

Successful Term Delivery Following Ruptured Sinus of Valsalva Aneurysm Repair at 29 Weeks of Pregnancy

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Abstract

Ruptured sinus of Valsalva aneurysm is a very rare cardiac condition occurring in pregnancy. A sinus of Valsalva aneurysm is thought to be caused by deficient fusion of the media of the aorta with annulus fibrosus of the aortic valve. The incidence of sinus of Valsalva aneurysm is less than 1% of all congenital cardiac anomalies. Though ruptured sinus of Valsalva aneurysm is rare, this condition when confronted can cause great difficulty in management. There is little discussion in literature regarding this condition in pregnant patients. Documented cases of ruptured sinus of Valsalva aneurysm have either presented at term or as a previously diagnosed congenital defect which did not require immediate surgical intervention. We describe a case which required surgical intervention at 29 weeks with the pregnancy continued till term.

Keywords: sinus of Valsalva aneurysm, ruptured sinus of Valsalva aneurysm in pregnancy, cardiopulmonary by-pass in pregnancy

Özet

Gebeliğin 29. Haftasında Valsalva Sinüs Anevrizma Rüptürü Tamiri Olan Bir Hastada Başarılı Term Doğum

Valsalva sinüs anevrizma rüptürü gebelikte görülebilen, oldukça nadir bir kardiyak durumdur. Valsalva sinüs anevrizmasının, aortadaki medial tabakanın aortik kapaklardaki annulus fibrozuslara olan yetersiz bağlantısından oluştuğu düşünülmektedir. Valsalva sinüs anevrizması tüm konjenital kardiyak anomalilerin %1'inden azını oluşturur. Valsalva sinüs anevrizması rüptürü nadir görülen bir durum olmakla birlikte, bu sorunla karşılaşıldığında tedavide büyük bir güçlük ortaya çıkabilir. Burada, gebe bir hastada bu durumun değerlendirilmesiyle ilgili literatür desteği ile kısa bir tartışma yapılmıştır. Literatürde, Valsalva sinüs anevrizma rüptürü vakaları, ya miad gebeliklerde ya da önceden tanı almış ve acil cerrahi girişim gerektirmeyen konjenital defektler olarak bildirilmiştir. Burada, 29. gebelik haftasında acil cerrahi müdahale gerektiren ve sonra gebeliği terme kadar devam eden bir vaka bildirilmektedir.

Anahtar sözcükler: Valsalva sinüs anevrizması, gebelikte rüptüre olmuş Valsalva sinüs anevrizması, gebelikte kardiyopulmoner by-pass

Case

Ruptured sinus of Valsalva aneurysm is a very rare cardiac condition occurring in pregnancy. A sinus of Valsalva aneurysm is thought to be caused by deficient fusion of the media of the aorta with annulus fibrosus of the aortic valve. The incidence of sinus of Valsalva aneurysm is less than 1% of all congenital

cardiac anomalies (1). Though ruptured sinus of Valsalva aneurysm is rare, this condition when confronted can cause great difficulty in management. There is little discussion in literature regarding this condition in pregnant patients. Documented cases of ruptured sinus of Valsalva aneurysm have either presented at term or as a previously diagnosed congenital defect which did not require immediate surgical intervention (2). We describe a case which required surgical intervention at 29 weeks with the pregnancy continued till term.

A 29-year primigravida at 29 weeks of gestation presented with acute onset of symptoms of cardiac failure for 6 hours. Her

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pregnancy had been uneventful till then. She had no prior significant medical or cardiac illness. Examination revealed sinus tachycardia of 122 per minute, blood pressure of 128/58 mmHg, continuous machinery murmur over the entire precardium, uterus 28 weeks size and foetal heart rate of 140 per minute. Initial blood investigations were within normal limits. Electrocardiogram showed sinus tachycardia. Echocardiography showed evidence of aneurysm of right coronary sinus with opening into right ventricle and right atrium, continuous flow from coronary sinus to right atrium, dilated aortic root, signs of hyperdynamic circulation and structurally normal other valves – suggestive of ruptured sinus of Valsalva aneurysm (Figure 1).

She was stabilised with decongestive therapy and treated conservatively. On fifth day following admission, she went into pulmonary oedema which necessitated the patient to be put on a ventilator. Ruptured sinus of Valsalva aneurysm repair was planned and obstetricians were consulted regarding the pregnancy management. After explaining to the patient and the relatives the risks to the mother and foetus, decision was taken to continue pregnancy.

Ruptured sinus of Valsalva aneurysm repair under cardiopulmonary bypass was undertaken. Intraoperatively there was dilated right atrium, right ventricle and left pulmonary artery, large sinus of Valsalva aneurysm rupture into right ventricle, no ventricular septal defect and interatrial septum. Sinus of Valsalva aneurysm rupture into right ventricle was closed with Dacron patch. Patient was not cooled during surgery. Postoperative period was uneventful. Repeat echocardiography showed no residual shunt. On day ten she was discharged on Digoxin and Frusemide after confirming foetal wellbeing. She had regular antenatal follow ups every 15 days and was continued on Digoxin and Frusemide.

She was admitted at 37 weeks for elective caesarean section in view of IUGR with oligamnios. However, there was no obvious cause detected for IUGR. Her general condition was fair with no residual symptoms. She delivered a 2 kg male baby with APGAR score of 8/10 and 9/10 at first and fifth minute respectively by elective caesarean under spinal anaesthesia. There was no intrapartum complication. Mother had uneventful postpartum period while neonate received phototherapy for neonatal jaundice for 4 days. Both mother and baby were discharged on 10th postoperative day. The mother was continued on Digoxin and Frusemide and advised to consult her cardiologist one month later.

Discussion

Sinus of Valsalva aneurysm is a very rare cardiac anatomic abnormality resulting from defect in the continuity between aortic media and annulus fibrosis. It is mostly congenital and may be associated with other structural defects. It can also be due to underlying connective tissue disorders or as a result of damage to cardiac tissue surrounding the sinus of Valsalva following inflammation or trauma. Patients are usually asymptomatic until the 3rd or 4th decade when it ruptures into

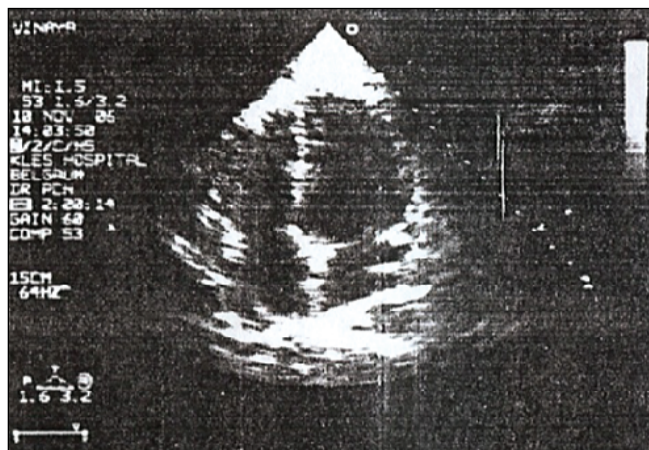


Figure 1. Echocardiography showing right and left ventricles and the aneurysmal dilatation of the aortic root.

any of the four cardiac chambers and produces symptoms of failure, arrest or tamponade (3). Clinical diagnosis can be easily made by the presence of continuous murmur and confirmed by echocardiography, computed tomographic angiography or magnetic resonance imaging.

All cases which have been reported so far usually presented at term or after previous diagnosis of aneurysm without clinical deterioration in the patient's condition. Surgical intervention was required 1 week postpartum in two cases (2,4). Our case of ruptured sinus of Valsalva aneurysm was not associated with previously known cardiac disease and presented for the first time at 29 weeks of gestation posing dual problem for the management. Clinical deterioration of the patient in the presence of hyperdynamic state questioned continuation of pregnancy, while termination of pregnancy could have resulted in preterm delivery. However pregnancy was continued after discussing with the relatives the risks of preterm birth and continuation of pregnancy during cardiopulmonary bypass which is known to be associated with increased maternal and foetal mortality. The patient underwent a successful ruptured sinus of Valsalva aneurysm repair and pregnancy was continued until week 37 under supervision. Caesarean section was carried out for IUGR with oligamnios. The patient could have had a normal vaginal delivery, as there had been no obstetric indication for caesarean section and there was no residual shunt. This is probably the first case of ruptured sinus of Valsalva aneurysm repair during pregnancy with good maternal and foetal outcome.

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