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Case report

Aortic diseases in pregnancy : clinical case

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Abstract

Introduction: Several hereditary disorders affect the thoracic aorta, predisposing patients with both aneurysm and aortic dissection. Pregnancy in patients with Marfan's syndrome carries an increased risk of cardiovascular complications, resulting in increased maternal and fetal mortality and morbidity.
Case report: We present a 36-year-old woman with Marfan's syndrome at the 10th week of pregnancy, who at the age of 24 had mechanical aortic valve implantation due to bicuspid aortic valve and aortic regurgitation. On a routine cardiac control transthoracic echocardiography, we discovered an aneurysm of ascending aorta with dissection. Mechanical valve on aortic position was well-functional. There was also prolapse of the mitral valve with 3+ mitral regurgitation, increased dimensions of the left atrium and left ventricle with a preserved ejection fraction. Patient had positive family history for aortic disection.
Conclusion: As aortic disease is reported as one of the leading causes of maternal mortality, it should be emphasized that pregnancy is a high risk period for all patients with aortic pathology.

Key words Marfan's syndrom, aortic dissection, pregnancy

Introduction

During pregnancy, women with Marfan's syndrome have serious risk for acute aortic dissection, around 3%[1], even with aortic root diameter < 40 mm. Mortality rates are high both for the mother and the fetus[2]. The cooperation of obstetrics and cardiologists in prenatal counseling and management of women with Marfan's syndrome is of outmost importance and it is advised that assessment and evaluation of adverse cardiac events should be done individually, because of the significant variations of the phenotypic expression[2].

Case report

A 26-year-old woman with Marfan's syndrome at the 10th week of pregnancy came to the routine cardiac control.

At the age of 24 she was operated for bicuspid aortic valve and aortic regurgitation and mechanical aortic valve was implanted. In family history, her father had aortic dissection.

Transthoracic echocardiography revealed well-functional mechanical valve. However, aneurysm of the ascending aorta with dissection was detected. There was also a prolapse of the mitral valve with 3+ mitral regurgitation and increased left atrium. Left ventricle was significantly enlarged, still with preserved ejection fraction (Figures 1,2,3,4 Table 1.).

The patient was advised to interrupt pregnancy and to perform surgery of the aorta and mitral valve.

The pregnancy was interupted and two months later she was operated. Resection of ascendent part of the aorta with Dacron graft and mechanical mitral valve implantation were done.

At the age of 31, she had a second pregnancy, that was performed under the strict supervision of cardiologist and obstetrician. The patient was given birth by cesarean section at eight and a half months' gestation to a healthy female infant.

Discusion

Several hereditary disorders affect the thoracic aorta, predisposing patients with both aneurysm and aortic dissection. Pregnancy is a high risk period for all patients with aortic pathology. Investigations have identified estrogen and relaxin as the most likely causes of the diseased aortic wall. Hormonal and haemodynamic changes during pregnancy increase the chance of

Table1. Echocardiographic parameters : Ao –aorta, LA- left atrium, LV-left ventricle, LV EDD-end-diastole dimension of the left ventricle, LVSD-end-systole dimension of the left ventricle, SEP- septum, PW- posterior wall, EF- ejection fraction, AR - aortic regurgitation, MR- mitral regurgitation.

Ao: 3.7 -5.0cm 7.5x6.5	**LA 4.8 cm**	**LV DD 6.4 cm**	**LV SD 4.0 cm**	**SEP 1.0cm**	**PW 0.9 cm**	**EF 65%**	**AR 1+**	**MR 3+**	**RV**	**TR**

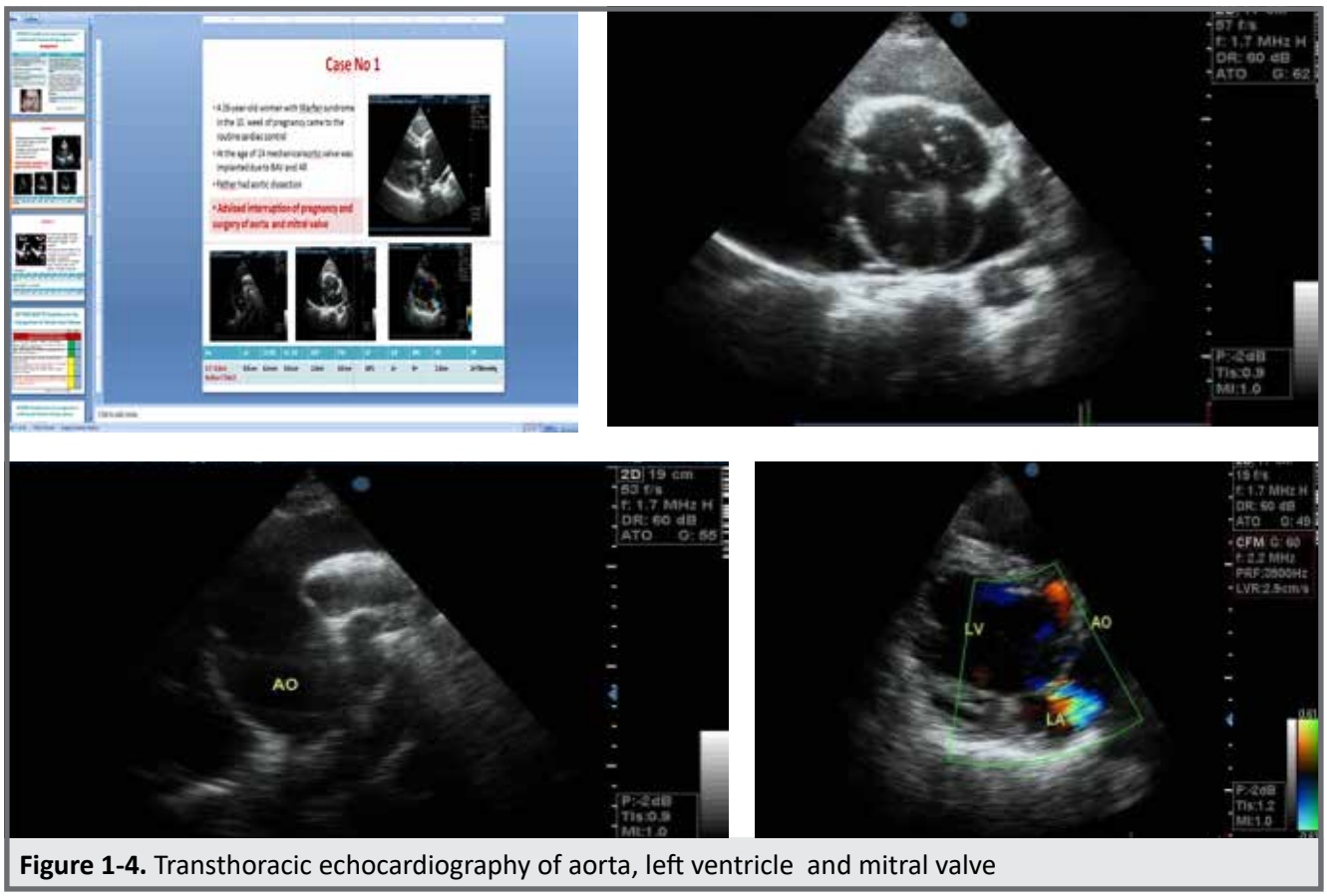

Figure 1-4. Transthoracic echocardiography of aorta, left ventricle and mitral valve

Table 2. Recommendations for the management of aortic disease during pregnancy

All aortic diseases	
Recommendations	**Class[a]/ Level[b]**
It is recommended that women with aortic disease have counselling about the risk of aortic dissection	I C
Imaging of the entire aorta (CT/MRI) is recommended before pregnancy in patients with a genetically proven aortic syndrome or known aortic disease	I C
In BAV patients imaging of the ascending aorta is recommended	I C
In pregnant women with known aortic dilatation, (history of) type B dissection or genetic predisposition for dissection, strict blood pressure control is recommended	I C
Repeated echocardiographic imaging every 4–12 weeks is recommended during pregnancy in patients with ascending aorta dilatation	I C
For imaging of pregnant women with dilatation of distal ascending aorta, aortic arch or descending aorta, MRI (without gadolinium) is recommended	I C
It is recommended to deliver all women with aortic dilatation or (history of) aortic dissection in an experienced centre with a pregnancy heart team, where cardiothoracic surgery is available.	I C
In patients with an ascending aorta <40 mm vaginal delivery is recommended	I C
In patients with an ascending aorta >45 mm caesarean delivery should be considered.	IIa C
Prophylactic surgery should be considered during pregnancy if the aortic diameter is ≥50 mm and increasing rapidly.	IIa C
In patients with an aorta 40–45 mm vaginal delivery with epidural anaesthesia and expedited second stage should be considered.	IIa C
In patients with an aorta 40–45 mm caesarean section may be considered.	IIb C
It is recommended to avoid pregnancy in patients with (or history of) aortic dissection.	III C
Specific syndromes	
β-blocker therapy throughout pregnancy should be considered in women with Marfan syndrome.	IIa C
It is recommended to avoid pregnancy in patients with severe dilatation of the aorta (Marfan syndrome >45 mm, Loeys–Dietz syndrome >45 mm, BAV >50 mm (or >27 mm/m2 body surface area), Turner syndrome ASI>25 mm/m2 body surface area).	III C
It is recommended to avoid pregnancy in patients with vascular Ehlers–Danlos syndrome.	III C

ASI = aortic size index; BAV = bicuspid aortic valve; CT = computed tomography; MRI = magnetic resonance imaging. a Class of recommendation. b Level of evidence.

the dissection. Dissection occurs most often in the last trimester of pregnancy (50%) or the early post-partum period (33%). All women with a genetically proven syndrome should have counseling on the risk of dissection.

Diagnosis and treatment of our patient was guided according to the recommendations for treating aortic diseases during pregnancy (Table 2.)[3].

Conclusion

In conclusion, it should be emphasized that pregnancy is a high risk period for all patients with aortic pathology, because the aortic disease is reported as one of the leading causes of maternal mortality.

References

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Sažetak

Uvod: *Poznato je nekoliko naslednih oboljenja koja dovode do izmene strukture vezivnog tkiva torakalne aorte i čine ih podložnijim razvoju aneurizme i disekcije aorte. Trudnoća u pacijentkinja sa Marfanovim sindromom nosi visok rizik od kardiovaskularnih komplikacija, rezultujući povećanim maternalnim i fetalnim morbiditetom i mortalitetom.*

Prikaz slučaja: *Prikazujemo slučaj 36-godišnje pacijentkinje sa Marfanovim sindromom u 10. nedelji trudnoće, kojoj je prethodno u 24. godini života implantirana veštačka mehanička valvula na terenu bivelarne aortne valvule i značajne aortne regurgitacije. Rutinski kontrolni transtorakalni ehokardiografski pregled otkriva aneurizmu ascendentne aorte sa disekcijom, uz dobro funkcionalnu veštačku mehaničku valvulu na aortnoj poziciji. Takođe, uočava se prolaps mitralne valvule sa 3+ mitralnom regurgitacijom, povećane dimenzije leve pretkomore, kao i leve komore, uz očuvanu sistolnu funkiju. Anamnestički, saznaje se da je porodična anamneza pacijentkinje pozitivna za disekciju aorte.*

Zaključak: *Kako su bolesti aorte prepoznate kao jedan od glavnih uzroka maternalnog mortaliteta, neophodno je istaći da je trudnoća visoko rizična kod svih pacijentkinja sa patologijom aorte.*

Ključne reči: *Marfanov sindrom, disekcija aorte, trudnoća*