Catastrophic Post-Cesarean Aortic Dissection in a Patient with Loeys-Dietz Syndrome

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Introduction: Loeys-Dietz syndrome (LDS) is an autosomal dominant connective tissue disorder, similar to Marfan and Ehlers-Danlos syndromes, caused by a heterozygous mutation of transforming growth factor-B receptor 1 or 2, which results in overproduction of collagen, loss of elastin, and disarray of elastic fibers (1,2). Clinical characteristics include: craniosynostosis, micrognathia, cleft palate, scoliosis, cervical spine instability, joint laxity, chest wall deformity, and arterial tortuosity and aneurysms. Median survival of LDS patients is 37 years, which is shorter than patients with Ehlers-Danlos (48 years) or Marfan’s (70 years) syndromes (2). Pregnancy is an extreme cardiovascular stress for LDS patients. Here, we report a LDS patient who had an uneventful pregnancy and cesarean delivery but developed postpartum aortic dissection.

Case: A 39 yo G1P0 woman with LDS, status post valve-sparing aortic root replacement, presented at 35 6/7 weeks with painful contractions. The patient had had an uneventful pregnancy. Multiple echocardiograms revealed stable aortic root diameter and a 3.4 cm ascending aortic aneurysm. Elective primary cesarean delivery had been scheduled.

On presentation, she had elevated blood pressure (140-150 mmHg systolic). We performed a cesarean section with spinal anesthesia. A 2080 g female infant was delivered uneventfully. The patient did well postoperatively and was transferred to the floor for routine postpartum care.

On POD2, the patient developed chest, back, and neck pain. A chest CT revealed a descending aortic dissection. Surgery was not performed because she was hemodynamically stable. She continued to have chest pain and a repeat chest CT the next day revealed extension of the dissection into the arch and left carotid artery with rupture into the mediastinum. She underwent emergent replacement of her descending thoracic aorta. Three days later, she suffered a cardiac arrest secondary to acute rupture of the proximal graft anastomosis, which was repaired with an endovascular stent. The next day a distal anastamotic leak was repaired with a second endovascular stent. She was discharged home one month later with cortical blindness and an aortic root dilated to 5.6 cm.

Discussion: Parturients with LDS are at risk for aortic dissection and rupture, uterine rupture, and damage to the vagina, perineum, and colon both during and after pregnancy (3). Avoiding hypertension, with the right antihypertensive agents, is imperative. Losartan is associated with slower, while calcium channel blockers are associated with more rapid, aortic root dilation than beta blocker therapy (4).

References: