Prenatal sonographic diagnosis of left pulmonary artery sling

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Objective

Firstly to report a rare case of early prenatal diagnosis of fetal left pulmonary artery sling, secondly to highlight the importance of early prenatal diagnosis of this rare anomaly and thirdly to define the ultrasound markers for prenatal diagnosis.

Methods

1. Describe the prenatal clinical and ultrasound findings of a case of fetal left pulmonary artery sling. 2. Compare the findings with a previous prenatally diagnosed case to look for ultrasound clue for making a prenatal diagnosis.

Results

Left pulmonary artery sling (LPAS) is a very rare anomaly in which the left pulmonary artery (LPA) arises and courses aberrantly and forms a sling around the trachea, causing distal airway obstruction by mechanical compression. But severe obstruction is more commonly due to the associated intrinsic tracheal stenosis, which carries a high morbidity and mortality. So far only one case of late prenatal diagnosis (PND) at 32 weeks' gestation has been reported. We present a case of early PND of LPAS at 23 weeks. A 34 year-old primigravida had amniocentesis at 17 weeks for high-risk Down syndrome screening. Fetal karyotype was normal. Routine anomaly scan at 22 weeks showed early onset intrauterine growth restriction (IUGR) with umbilical artery absent end diastolic velocity. Mesocardia was also suspected. Fetal echocardiography at 23 weeks showed mild rightward cardiac displacement. The right pulmonary artery (RPA) was normal but normal LPA was not seen. 2D and colour Doppler examination found that the LPA arose from the RPA. After running a short distance posteriorly, it turned sharply leftward around right side of the trachea and then coursed behind it to reach the left lung. Pulsed Doppler study of this aberrant artery showed the characteristic flow pattern of a branched pulmonary artery. The couple opted for continuation of pregnancy. Intrauterine fetal death occurred at 27 weeks because of severe IUGR. Labour was induced and a 560-gram female baby was stillborn. Postmortem examination confirmed the presence of LPAS. PND of LPAS is rare because of lack of ultrasound clue for its identification. In our and the previous reported case, fetal cardiac dextroposition was present.

Conclusion

PND may prevent serious outcome of LPAS by allowing early postnatal detection. Early PND also gives the parents the option of termination of pregnancy. Cardiac dextroposition may serve as an ultrasound clue for PND of LPAS.

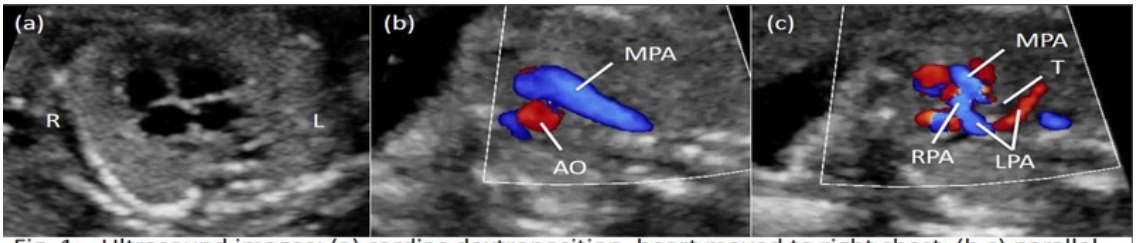

Fig. 1 – Ultrasound images: (a) cardiac dextroposition, heart moved to right chest; (b,c) parallel axial planes showing the left pulmonary artery arising from the right pulmonary artery and coursing around the right side of and behind the trachea to reach the left lung. MPA main pulmonary artery, LPA left pulmonary artery, RPA right pulmonary artery, AO aorta, T trachea

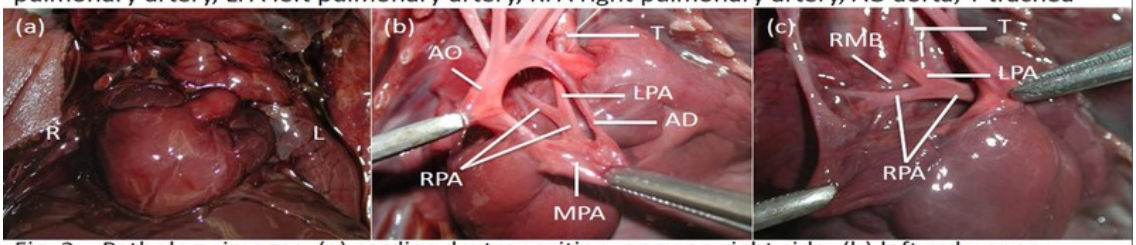

Fig. 2 – Pathology images: (a) cardiac dextroposition, apex on right side; (b) left pulmonary artery arising from the right pulmonary artery; (c) left pulmonary artery coursing over the right main bronchus and behind the trachea. MPA main pulmonary artery, LPA left pulmonary artery, RPA right pulmonary artery, AO aorta, AD arterial duct, T trachea, RMB right main bronchus