Aortic and pulmonary artery calcification: An unusual manifestation of twin-to-twin transfusion syndrome

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ABSTRACT
Twin-to-twin transfusion syndrome (TTTS) at times complicates monochorionic twin gestations, resulting in conditions ranging from discordant sizes to fetal demise of one baby. Various types of cardiac defects have been described in the recipient twin of this syndrome. Isolated great artery calcification, i.e. aortic and pulmonary artery calcification is one such uncommon condition associated with TTTS. Calcification of the walls of great vessels may be due to chronic vascular injury sustained as a result of circulatory volume overload in the recipient twin. It may also cause severe systemic hypertension and cardiomyopathy. An accurate diagnosis is important for an optimal follow-up and appropriate genetic counseling. We report a case of aortic and pulmonary artery calcification in association with TTTS.

Keywords: Aortic calcification, pulmonary artery calcification, twin-to-twin transfusion syndrome

INTRODUCTION
Twin-to-twin transfusion syndrome (TTTS) is a rare disorder occurring as a result of communicating vascular anastomosis between the circulations of one twin with that of the other. The incidence of TTTS is 4–26% in monochorionic twins. Cardiac findings in this condition are varied and may include ventricular hypertrophy, pulmonary stenosis, tricuspid regurgitation, congestive cardiac failure, left ventricle hypoplasia with hypokinesia, and subaortic obstruction seen in the recipient twin.[1–4]

CASE REPORT
The mother was a 24-year-old primigravida with twin gestation (monochorionic diamniotic) of spontaneous conception. An ultrasound examination at 20 weeks of gestation showed TTTS, and hence, laser photocoagulation of placental vascular anastomoses was done. Postprocedure, the donor fetus developed hydrops fetalis and the recipient fetus showed a high middle cerebral artery peak systolic velocity. Subsequently, the donor fetus showed ventriculomegaly on neurosonography though hydrops had significantly reduced. Both twins developed polyhydramnios possibly giving the diagnosis of twin reversed arterial perfusion sequence. Bipolar cord occlusion of the donor twin was done as per the parents’ decision. Thereafter, the recipient twin showed calcifications in ascending aorta and proximal portion of the pulmonary artery with high-velocity flow in both vessels on fetal ultrasonography. Pericardial effusion was consistent and persisted throughout the entire gestation. Fetal echocardiography of the recipient twin showed hyperechogenicity in the root of the pulmonary artery and aorta, excluding the valves and extending beyond the sinotubular junction in the aorta. The antegrade flow was seen across pulmonary artery and aorta with high velocity, and also calcifications were noted in the distal descending aorta. Institutional delivery was advised and regular monitoring was carried out till delivery. Baby was delivered by cesarean section at a gestational age of 32.1 weeks, with the weight of 1840 g.

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Venkatesh, et al.: Great vessels calcification

Serum calcium was 9.4 g/dl with ionic calcium of 1.18 mmol/L. Chest X-ray was unremarkable. Neonatal echocardiography was performed in the Neonatal Intensive Care Unit (NICU), which showed calcification of the main pulmonary artery and descending thoracic aorta with a maximum gradient of 10 mm Hg, a small patent ductus arteriosus, and a patent foramen ovale. Serial echoes showed no increase in the flow velocities across the great vessels. Cardiac computed tomography showed calcification of the great vessels (both aorta and pulmonary artery). There were no calcifications elsewhere in the body.

Genetic studies for Idiopathic Arterial Calcification of Infancy (IACI) were negative. Initially, bisphosphonate was planned as therapy but was withheld in view of no progression of calcification or deterioration of clinical status. The neonate was closely monitored in the NICU and discharged with a good weight gain on day 25 of life. The baby has been asymptomatic and follows-up regularly with blood pressure monitoring. Following Figures 1 and 2 shows calcifications in descending aorta and pulmonary artery while Figure 3 shows cardiomegaly in fetal heart.

**DISCUSSION**

TTTS is a rare, nonhereditary entity with a worldwide incidence of 8–12% and a poor fetal prognosis when untreated. Neonatal mortality reduces to 20–30% on serial amnioreduction in such cases. This procedure reduces premature labor and thereby the mortality rate.

Events leading to TTTS in pregnancy include the time of twinning, sharing of the placenta, and connecting vessels by the twins.

The reported cardiac complications seen in TTTS include ventricular hypertrophy, right ventricular outflow tract obstruction (pulmonary stenosis), tricuspid regurgitation, congestive cardiac failure, left ventricle hypoplasia with hypokinesia, and subaortic obstruction, with incidence being more in the recipient twin. No definitive therapy and a high mortality rate have been associated with this condition in most of the case reports mentioned in literature.

Great vessel calcification represents a clinical spectrum, in which there is calcification of large- and medium-sized blood vessels. Vascular injury as a result of excessive volume overload can cause calcification of the great vessels and so are seen more in the recipient twin. Calcification can occur in either systemic or pulmonary vessels, as there is no pressure difference between the two during fetal life. According to the study by Zosmer et al., volume overload and resistance to flow through umbilical artery cause an increase in both preload and afterload leading to cardiac dysfunction in the recipient twin. The various substances released from placenta such
as renin, troponin, and endothelin are also responsible for the cardiac dysfunction.[1,3]

IACI is the only differential diagnosis for the condition which is a rare, autosomal recessive disorder where widespread calcifications occur in large- and medium-sized blood vessels such as aorta, pulmonary vessels, coronary arteries, renal arteries, and extravascular sites such as adipose tissues, kidneys, and periarticular region.[8]

Genetic study in the form of mutation analysis is the only investigation for confirmation. The study by Hamazaki[9] mentions that idiopathic calcification of infancy is an elastic fiber defect where calcification occurs in the internal elastic lamina, followed by the calcium encrustation. Materials that stain with mucopolysaccharides accumulate around the elastic fibers. Deposition of hydroxyapatite and smooth muscle cell proliferation occurs in association with periarticular calcification.

Karatza et al.[10] found that there was no hemodynamic difference in monochorionic and diamniotic twins. Aortic and pulmonary velocities were more in recipient twin as compared to donor twin. Furthermore, Samon et al.[11] mention that idiopathic calcification of the great vessels occurs in the recipient twin in a case of TTTS, as in our case.

CONCLUSION

Calcification of the great vessels is seen due to excessive volume overload in the recipient twin in cases of TTTS. Antenatal ultrasonography is useful to identify hyperechogenicity of vessel walls or polyhydramnios in cases of multiple gestation which aids early recognition of the condition. Serial monitoring during pregnancy and postnatal life is imperative to reduce morbidity and mortality associated with this syndrome.

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Conflicts of interest

There are no conflicts of interest.

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