially when it is not correctable with supplemental oxygen. In this case, other sources of hypoxemia were first ruled out, including V/Q mismatch, hypoventilation, and diffusion limitation. Though typically benign, PFOs may be of great hemodynamic consequence in certain scenarios and can be easily overlooked if not considered.

Given that our patient’s shunting through the PFO was due to elevated cardiac pressures from constrictive pericarditis, it was initially thought to pursue pericardiectomy in order to decrease the cardiac pressures and therefore decrease the shunting. However, despite this action, the patient remained hypoxic and post-pericardiectomy imaging showed continued shunting. Most patients who are symptomatic due to constrictive pericarditis (though not necessarily with shunting through PFO) improve rapidly after
pericardiectomy [7,8]: however, some studies have shown that the hemodynamics may remain abnormal for up to several months in certain patients [8]. The reason for this has been related to the degree of surgical success as well as degree of myocardial inflammation and fibrosis [8]. Our patient did have challenging anatomy, especially in respect to the left ventricular pericardium, but was able to get near-complete liberation of the right ventricle and then 80% of the right atrium; however, he continued to have constrictive physiology demonstrated postoperatively. This may suggest that even in the setting of adequate surgical success, certain patients may not have immediate normalization of hemodynamics.

Because they are typically a benign entity, there is no general recommendation for shunt closure. However, there are situations in which it is recommended to undergo surgical PFO closure. The only steadfast recommendation is for closure of hemodynamically significant PFOs with resultant RV overload; closure can be considered in platypnea-orthodeoxia syndrome, evidence of paradoxical embolization, as well as other situational circumstances [6]. In this patient, the continued right-to-left cardiac shunting despite pericardiectomy lead to the requirement of trans-catheter closure of the PFO, after which he had marked improvement. He did not undergo post-operative cardiac catheterization after closure of the PFO, but did have a trans-thoracic echo done which did not demonstrate shunting, and his symptoms completely resolved. This would indicate that the shunting was the etiology of his profound, uncorrectable hypoxia, even after pericardiectomy.

4. Conclusion

PFOs are not always benign, and are an important and often unrecognized cause of hypoxic respiratory failure. Right-to-left shunting occurs through a PFO in the setting of elevated right sided cardiac pressures, such as in constrictive pericarditis. In theory, removal of the element of constriction would lead to decreased pressures, and therefore decreased shunting. However, in some patients, abnormal hemodynamics may persist for weeks-months and lead to ongoing shunting. In these situations, it may be necessary to undergo PFO closure to resolve ongoing hypoxia.

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