Case Report

Anaesthetic management of a parturient with an unrepaired coronary arteriovenous fistula for caesarean section

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Summary

Coronary arteriovenous fistulas are an abnormal conduit between a coronary artery and another cardiovascular lumen, without an intervening capillary bed. The reported prevalence is 0.002–0.3%. Physiologic consequences such as congestive heart failure, coronary steal phenomenon and fistula aneurysm formation and rupture are possible. There are limited reports of symptomatic coronary arteriovenous fistulas in association with pregnancy. We describe a 19-year-old woman with symptomatic left circumflex artery to coronary sinus fistula, terminating into a large exophytic varix in the right atrium, presenting for an elective caesarean section at 37 weeks gestational age. Our anaesthetic management strategy aimed to optimise myocardial perfusion, maintain euvolemia, avoid right ventricular obstruction from exophytic varix and avoid sympathetic stimulation or sudden increases in pulmonary vascular resistance. A slowly titrated epidural was used as the primary anaesthetic. Our patient tolerated the procedure well and was discharged home on postoperative day two. Understanding of the potential physiologic consequence of coronary arteriovenous fistulas, and interaction with the physiologic changes of pregnancy and delivery, are essential for the management of these cases.

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Introduction

Coronary artery fistulas (CAF) are an abnormal conduit between a coronary artery and another cardiovascular lumen, such as another vessel (coronary arteriovenous fistula) or cardiac chamber (coronary-cameral fistula), without an intervening capillary bed [1]. Most patients with coronary arteriovenous fistulas (CAVF) are asymptomatic. If symptoms do appear, they typically present after the age of 20 [2]. Presenting signs and symptoms include exertional dyspnea, fatigue, palpitations, atrial arrhythmias, continuous cardiac murmur, angina and congestive heart failure. Surgical intervention is the definitive therapy for patients with symptoms or complications related to their CAF.

There are very few published reports of CAVF in association with pregnancy [3–5]. In this report, we describe the anaesthetic management of a 19-year-old woman with symptomatic CAVF of her left circumflex to coronary sinus, presenting for an elective caesarean section at 37 weeks gestational age.

Report

A 19-year-old, nulliparous woman presented to the emergency department with a 1-month history of intermittent exertional chest pain, exertional dyspnea (New York Heart Association class IV), palpitations, and two episodes of syncope. There was no other notable medical or surgical history, and her family history was not known to her. She appeared well on initial physical examination, with no apparent signs of distress or congestive heart failure. Chest radiography revealed a slightly enlarged pericardial silhouette, with normal pulmonary vascularity. Transthoracic echocardiogram (TTE) was performed which revealed a large ill-defined, ovoid, mobile mass in the right atrium, with normal left and right ventricular function. Further investigation with cardiac computerised tomography (CT) then characterised a macrofistulous communication of the distal left circumflex artery with the coronary sinus and exophytic varix extending into the right atrium. Transoesophageal echocardiogram (TOE) demonstrated a 3 × 2 cm right atrial mass which was continuous with the coronary sinus (Fig. 1a and b). High-velocity flow was noted from the mass in systole and diastole emptying into the right atrium from two distinct areas, with a maximum gradient of 70 mmHg (Fig. 1c). In anticipation of surgical intervention, a coronary angiogram was performed, which showed a diffusely dilated left circumflex artery and distal fistula to the coronary sinus, but otherwise structurally normal coronary anatomy. The mass was determined to be formed secondary to the shunt jet distending the Eustachian valve of the coronary sinus. Finally, she underwent a myocardial perfusion study (MIBI) which showed no evidence of significant ischemia, but her ejection fraction reduced from 60% to 53% with stress. Ultimately, coronary fistula ligation, resection of eustachian valve and possible circumflex bypass were planned.

Six months later, and just a few weeks before her scheduled surgery, it was discovered that she was now 7 weeks pregnant. The decision was made to delay surgical intervention until after her delivery. During her pregnancy, she developed gestational
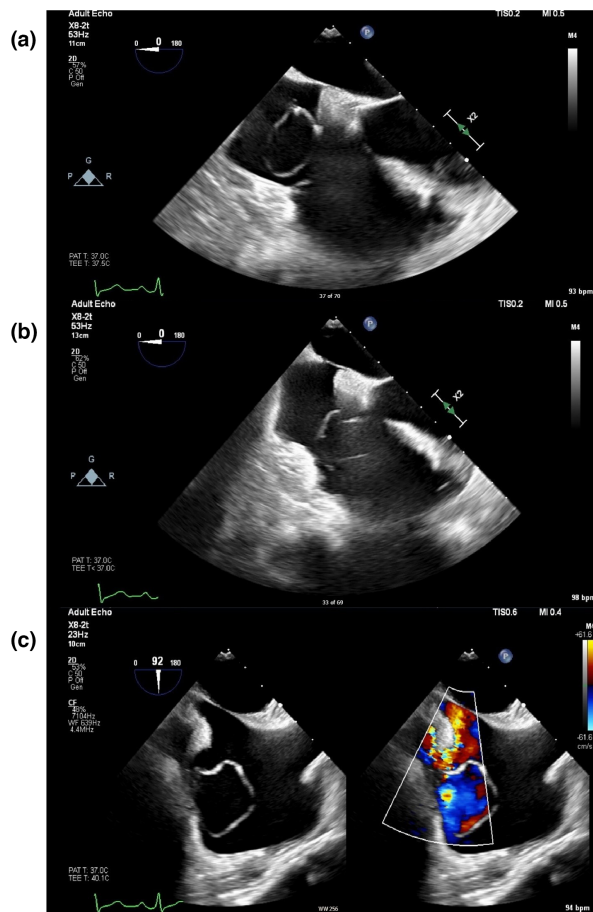
Figure 1 (a, b) transoesophageal echocardiography images demonstrating a 3 × 2 cm right atrial mass, continuous with the coronary sinus; (c) colour Doppler demonstrating flow into the right atrium.

hypertension which was controlled with metoprolol. She also developed worsening fatigue, dyspnea and presyncope beginning at 20 weeks gestational age. Despite this, her clinical course throughout her pregnancy was relatively uneventful, and repeat TTE was unchanged. Fetal ultrasound demonstrated fetal macrosomia above the 98% percentile, thus an elective caesarean section was arranged at 37 + 5 weeks gestation.

Anaesthetic management included insertion of two large-bore intravenous cannulae, an arterial line and a lumbar epidural. A crystalloid fluid bolus was administered before epidural titration. Left uterine displacement was maintained throughout the procedure. Her epidural was titrated slowly until a block at the level of T5 was achieved, with a total of 18 ml of 2% lidocaine with 1:100,000 adrenaline and 75 mcg fentanyl. A phenylephrine infusion was used intermittently to maintain a mean arterial pressure above 70 mmHg. After delivery, 100 mcg of carbetocin was administered via slow intravenous injection; no additional uterotonics were required and the estimated blood loss was 500 ml. She was transferred to the recovery room where she was continuously monitored and remained in stable condition. The remainder of her postpartum care was uneventful, and she was discharged home on postoperative day two.

Discussion

The reported prevalence of CAFs in the general population varies from 0.002% to 0.3% by invasive angiography, and up to 0.9% by CT angiography [6, 7]. Complex malformations causing significant haemodynamic consequences are exceptionally rare. Most CAFs are congenital in origin, but they can also occur secondary to trauma, infection, or iatrogenic injury [2]. Coronary arteriovenous fistulas originating from the left circumflex occur in 18% of cases, while 50–60% originate from the right coronary artery, 30% from the left anterior descending, 1.9% from the diagonal branch, and 0.7% from the left main coronary [6, 8]. They often terminate into right-sided structures such as the pulmonary artery, right ventricle, right atrium, coronary sinus, or superior vena cava, however, termination into the left ventricle, pulmonary veins and left atrium is also possible [2, 6].

The pathophysiology of CAFs involves shunting of blood from a high-pressure arterial system into a low-resistance venous system, bypassing the capillary bed in the myocardium. The size of the shunt is determined by fistula resistance, the pressure differential between the two systems, and the site of fistulous termination [7]. The potential physiologic consequences and considerations may include [2]: congestive heart failure and pulmonary hypertension from excessive left-to-right shunt; coronary steal phenomenon whereby coronary blood flow is diverted from the myocardium into the low resistance fistula, leading to myocardial ischemia; massive dilatation and aneurysm formation of the coronary artery proximal to the fistula, increasing risk of fistula rupture; valvular and endocardial complications such as valvular regurgitation and infective endocarditis; and atrial and ventricular arrythmias from chamber dilatation.

In our patient, the CAF also led to the formation of a large exophytic varix formed secondary to the shunt jet distending the Eustachian valve. The concern of this mobile mass is the potential obstruction of blood flow across the right atrium and/or tricuspid valve, which could theoretically impede right ventricular preload and therefore cardiac output. Although our patient did not show any signs of obstruction on TOE, dynamic obstruction during periods of hypovolemia may have been possible. Despite exertional angina and concern for coronary steal phenomenon, there was no evidence of stress-induced myocardial ischemia on her stress MIBI. Interestingly, her ejection fraction did reduce significantly with stress, which was concerning for a limited cardiac reserve. She showed no signs of congestive heart failure, pulmonary hypertension, valvulopathies, or other cardiac anomalies during her initial investigations, which was reassuring. Since these investigations, however, significant physiologic changes associated with pregnancy occurred, along with increasing severity of symptoms. Additional considerations associated with pregnancy and caesarean section delivery included [9]: increased cardiac output, sympathetic stimulation and myocardial oxygen demand, particularly during the time of delivery. These factors may lead to myocardial ischemia, worsen coronary steal phenomenon and cardiac decompensation; increased total blood volume and auto-transfusion after delivery, increasing risk for congestive heart failure; aortocaval compression and blood loss during delivery, which may reduce pre-load and potentially exacerbate right atrial mass obstruction; and administration of uterotonics such as carbetocin or oxytocin, which can cause systemic vasodilation and hypotension, tachycardia and increased cardiac output [10]. This may increase myocardial oxygen demand and reduce coronary perfusion pressure, which can lead to myocardial ischemia.

A multi-disciplinary approach involving anaesthesia, obstetrics, cardiology and cardiovascular surgery was imperative for pre-operative planning and optimisation. Our anaesthetic management was aimed to optimise myocardial perfusion, maintain euvolemia, avoid obstruction from exophytic varix and avoid sympathetic stimulation or sudden increases in pulmonary vascular resistance. An arterial line was inserted to allow rapid detection of haemodynamic instability. A slowly titrated epidural was selected as the primary anaesthetic given the haemodynamic stability, exceptional pain control and favourable bleeding risk

profile. Large bore intravenous access was obtained, and a fluid bolus was given before epidural titration to avoid sudden changes in preload. Intravenous carbetocin was administered slowly over 2–5 min, as opposed to a bolus, to minimise the haemodynamic side effects [10]. Central venous access and/or pulmonary arterial catheterisation may also be considered. Peri-operative cardiovascular collapse, ST-segment changes, myocardial infarction, pericardial tamponade, fistula dissection and coronary spasm have been all described, therefore, peri-operative vigilance was essential [5, 11]. Appropriate understanding of the potential physiologic consequences of CAVF and how they interact with the physiologic changes of pregnancy and delivery are essential for the management of these cases.

Acknowledgements

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