Chest Pain in a Pregnant Patient with Takayasu Arteritis

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Patient: 30 years, female

Clinical History:

A 31 week pregnant patient with Takayasu arteritis (TA) presents to the emergency department with one day of low back pain, chest pain and shortness of breath.

Imaging Findings:

A 30 year old patient, with one prior uncomplicated pregnancy, presented at 31 weeks of pregnancy to our emergency department with complaints of low back pain radiating to her chest and shortness of breath for 1 day. Physical exam revealed sinus tachycardia and a low grade fever. Fetal and cervical exams were unremarkable.

A computed tomography angiogram (CTA) was performed to evaluate primarily for pulmonary embolism (PE) but also to rule out aortitis secondary to TA. This showed complete occlusion of the right main pulmonary artery and significant narrowing of the lower thoracic aorta (Fig 1). In retrospect the patient's prior chest x-rays and non-contrast CT scan of the patient's chest performed 4 years earlier supported the impression that the findings were chronic in nature (Fig 2). Echocardiogram demonstrated normal ventricular function without pulmonary artery hypertension.

The patient's work up for PE, myocardial infarct, and aortic dissection were negative. The fetal heart rate remained reassuring throughout her hospitalization but she was found to have preterm contractions and a positive influenza A titer. After tocolysis and subsequent decrease in preterm contractions her pain and shortness of breath significantly improved, she was discharged home with scheduled follow up. Dehydration secondary to influenza infection leading to preterm contractions were thought to be the cause of the patient's presenting symptoms.

The patient returned to the hospital the following week with premature rupture of membranes at 32 weeks, 1 day and delivered a healthy baby with Apgar scores of 8 and 8.

Discussion:

Takayasu arteritis (TA) is a chronic inflammatory disease that affects the aorta and its major branches. The disease usually affects women in their 2nd and 3rd decades of life during childbearing age. Cellular inflammatory infiltrates in the wall of the artery, primarily within the media, cause thickening of the arterial wall.[1, 2] This is followed by intimal and adventitial fibrosis. This process leads to luminal narrowing which can eventually cause complete occlusion. Alternatively, destruction of the media or elastic lamina of the vessel can lead to diffuse dilatation and formation of aneurysms.[1, 2] Traditionally, TA was thought to follow a triphasic pattern of expression consisting of a systemic/preinflammatory phase, a vascular inflammatory phase and finally a "burn out" phase characterized by fibrotic stenotic lesions.[3] However, currently it is recognized that all phases can coexist.[3] The distribution of vessels involved varies between patients. The aorta, the great vessels of the aortic arch and pulmonary arteries are
most commonly affected. Renal and mesenteric arterial involvement is also seen.

A search of “Takayasu arteritis + pregnancy” on both OVID/Medline and Pubmed revealed 88 and 80 articles respectively. A search of “Takayasu arteritis + pregnancy + pulmonary” revealed only 2 articles. Reviewing these results many articles reported outcomes of pregnant women with TA, however, only two reported outcomes of pregnant women with documented pulmonary artery involvement [4, 5]. These two cases both end with the delivery of babies with good Apgar scores, in the first case mother did well. In the second the mother, who was diagnosed with TA during her pregnancy, died of postoperative complications after repair of a thoracic aortic aneurysm. Both articles quote the high rate of pulmonary artery hypertension in women with TA, 50% [6]. The article by Jacquemyn et al. mentions the mortality of pulmonary artery hypertension in pregnant women to be 30 – 36 % and the general recommendation to terminate the pregnancy in such cases [4]. The article they reference states maternal mortality to be 30% in women with primary pulmonary artery hypertension and even higher, 56%, in women with secondary vascular pulmonary hypertension, the group that includes pulmonary hypertension secondary to TA [7]. However, this paper states that SVPH is a very heterogeneous group of disorders some of which lead to severe pulmonary hypertension but in reference to TA they state, “On the other extreme, in Takayasu’s arteritis … the pregnancy, delivery and postpartal period were experienced without difficulties” [7].

The case presented here is of a patient with TA and severe pulmonary artery involvement but without pulmonary hypertension. This case documents pulmonary artery involvement in a pregnant woman with TA, more severe than the previous two cases reported in the literature, further supporting the claim that women with TA and pulmonary artery involvement can tolerate pregnancy with a good outcome for themselves and their babies.

**Differential Diagnosis List:** Chronic obstruction of the right pulmonary artery from Takayasu Arteritis

**Final Diagnosis:** Chronic obstruction of the right pulmonary artery from Takayasu Arteritis

**References:**


Description: Contrast enhanced CT of the chest shows complete occlusion of the right main pulmonary artery (long arrow) but normal enhancement of the left main pulmonary artery (short arrow).

Origin:
Description: The same scan more inferior shows filling of peripheral pulmonary arteries on the left but no filling on the right. **Origin:**
Description: A coronal reformatted image further shows the lack of contrast within the right pulmonary arteries (long arrow) but enhancement of the left pulmonary arteries (short arrow). Also note the narrowing of the descending aorta. Origin:
**Description:** A more inferior axial image shows narrowing of the inferior thoracic/upper abdominal aorta (arrow). **Origin:**
Description: Chest radiograph taken prior to the CT scan in figure 1 shows a subtle Westermark sign (cutoff of the pulmonary vessels) in the right hemithorax. Origin:
Description: A non-contrast CT from 4 years earlier shows narrowing of the right pulmonary artery (arrow). Origin: