Anesthetic implications for coexisting cardiac capillary hemangioma and multiple coronary artery to pulmonary artery fistulas

ABSTRACT
Abnormalities of the coronary vascular branches and cardiac hemangiomas represent together unusual clinical entities, with an incidence difficult to establish for the former as the vast majority of the patients with these anomalous vascular connections are usually asymptomatic and 2.8% for the latter. Symptomatic patients may develop dyspnea on exertion or chest pain secondary to a “coronary steal” phenomenon as part of the underlying pathophysiology of the disease. The authors report a case of a patient with concomitant cardiac capillary hemangioma with multiple coronary to pulmonary artery fistula connections that was successfully treated with surgical resection of the tumor and ligation of the fistula tracts. A comprehensive and balanced anesthesia management aimed to preserve tight hemodynamic stability to avoid increased myocardial demand and worsening of the coronary steal becomes essential in these patients.

Key words: Anesthesia; cardiac hemangioma; coronary artery fistula; coronary steal; pulmonary artery fistula

Introduction
Abnormal connections between the main coronary artery and its branches with other major thoracic vessels or with the heart chambers represent a rare clinical entity with an incidence of 0.002%–0.32% among the general population, with a male to female ratio of 1.9:1 although at least 50% of these cases are asymptomatic, discovered incidentally and more often congenital than other acquired forms (traumatic, iatrogenic, or associated to cardiac neoplasms).[1‑5] Meanwhile, the incidence reported for cardiac hemangiomas is 2.8% among all primary heart neoplasms and are classified as cavernous, capillary, or arteriovenous hemangioma according to their histological features.[4,6]

Case Report
A 76-year-old male with previous medical history of hypothyroidism and gastroesophageal reflux disease presented to our institution for thoracotomy and surgical ligation of coronary to pulmonary artery fistulas (CPAFs). The patient was initially evaluated by cardiology for dyspnea on exertion that onset 3 months prior; consequently, a dobutamine stress trans-thoracic echocardiogram was
performed and revealed a normal left ventricular ejection fraction (65%) with a reversible hypokinesis of the anterior and septal portions on maximum dobutamine stimulation.

Further work-up included right coronary catheterization which displayed normal pulmonary artery pressures, cardiac output, and index within normal limits calculated by Fick (5.43 L/min and 2.33 L/min/m² respectively), “step-up” in the oxygen saturation levels from the right ventricle (59%) to the right and left pulmonary arteries (65% and 61%, respectively). Left coronary catheterization reported a “tortuous-appearing and tangling vessels” at the proximal and mid-left ascending artery with a fistula run-off into the pulmonary artery system [Figure 1 and Supplemental Video 1]. Additional imaging through coronary computed tomography (CT) portrayed more precisely the “vessel tangling” between the main pulmonary artery and the left atrial appendage with unclear source of feeding, and a conus artery (branch of right coronary artery) run-off into the main pulmonary artery [Figure 2].

The patient was taken to the operating room for surgical ligation through median sternotomy approach under general anesthesia. Postinduction transesophageal echocardiography revealed the presence of jets from the coronary circulation into the pulmonary artery during diastole [Figure 3 and Supplemental Video 2]. Direct visualization revealed a tumor structure at the distal aspect of the main pulmonary artery, and since the surgeon was not able to visualize the origin of the vessels feeding the tumor and the fistula tracts, the patient was started on cardiopulmonary bypass (CPB) after full heparinization. Several fistula tracts were ligated including one from the conus artery and entry points at the anterior and lateral aspect of the main pulmonary artery as well as resection of the mass, which was found to be a hemangioma after surgical pathology examination. The patient was separated from CPB, heparinization was reversed, mediastinum was closed after adequate hemostasis, and the patient was extubated in the operating room and sent to the intensive care unit for further recovery.

Discussion

CPAFs are classified based on their origin as congenital or acquired and based on their hemodynamic repercussions over the myocardium as insignificant (asymptomatic) or significant (symptomatic).[7] According to previous studies, the presence of symptoms is seen in 19%–63% of patients, with dyspnea on exertion as the most common presentation, although atrial arrhythmias, pulmonary hypertension, congestive heart failure, presence of aneurysms, hemoptysis, angina, and endocarditis have also been reported.[1] Verdini et al. in a systematic review of 103 patients with CPAFs found a 63% male prevalence, the presence of a murmur as the most common physical examination finding (37%), with the left main and left anterior descending artery as the
most common site of origin (84%) and the main pulmonary artery as the most common site of merging (89%). These patients may undergo either surgical ligation or percutaneous transcatheter embolization of the fistula tract as part of the therapeutic options.[1]

Cardiac hemangiomas follow a variable and unpredictable clinical course, with no predilection for any particular age group and with a clinical presentation based on its location and the structures surrounding.[4,6] A literature review performed by Han et al., of 56 cases revealed that the slight majority of these tumors arise in the right ventricle (35.7%) followed by the left ventricle (33.9%), at least one-third of the cases had multiple tumors and patients presented arrhythmias, pericardial effusion, congestive heart failure, right ventricular outflow tract obstruction, or coronary insufficiency as presenting symptomatology.[6,9]

Xu et al. described a similar entity with an associated dilation of the tricuspid annulus leading to moderate regurgitation that required resection of the tumor and ligation of the fistula tract tricuspid valvuloplasty was also performed.[6] Anesthesia management plays a paramount role in the perioperative care of these patients as they pose an elevated risk for cardiovascular decompensation. Normovolemia and normal heart rate to decrease coronary steal and pulmonary artery hyperflow as well as maintaining an anesthetic plane to prevent sympathetic hyperactivity are utmost important for the favorable outcome of these patients.[5]

The case presented above depicted a symptomatic patient with no prior cardiac history who was found on further work-up the presence of CPAFs in the setting of an underlying cardiac capillary tumor. Due to the complexity of the anatomy displayed in the catheterization and coronary CT imaging, it was determined that the patient would be a more appropriate candidate for open surgical ligation with further exploration of the “tangling of vessels,” which later on resulted in the presence of a hemangioma. A thorough literature review revealed as the best to the authors’ knowledge the second case reported with coexisting multiple CPAFs and cardiac hemangioma. Cardiac catheterization, coronary CT, and transesophageal echocardiography provide invaluable information for the anatomic delineation and hemodynamic impact of both pathologies and ultimately outlining the best therapeutic decision for these patients.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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