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Rare antenatal presentation of cardiac panvalvular calcification with coarctation of aorta resulting in Hydrops fetalis in a fetus with Trisomy 21

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Abstract
Non immune hydrops fetalis may be associated with multiple fetal anatomic and functional abnormalities including cardiac structural defects and rhythm abnormalities which cause pressure and volume overload of right side of fetal heart and eventually venous hypertension and systemic edema. Fetal panvalvular calcification with Coarctation of aorta is an unusual and rare ultrasound finding associated with defunctioning myocardium leading to Non-immune hydrops fetalis (NIHF).We are reporting a case of 32 year old G3P2L2 reported at 23 weeks period of gestation from primary health centre with ultrasound examination report suggestive of all panvalvular cardiac calcifications. Fetal echo was performed and provisional diagnosis of interrupted aortic arch, panvalvular calcification with biventricular hypertrophy and moderate pericardial effusion was made. Patient was evaluated for non immune hydrops fetalis and cordocentesis was performed at 27 weeks and karyotype of lymphocyte culture with GTG revealed 47, XX, +21. On serial imaging fetal hydrops ensued followed by intrauterine fetal death. Subsequent reports confirmed all valve calcification with coarctation of aorta in fetus with Down syndrome.

Keywords: immune, intrauterine, Down syndrome, hydrops, panvalvular

Introduction

A 32 –year-old G3P2L2 was referred in our hospital with ultrasound report suggestive of fetal panvalvular calcification. Detailed anomaly scan in our hospital reconfirmed panvalvular calcification rest no gross congenital abnormality noted. Fetal echo was performed at 25 week period of gestation and provisional diagnosis of interrupted aortic arch with hyper echoic valves, biventricular hypertrophy and moderate pericardial effusion was made. Meanwhile the patient was evaluated for causative factors associated with non immune hydrops fetalis and was prognosticated regarding risk of aneuploidies i.e. trisomy 13, 18, and 21, post delivery ventilation and ICU care with poor chances of survival. Patient was offered cordocentesis at 27 weeks. Patient blood group was A positive and her husband's blood group was B positive, her blood sugar and blood pressure records were normal. Maternal infection screen was negative except CMV and Rubella IgG positive suggestive of old infection. Maternal autoantibody screen was negative. Fetal karyotype was trisomy 21. On further follow up, patient developed polyhydramnios and there was progressive pericardial effusion with fetal ascites. Fetal death was observed at 31 weeks. Patient delivered a macerated female baby with facies suggestive of down phenotype and gross fetal ascites and pericardial effusion. Fetal autopsy confirmed ultrasound findings of all cardiac valve stenosis likely due to calcification with constriction of aorta distal to ductus arteriosus and retrograde dilatation of ductus arteriosus, interrupted aortic arch was not seen. Placenta and membrane examination were suggestive of two vessel cord with mild perivillous fibrous exudates. Patient was discharged after 24 hours of observation. Breast suppression was given.

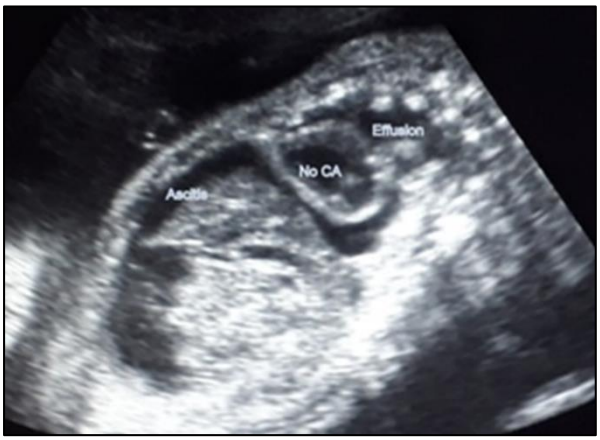

Fig 1

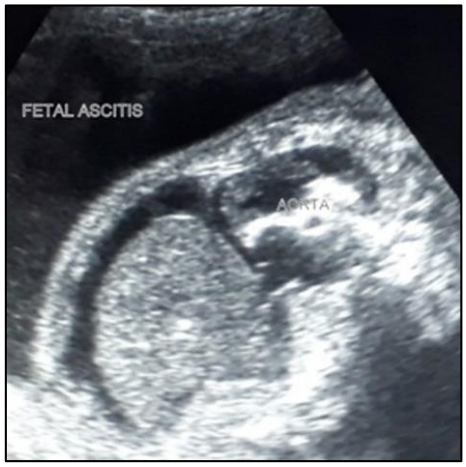

Fig 2

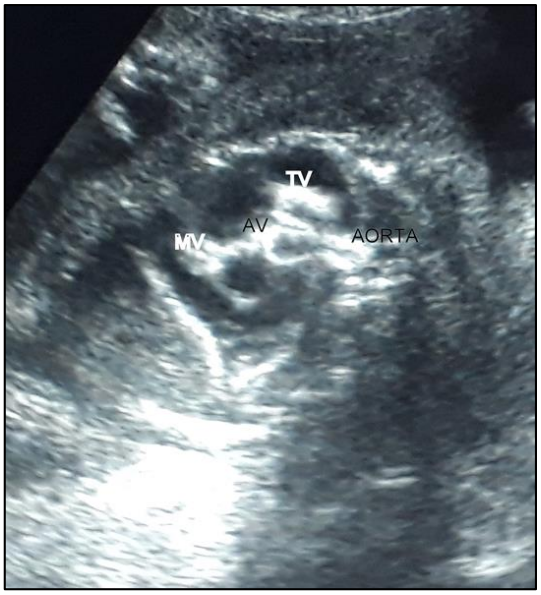

Fig 3

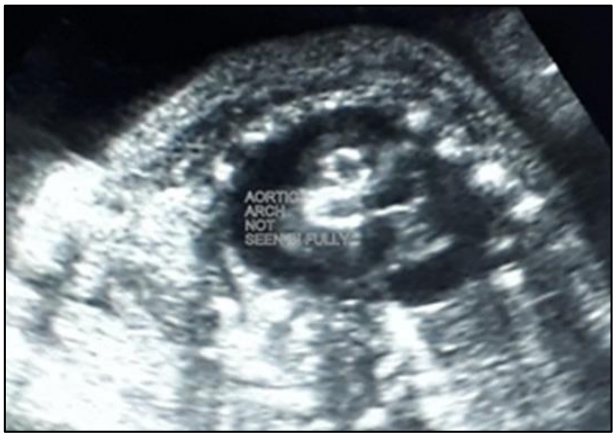

Fig 4

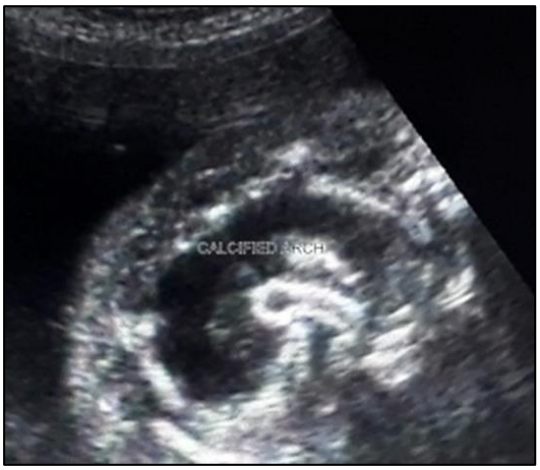

Fig 5

Discussion

Dystrophic calcification resulting in myocardial and valvular calcification is found to be associated with maternal autoimmune conditions like SLE, scleroderma etc, inherited thrombophilias, maternal infections and chromosomal abnormalities in fetus [1]. Fetal cardiac valve calcification is a rare entity observed where valves appear hyper echoic with echogenecity comparable to bones. The calcifications can be very subtle in the form of echogenic intracardiac foci to dense involving myocardium and valves. The probable causes are excessive calcium deposition in areas with infection, inflammation, haemorrhage, necrosis and fibrosis. Non – immune hydrops fetalis developing after 24 weeks is associated with cardiac, pulmonary defect, and fetal infections in most of the cases [2, 3]. Cardiac abnormalities including structural abnormality, arrhythmias and vascular defects constitute 40% cases of non immune hydrops fetalis [4]. Severe fetal anaemia accounts for 10-27% cases of NIHF due to high output cardiac failure [4]. Aneuploidies causes 7% to 16% cases, of which turners constitute 42%-67% cases, trisomy 21 (23%-30%), trisomy 13, 18, 12 (10%) [4, 5]. Syndromic association was observed in 5-10% of cases [6]. Fetal infection constitute 5-10% cases of NIHF of which Parvovirus B 19 infection constitutes majority of cases [3, 7]. Other common infections having causative association includes cytomegalovirus, toxoplasma, syphilis, varicella, adenovirus, coxsackievirus and leptospirosis etc. Inborn errors of metabolism were responsible for 1%-2% of cases [4]. Monochorionic twin gestation with complications like TTTS and TRAP constitutes minority of cases of non immune hydrops fetalis. M.J.Simchen *et al.* [8] reported four cases with antenatally diagnosed cardiac calcifications with extensive myocardial calcification in 3 cases leading to cardiac failure with hydrops and in fourth case was likely due to fetal infection, though no corroborative evidence can be found. Jami C. Levine *et al.* [9] reported an antenatally diagnosed fetal cardiac calcification which on evaluation was diagnosed with infantile idiopathic arterial calcification, here also fetal death was observed at 35 weeks and on autopsy the antenatal findings were confirmed and calcium hydroxyapetite deposition was noted in internal elastic lamina. Thomas C. Wheeler *et al.* [10] reported case of rare antenatal presentation of interrupted aortic arch in elderly multigravida presented with fetal hydrops in target scan along with maternal polyhydramnios, on evaluation complex cardiac abnormality was noted, diagnosis of interrupted aortic arch was noted after termination of pregnancy in fetal

autopsy and karyotype was 47XX+21. Cuillier F *et al*. [11] reported an abnormal case of fetal cardiac calcification diagnosed at term in fetus with all previous ultrasounds normal. Postnatally the diagnosis of dystrophic calcification was made with tricuspid dysplasia, the infant was doing well at 1 month post delivery. Yap *et al*. [12] reported two cases of fetal myocardial calcification having association with maternal cocaine abuse, both fetus survived. Although literature was available for association of interrupted aortic arch with Trisomy 21, the literature for association of coarctation of aorta with trisomy 21 was very scanty. Al Jarallah *et al*. [13] detected no cases of coarctation of aorta in Trisomy 21 patients. Stoll C *et al*. [14] reported incidence of coarctation of aorta in trisomy 21 patients was 5% in their study.

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