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Case Report:

Idiopathic Infantile arterial calcification –A Very rare case

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Abstract:
A rare case of Idiopathic Arterial Calcification of Infancy (IACI), inherited as an autosomal recessive disease, is reported.
Key Words: Arterial calcification, Infancy, Autosomal recessive

Introduction:
Calcification of heart and vessels in fetus is a very rare condition. It may be dystrophic or metastatic. An extremly rare form of vascular calcification has been termed Idiopathic Arterial Calcification of Infancy (IACI) which is inherited in an autosomal recessive pattern. We report a case of IACI diagnosed in our hospital.

Case Report:
A 26 year old woman, gravida 2, para 1 with no living child, with 20 weeks amenorrhea came for antenatal check up to our clinic. Her previous pregnancy was full term normal delivery and baby died at 11th month of age, reasons not being clear. Anomaly scan done at 20 weeks was reported as normal fetus. Patient came with preterm labor at 28 weeks of pregnancy and delivered a preterm baby weighing 1.2 kg which died 5 min after birth. Baby was sent for autopsy, as there was history of loss of first child. Autopsy report revealed fetal arterial calcification which involved massive calcification of aortic vessels, coronary vessels, pulmonary vessels, pancreatic vessels and vessels in the kidney. Liver showed extramedullary hematopoiesis and there was also hypertrophy of myocardium.

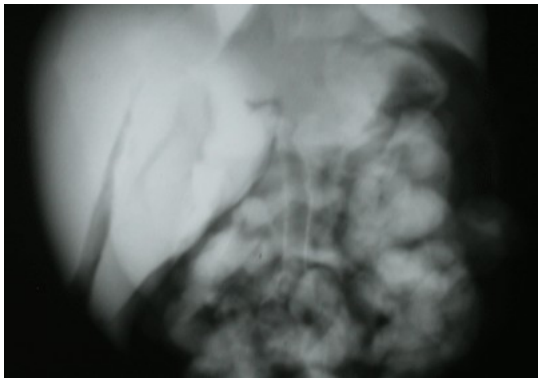

Fig 1: Calcification of descending aorta

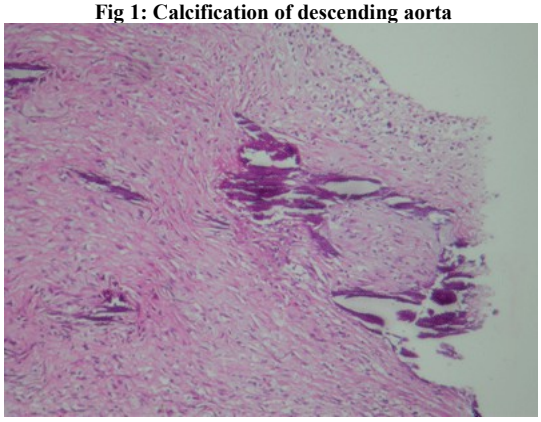

Fig 2: Calcification of aorta

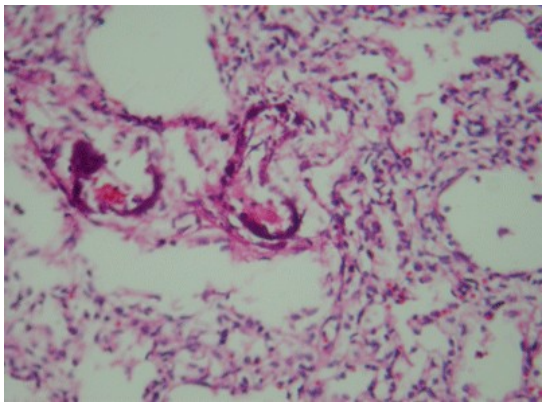

Fig 3: Calcified pulmonary vessels

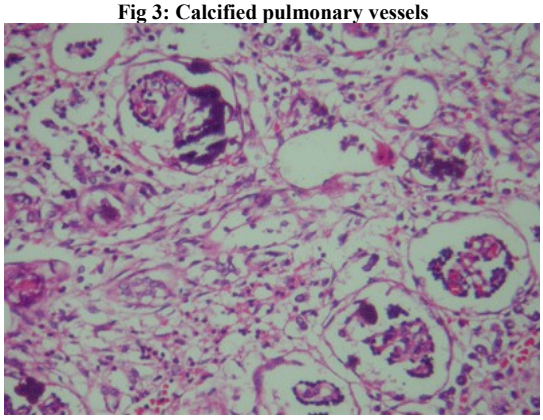

Fig 4: Calcified renal vessels

Fig 5: Myocardial hypertrophy

Discussion:

Idiopathic arterial calcification of infancy or occlusive infantile arterial calcification[1] is a rare congenital disorder which is usually fatal in first six months of life[2] and very few have survived more than one year. This condition is inherited as an autosomal recessive pattern[3] and diagnosis is usually made at autopsy in most of the cases.[4] Only about 161 cases have been described by literature to date, with antemortem diagnosis being made on the radiographic or sonographic demonstration of arterial calcification in only a few cases. It was first described by Bryant and White in 1901. It is characterized by widespread and extensive calcification and stenosis of large and medium sized arteries. This results from the deposition of calcium hydroxyapatite in the arterial internal elastic lamina layer, which leads to rapid progressive ischemic heart failure and refractory hypertension either prenatally or most commonly in neonatal and early infancy period.[5] Biochemical investigations are usually normal and there is no identifiable abnormality of calcium metabolism.[2]

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