Case Reports

Peripartum cardiomyopathy and Klippel-Trenaunay syndrome

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Summary

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder of unknown etiology characterized by venous malformations or varicose veins, cutaneous capillary malformation and hypertrophy of soft tissues with limb (usually asymmetric lower extremity) involvement. Peripartum cardiomyopathy (PPCM) is characterized by rapid onset heart failure during the final month of pregnancy or within five months of delivery, in the absence of identifiable risk factors or previous heart disease. The aim of this study was to illustrate the correlation between the KTS and the onset of PPCM in women with twin pregnancies. Our case is a 35-year-old woman, gravida II para I, with KTS, twin pregnancy and PPCM. We can assume that, as the heart of a women with KTS usually works with a low preload reserve due to the widespread venous varicosities, if a significant increase in preload occurs, it may lead to the onset of cardiac dilatation and thus PPCM.

Key words: : Peripartum cardiomyopathy; Klippel-Trenaunay Syndrome; Twin pregnancy.

Introduction

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder characterized by a specific symptom triad: cutaneous capillary malformations, venous malformations or varicose veins, hypertrophy of soft tissues [1-4] with limb (usually asymmetric lower extremity) involvement. The first description of this syndrome was reported in 1900 [5] but the molecular mechanism remains unknown and realistically multiple genetic factors are involved [6]. KTS is uncommon and no consensus yet exists on the correct approach to its investigation and treatment. The syndrome may present local and systemic complications (Table 1). A tendency of KTS to recurrent thromboembolic events is well known with a reported incidence of up to 22% [7]. Peripartum or postpartum cardiomyopathy (PPCM) is characterized by rapid onset heart failure during the final month of pregnancy or within five months of delivery in the absence of identifiable risk factors or previous heart disease [8]. The diagnosis of PPCM is made according to criteria provided by the World Health Organization/International Society and Federation of Cardiology [9], the Guidelines of the National Heart, Lung, and Blood Institute Workshop on the "Prevalence and the Etiology of Idiopathic Dilated Cardiomyopathy" [10] and the more recent Guidelines for the "Study of Familial Dilated Cardiomyopathies" [11], designed to improve the sensitivity and specificity of the old classification criteria (Table 2). Early signs and symptoms of heart failure can be obscured by pregnancy, because often the patient considers them to be normal. The incidence of peripartum cardiomyopathy ranges from one in 1,300 to one in 15,000 pregnancies. The mortality rate of peripartum cardiomyopathy is 30-60% and death may be caused by severe pulmonary congestion [12]. Etiology of PPCM is unknown [13-15] and several hypotheses have been proposed: myocarditis, viral infection, autoimmune factors, inflammatory cytokines, and abnormal hemodynamic response to physiological changes in pregnancy. Common reported risk factors for PPCM are advanced maternal age, multiparity, multiple gestations, black race, obesity, malnutrition, gestational hypertension, preeclampsia, poor antenatal care, breast feeding, cesarean section, alcohol, cocaine and tabacco abuse, low socio-economic conditions and family history [15, 16]. No strong hereditary association has been identified [16].

Case Report

The patient was a 35-year-old woman, gravida 2 para 1, with KTS syndrome, twin pregnancy and peripartum cardiomyopathy. No prenatal or obstetric complications occurred during her prior pregnancy and delivery. There was no history of heart disease, excessive alcohol consumption, recent viral infection, hypertension, diabetes mellitus, or preeclampsia. The patient came to our observation at the 34th week of gestation with early labor contractions. She had elephantiasis of the lower left limb (Figure 1), cutaneous capillary hemangioma (Figure 2) spread all over the body surface and vulval varicosities (Figure 3). Dyspnea and asthenia (NYHA class II) occurred at the 32nd week of gestation. A prompt vascular examination was carried out and a compression bandage was applied to the lower limbs. Administration of subcutaneous heparin (Clexane 4000 UI) for prophylaxis of deep vein thrombosis (TVP) was initiated. Due to the vulval varicosities, the elephantiasis of the lower right limb and the unfavorable cervix a cesarean section was performed. The operation was carried out without complications and the fetal outcome was good. Both twins' Apgar scores were 8-9. Two hours later the patient showed symptoms of increasing dyspnea and chest pain. ECG, chest X-ray, total body computed tomography (CT), with and without contrast agent, and

Revised manuscript accepted for publication February 2, 2010

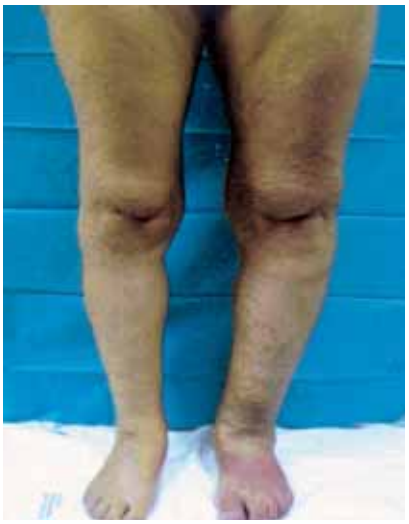

Figure 1. — Hypertrophy of lower left limb.

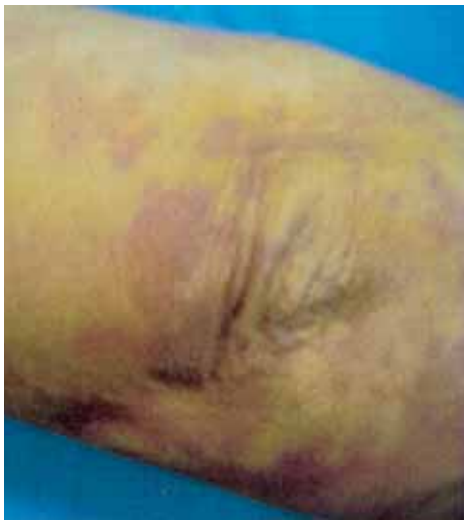

Figure 2. — Cutaneous capillary malformation.

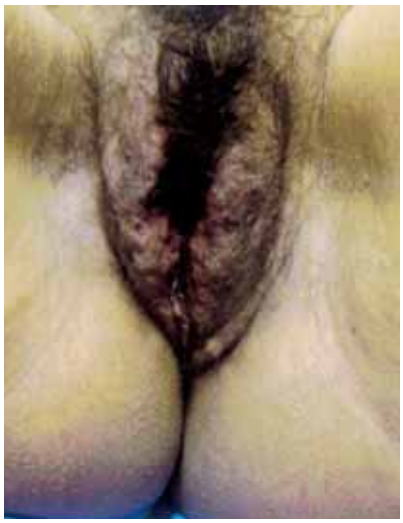

Figure 3. — Vulvar varicose veins.

echocardiogram were performed, assuming the onset of an acute pulmonary embolism. The electrocardiogram showed sinus rhythm with short P-R and nonspecific ST-T abnormalities. The chest X-ray showed pulmonary edema and cardiomegaly. A filling defect in the areas of the main and peripheral pulmonary arteries was reported by CT with bilateral pleural effusion, more marked on the right, and thickened areas in the lower lung most probably referring to acute pulmonary edema. Echocardiography demonstrated a 35-40% left ventricular ejection fraction, a 25.4% fractional shortening and a 5.9 cm/m^2 left ventricular end-diastolic dimension with signs of congestive heart failure (CHF). A mild mitral regurgitation also occurred. Data were consistent with peripartum cardiomyopathy. As a result, the patient was transferred to the Intensive Care Unit. On admission to the emergency department the patient presented with mild respiratory distress, with oxygen saturation of 84% on room air, heart rate of 84 bpm and blood pressure 100/50 mmHg. Her laboratory evaluation, including a complete blood count and chemistries, were remarkable only for a elevated white blood cell count of 19.4. D-dimer was elevated (5052). She was treated with furosemide, angiotensin converting enzyme (ACE) inhibitor (ramipril), low molecular weight heparin 4000 UI and potassium canrenoate. Seven days later the electrocardiogram showed an improvement of the pathology. Left ejection fraction was 59%, fractional shortening 46% and left ventricular end-diastolic dimension 5.0 cm/m^2. The patient did well for the following six months and returned to active employment. Subsequent investigations showed a normal echocardiographic pattern.

Discussion

Pregnancy has rarely been reported in patients with KTS and since 1989 there have been only 13 cases of pregnant women with KTS reported in the literature [17]. Maternal and fetal risks associated with pregnancy in women with KTS are proportional to the severity of the disease, which can be exacerbated by pregnancy. An association between KTS and fetal growth restriction has been reported, maybe due to placental insufficiency caused by angiomatosis related to the syndrome [18]. Complications that may arise in pregnancy in patients with KTS include bleeding, DIC, thromboembolic events and pain. The occurrence of pulmonary embolism has also been reported [19] so careful monitoring of coagulopathic disorders is advisable [20]. However, literature data do not report an increased risk of peripartum cardiomyopathy in patients with KTS. We can assume that, as the heart of a women with KTS usually works with a low preload reserve due to the widespread venous varicosities, if a significant increase in preload occurs, this may lead to the onset of cardiac dilatation and thus peripartum cardiomyopathy. In our case the twin pregnancy may have played a determinant role.

Table 1. — *KTS: complications*.

KTS: complications local
– Pain
– Cellulitis
– Ulceration
– Thrombophlebitis
– Gangrene
Involvement of internal organs
– Neurovascular anomalies
– Pulmunary vein varicosities
– Pleuro-pericardial effusions
– Pulmonary embolism
– Colorectal and urinary tract
– Hemorrhage
Systemic
– Consumptive coagulopathy high-output cardiac failure

Table 2. — *PPCM: diagnostic criteria*.

Demonstrable echocardiographic criteria of left ventricular dysfunction
– Ejection fraction < 45%
– Left ventricular fractional shortening < 30%
– Left ventricular end-diastolic dimension > 2.7 cm/m^2 body surface area

Conclusion

Although the KTS was previously considered a contraindication to pregnancy, close monitoring can greatly improve the maternal and fetal outcome. Regardless of adeguate preconception counselling, systematic clinical and instrumental monitoring is recommended and a close collaboration between the obstetrician and the cardiologist is mandatory to reduce morbidity and mortality related to the onset of peripartum cardiomyopathy.

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