Spontaneous coronary artery dissection in a parturient with Nail–Patella syndrome

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ABSTRACT
Spontaneous coronary artery dissection is an uncommon cause of acute coronary syndrome, occurring predominantly in women during and immediately after pregnancy; it carries a mortality rate of greater than 50%. While the exact etiology is unknown, possible contributing factors include pregnancy-related hormonal, connective tissue and hemodynamic changes. We present a case of a 35-year-old multigravid woman with Nail–Patella syndrome who developed an acute myocardial infarction secondary to spontaneous coronary artery dissection during labor which was not diagnosed until after delivery. We hypothesize that abnormal collagen fiber formation found in Nail–Patella syndrome may have put her at an increased risk of coronary dissection and myocardial infarction. Regardless of etiology, a delay in diagnosis of myocardial ischemia can lead to significant morbidity and mortality. In light of the increasing burden of cardiac disease in the obstetric population, clinicians should remain vigilant for signs of myocardial infarction and prepare for definitive diagnosis and treatment.

Keywords: Acute coronary syndrome; Coronary artery dissection; Nail–Patella syndrome

Introduction

Spontaneous coronary artery dissection (SCAD) is rare in the non-pregnant population, but is the primary cause of acute myocardial infarction in the immediate peripartum period.[1] The reason for its higher prevalence continues to be investigated; however, it is postulated that progesterone excess, resulting in structural endothelial changes or eosinophilic infiltration, is associated with collagen damage and weakening of the vessel adventitia making dissection more likely.[2–4] Some underlying diseases associated with SCAD include connective tissue disorders such as Marfan and Ehlers-Danlos syndrome, vasculitic disorders such as polyarteritis nodosa and systemic lupus erythematosus, antiphospholipid antibody syndrome and inflammatory bowel disease.[5,6]

Nail–Patella syndrome, also known as hereditary onycho-osteodysplasia, Fong disease or Turner–Kieser syndrome, is a collagen vascular disorder which is traditionally associated with dysplastic nails, aplastic or hypoplastic patellae and renal pathology. To our knowledge SCAD has not previously been reported in a patient with Nail–Patella syndrome.

Case report

A 35-year-old G2P1 woman was admitted to the antepartum service for threatened preterm labor at 30 weeks of gestation. Her height was 152 cm, weight 62 kg, and body mass index 26.8 kg/m^2. Her pregnancy was marked by several evaluations and two admissions for preterm contractions, cerclage placement, tocolysis and a course of steroids for fetal lung maturity. The patient had a past obstetric history of an emergency cesarean delivery due to preterm labor at 27 weeks with breech presentation. Her past medical history included Nail–Patella syndrome, gestational diabetes, Hashimoto's thyroiditis, vitiligo and gastroesophageal reflux. She had no additional known risk factors for coronary artery disease and denied a history of tobacco, alcohol or recreational drug usage. Her home medications included prenatal vitamins.

On the antepartum floor she experienced sudden onset of chest heaviness and left arm pain. Vital signs included blood pressure 120/70 mmHg, heart rate 94 beats/min and oxygen saturation 100% on room air. An electrocardiogram (ECG) (Fig. 1) was interpreted as normal by the obstetric team and she was given sodium citrate to treat presumed gastroesophageal reflux. Symptoms resolved quickly, and the anesthesia and cardiology teams were not informed of her chest pain: no further investigations were pursued by the obstetric team.

The next morning the patient experienced more uterine contractions and variable fetal heart rate decelerations. Given a past history of vertical hysterotomy contraindicating trial of labor together with the non-reassuring fetal heart tracing, a decision was made to proceed with urgent cesarean delivery. During preoperative assessment, the patient denied any history of chest pain or shortness of breath. Before neuraxial placement,

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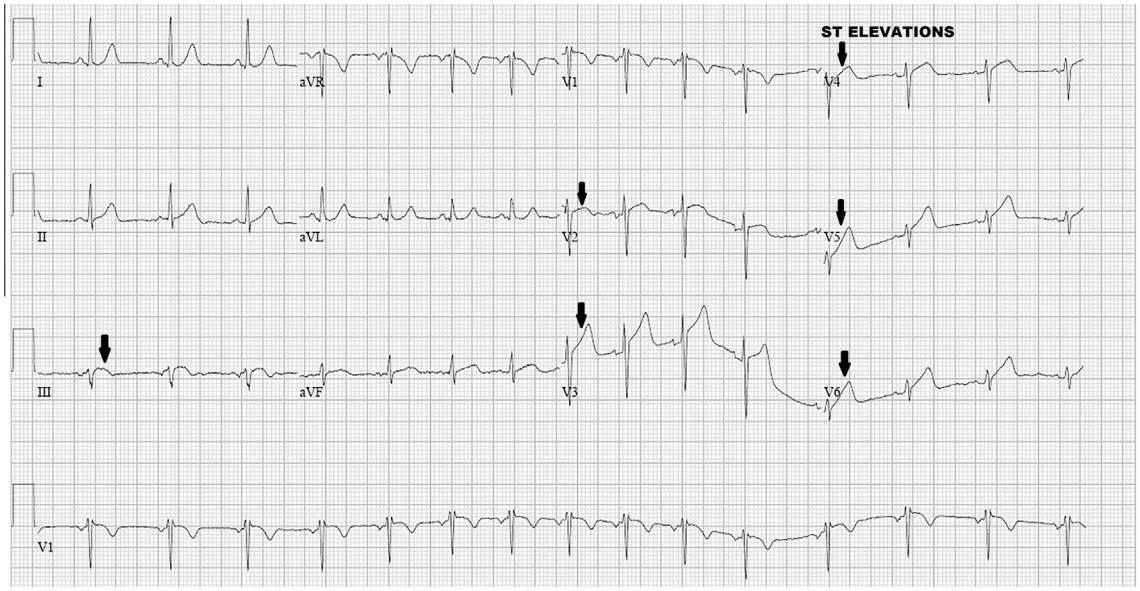

Fig. 1 ECG performed during patient's initial episode of chest pain one day before her cesarean section. There are subtle ST changes in the inferior and lateral leads.

vital signs included blood pressure 127/63 mmHg, heart rate 145 beats/min, and oxygen saturation 98% on room air. Her tachycardia was presumed to be related to anxiety. One provider noted the presence of possible Q waves on the five-lead intraoperative ECG but assumed it was related to the poor quality of the tracing.

Spinal anesthesia was performed with the patient sitting using 0.75% hyperbaric bupivacaine 1.6 mL, fentanyl 10 μg, and preservative-free morphine 0.2 mg. A bilateral T4 level of anesthesia was established after testing sensation to pinprick. Maternal tachycardia persisted (130–140 beats/min) throughout surgery but she denied discomfort. Phenylephrine boluses of 80 μg were initially required for blood pressure support after neuraxial anesthesia, and subsequently as an infusion at 40 μg/min. Oxygen saturations remained stable at 98% with 6 L/min facemask oxygen. The baby was born with Apgar scores of 7 and 9 at 1 and 5 min, respectively, breathing spontaneously, and was admitted to the neonatal intensive care unit for monitoring due to prematurity. Surgery was uncomplicated; estimated blood loss was 900 mL and the patient received lactated Ringer's solution 3 L and oxytocin 20 U in 500 mL 0.9% saline as an infusion over 45 min. Intrathecal morphine served as her primary postoperative analgesic medication in conjunction with oral ibuprofen for the first 24 h postoperatively.

Approximately one hour after delivery, the patient started coughing violently and complained of shortness of breath. Oxygen saturation was 88% and she was placed on a non-rebreathing oxygen mask. Vital signs included blood pressure 124/63 mm/Hg, heart rate 120 beats/min and respiratory rate 22 breaths/min. Bilateral crackles at the lung bases were heard and an emergency chest X-ray suggested acute pulmonary edema. She was given intravenous furosemide 20 mg and her symptoms improved over 20 min. A further ECG (Fig. 2) showed incomplete right bundle branch block, loss of R wave progression and anterior forces and Q-waves in V4-V5. Troponin-T was elevated at 2.8 ng/mL. A presumed diagnosis of myocardial infarction was made 20 h after her initial presentation with chest pain. An echocardiogram showed anteroseptal hypokinesis and apical akinesis, with an overall ejection fraction of 45–50%.

Further management took place in the cardiac step-down unit where she was started on metoprolol, aspirin and a heparin infusion. Cardiac catheterization, performed the following day, showed a completely occluded left anterior descending (LAD) artery and possible intramural hematoma with significant LAD dissection extending proximally into D1 (Fig. 3). Radial artery vasospasm precluded further intervention. Two days later, two drug-eluting stents were placed in the LAD (Fig. 4). She was discharged home on aspirin, clopidogrel, verapamil and metoprolol.

Discussion

The most common etiology of myocardial infarction during pregnancy is coronary atherosclerosis.[1] However, when acute myocardial infarction occurs in the immediate peripartum period, the cause in 50% of cases is coronary artery dissection.[7–9] Dissection of the coronary artery occurs when the layers of the arterial wall separate to create a false lumen. The dissection occurs