Atypical HELLP Syndrome in a Pregnant Patient with Takayasu Arteritis and Subclavian to Axillary Artery Saphenous Vein Bypass Graft: A Case Report

Hediye DAĞDEVİREN[1], Hüseyin CENGİZ[1], Sema Süzen ÇAYPINAR[1], Cihan KAYA[1], Ammar KANAWATI[1]

İstanbul, Turkey

Takayasu arteritis (TA) is a rare chronic granulomatous inflammatory disease of the aorta and/or its major branches, affecting mostly the vertebral, carotid, subclavian, iliac, and renal arteries. The disease shows a striking predilection for women during the child-bearing years, and it is reasonable to expect at least 1 pregnancy event in these women. Therefore, the management of pregnancies in patients with this disease is of great importance to obstetricians. Here, we present the case of a patient with atypical hemolysis, elevated liver enzymes, and low platelets (HELLP syndrome); this patient was receiving corticosteroid therapy and had undergone subclavian-axillary artery saphenous vein graft for TA.

Keywords: Hellp syndrome, Takayasu arteritis, Vein bypass graft

Gynecol Obstet Reprod Med 2014;20:163-165

Introduction

Takayasu arteritis (TA) is a rare chronic granulomatous inflammatory disease of the aorta and/or its major branches, affecting mostly the vertebral, carotid, subclavian, iliac, and renal arteries; some authors speculate that TA is a spectrum of a single disease within giant cell arteritis.[1] The disease shows a striking predilection for women during the child-bearing years,[2] and it is reasonable to expect at least 1 pregnancy event in these women. Therefore, the management of pregnancies in patients with this disease is of great importance to obstetricians. Although the inflammatory activity of the disease is thought to be unaffected by pregnancy, complications such as intrauterine growth restriction, fetal death, hypertension, heart failure, and aneurysm rupture have been described.[3-5] The few series published so far have evaluated maternal and fetal outcomes in pregnant patients with TA, and since most articles are case reports, the data are not conclusive. Here, we present the case of a patient with atypical hemolysis, elevated liver enzymes, and low platelets (HELLP syndrome); this patient was receiving corticosteroid therapy and had undergone subclavian-axillar artery saphenous vein graft for TA.

Case Report

A 33-year-old primigravid who was at 19 weeks' triplet gestation presented to the emergency room with a 2-day history of worsening epigastric pain, headache, nausea, and vomiting. The patient did not have chest pain, shortness of breath, or palpitations. She was under the care of a medical doctor in another hospital. She had a medical history of hypertension and anemia since 2008 and had undergone an arm operation 5 years previously. She had a history of TA since 2008. On admission to the emergency room, her obstetrical and medical records were not available. The patient was taking amlodipine, infliximab, prednisolone, prenatal vitamins, and iron supplements. She was subsequently admitted to the obstetric ward.

On physical examination, blood pressure (BP) was 180/120 mm Hg in the right arm and 170/110 mm Hg in the left arm, pulse rate was 100/minute, and the oxygen saturation in room air was 100%. A Grade II systolic murmur, optimally heard at the left sternal border with radiation to the carotids, was apparent. The pulse was palpable in all extremities, although the pulse in the left arm was slightly diminished as compared to that in the right. Electrocardiogram (ECG) revealed a normal sinus rhythm, and cardiac enzymes were not elevated. Abdominal examination revealed positive bowel sounds and a tender epigastrium with no fundal tenderness, rebound, or guarding. Lower extremities revealed 2+ pitting edema. On vaginal examination, there was no dilatation or effacement of the cervix. Obstetric ultrasonography revealed all growth-restricted viable triplet pregnancy and bilateral uterine artery notching Doppler studies showed normal findings for the carotid artery, but the subclavian arteries were not clearly visualized. Considering the findings of vascular and cardiac examinations, the cardiology and rheumatology service was consulted. Treatment with steroids, nifedipine, and methyldopa was initiated.

[1]*Bakirkoy Dr. Sadi Konuk Teaching and Research Hospital, Istanbul,*

Address of Correspondence: *Hediye Dağdeviren*
Tevfik Sağlam Street, No: 11,
Zuhuratbaba, Bakirköy İstanbul, Turkey
hediyedagdeviren@gmail.com

Submitted for Publication: *09. 08. 2013*
Accepted for Publication: *24. 09 2013*

Initial laboratory values were as follows: hemoglobin, 11 g/dL; hematocrit, 32%; platelets, 80.000 103/mm^3 (normal range, 150.000-400.000 103/mm^3); aspartate aminotransferase (AST), 358 U/L (normal range, 0-34 U/L); alanine aminotransferase (ALT), 233 U/L (normal range, 0-55 U/L); and lactate dehydrogenase (LDH), 1181 U/L (normal range, 125-243 U/L). Urinalysis revealed grade 3+ proteinuria. Erythrocyte sedimentation rate and blood levels of urea, creatinine, C-reactive protein, alkaline phosphatase (ALP), and gamma glutamyl transpeptidase (GGT) were within the normal range. On follow-up (1 hour after administration), gradual increases and decreases in liver enzymes and platelet levels were noted, respectively. Platelet levels decreased to 16.000×103/mm^3. A transfusion of 4 units fresh frozen plasma, 4 units erythrocyte suspension, and 12 units platelet suspension was initiated. Patient with elevated BP, thrombocytopenia, and elevated liver enzymes was diagnosed with HELLP syndrome. During laboratory analyses, the patient reported worsening epigastric abdominal pain and had elevated BP (200/110 mm Hg), as measured in the left arm. The patient's medical records indicated TA. Magnetic resonance imaging (MRI) angiography and peripheral angiography, performed in 2008, showed stenosis in the right subclavian artery and total occlusion in the left subclavian artery (Figure 1). In February 2008, she had undergone a left subclavian-axillary artery bypass with a saphenous vein graft. Further tests revealed fragmented red blood cells. After this work-up, we initiated treatment with nitroglycerin for controlling the BP and magnesium sulphate for prevent of convulsions. Considering these findings, an emergency cesarean section was planned. The patient underwent a low transverse cesarean section, and fetuses weighing 300 g, 310 g, and 260 g were delivered without complications.

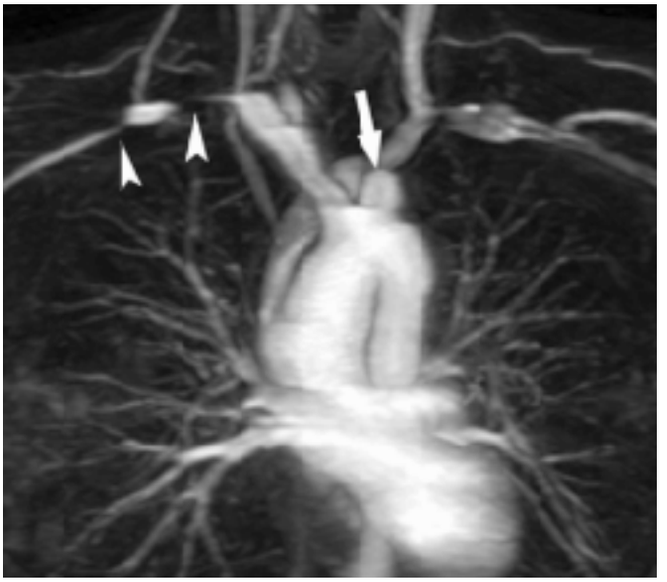

Figure 1: Shows two stenotic segments of the right subclavian artery (small arrows) and occlusion of the left subclavian artery (long arrow) in MR angiography

In the fifth postoperative hour, the patient reported acute onset palpitations and abdominal pain. Her BP was 100/50 mm Hg; pulse rate, 140 beats/min; oxygen saturation, 98% in room air; hemoglobin level, 7 g/dL; and hematocrit level, 22%. An abdominal ultrasonogram revealed extensive areas of hematoma, and the patient's vital signs were worsening. Therefore, bleeding from the incision site was suspected, and a relaparotomy was performed. Hematomas were observed on the rectus sheath, but an active bleeding point was not discovered during the operation. After the surgical procedure, the patient was transferred to the intensive care unit, where she remained for 5 days without complications. Postoperative recovery was good.

Discussion

TA is a potentially life-threatening condition during pregnancy, with a worldwide maternal mortality rate of 4-5% (130/100.000 in the general population) 6 and the highest incidence in Japan, East and South Asia, and India.[7] Various hypotheses have been proposed regarding its etiopathogenesis, including an association with rheumatic disease, an autoimmune mechanism, antiaorta antibodies, HLA antigens, and tuberculosis4. Most cases of TA during pregnancy have been reported in patients with a known prenatal diagnosis. In studies by Wong et al. and Suri et al., 53% and 60% of the patients, respectively, were diagnosed with TA for the first time during pregnancy.[3,8] In the present case, the patient was diagnosed prenatally, but detailed medical records were not available on first admission.

In most series, the majority of patients with TA present with absent or unequal peripheral pulses and/or hypertension or its complications.[3,4,9] Women in the Ishikawa group IIb and III have worse fetomaternal outcome,[3] but the present patient was Ishikawa group IIa. In the series reported by Suri et al. and Sharma et al.[3,9] abdominal aorta involvement was maximal, followed by the subclavian and renal arteries. Here, the patient had bilateral subclavian artery involvement but no renal artery involvement.

Matsumura et al.[10] studied levels of C-reactive protein in 16 patients with TA to evaluate the inflammatory activity of the disease 1 year before, during, and after pregnancy. The authors showed an improvement of inflammatory activity and hemodynamic findings during pregnancy. In the present case, the patient had normal C-reactive protein level, which is associated with a favorable pregnancy outcome.[3,9] In the present case, the outcome was surgical delivery by C-section. Despite the mode of delivery being vaginal, curtailing the second stage of labor has been recommended in some studies. The indications of operative interference were intrauterine growth retardation pre-eclampsia, eclampsia, and an inability to measure

BP in both arms. A C-section was performed in the present case because of the presence of HELLP syndrome with progressive disease. In the present case, there was no evidence of congestive heart failure, progression of renal insufficiency, or antepartum hemorrhage related to accelerated hypertension, but rectus hematoma requiring further surgery was evident and was presumably caused by thrombocytopenia.

Medical management of a pregnant patient with TA does not differ significantly from that of a non-pregnant patient. The HELLP syndrome occurs in about 0.5-0.9% of all pregnancies and in 10-20% of cases with severe preeclampsia.[11] The onset of the HELLP syndrome is usually rapid, and cases that develop before 20 weeks gestation are considered atypical, as was the case in the current report. Cases of TA complicated by HELLP syndrome have not been reported previously. This case report attempts to illustrate the clinician's dilemma in diagnosing an atypical presentation of HELLP syndrome with TA.

In the literature, there is debate about the effect of pregnancy on TA, but pregnant patients with TA appear to have significant risks for pregnancy-induced hypertension, preeclampsia, postpartum hemorrhage, and intrauterine growth restriction. Here, TA may have resulted in uncontrollable hypertension, severe preeclampsia, and atypical HELLP syndrome. Few data are available on the relationship between a triplet pregnancy and hypertension. To the best of our knowledge, preeclampsia is common in high-order multifetal gestations and often presents in an atypical manner.

In summary, we described the case of a patient with TA who developed atypical HELLP syndrome that resulted in maternal morbidity and perinatal mortality. Based on the results of this case, we suggest that patients with TA syndrome should be informed of the potential risks before conceiving a child. In addition, they should be carefully monitored during the perinatal period by collaboration between the rheumatology and obstetrics departments. Strict control of blood pressure during pregnancy and regular antenatal care is essential for an optimal outcome.

Takayasu Arteritli Subklavyen Arter - Aksiller Arter Safen Ven Grefti Olan Atipik Help Sendromlu Gebe: Olgu Sunumu

Takayasu arteriti (TA) aort ve ana dallarından sıklıkla vertebral, karotid, subklavyan, iliak ve renal arterleri tutan nadir görülen kronik granülomatöz bir hastalıktır. Hastalık sıklıkla reprödüktif dönemdeki kadınları etkiler ve bu yaşlardaki kadınlarda en azından bir gebelik meydana gelir. Bu nedenle hastalıklı kadınlarda gebeliğin yönetimi obstetrisyenler açısından büyük önem taşır. Biz yazımızda atipik hemoliz, yükselmiş karaciğer enzimleri ve düşük platelet sayısına sahip (HELP sendromu) kortikosteroid tedavisi alan ve daha önce subklavyen-aksiller arter safen ven grefti olan bir olguyu sunduk.

Anahtar Kelimeler: Help sendromu, Takayasu arteriti, Ven baypas greft

References

1. Maksimowicz-McKinnon K, Clark TM, Hoffman GS. Takayasu arteritis and giant cell arteritis: a spectrum within the same disease? Medicine (Baltimore) 2009;88: 221-6.
2. Seo P. Pregnancy and vasculitis. Rheum Dis Clin N Am 2007;33:299-317.
3. Suri V, Aggarwal N, Keepanasseril A, Chopra S, Vijayvergiya R, Jain S. Pregnancy and Takayasu arteritis: a single centre experience from North India. J Obstet Gynaecol Res 2010;36:519-24.
4. Mandal D, Mandal S, Dattaray C, et al. Takayasu arteritis in pregnancy: an analysis from eastern India. Arch Gynecol Obstet 2011;285:567-71.
5. Manika A, Ananya D. Pregnancy in Takayasu's arteritis with bilateral renal artery and abdominal aorta stents. Aust N Z J Obstet Gynaecol 2010;50:203-4.
6. Mwipatayi, BP, Jeffery PC, Beningfield SJ, et al. Takayasu arteritis: clinical features and management: report of 272 cases. ANZ J Surg 2005;75:110-7.
7. Johnstone SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. J Clin Pathol 2002;55:481-6.
8. Wong VC, Wang RY, Tse TF. Pregnancy and Takayasu's arteritis. Am J Med 1983;75:597-601.
9. Sharma BK, Jain S, Vasishta K. Outcome of pregnancy in Takayasu arteritis. Int J Cardiol 2000;75:159-62.
10. Matsumura A, Moriwaki R, Numano F. Pregnancy in Takayasu arteritis from the view of internal medicine. Heart Vessels Suppl 1992;7:120-4.
11. Karumanchi SA, Maynard SE, Stillman IE, Epstein FH, Sukhatme VP. Preeclampsia: a renal perspective. Kidney Int 2005;67:2101-13.