Case Report

Mesangioproliferative Glomerulonephritis in a case of Pulmonary Atresia with Ventricular Septal Defect (Pseudotruncus arteriosus) : An Interesting Case Report

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Pulmonary atresia with ventricular septal defect (PA-VSD) with pulmonary arterial supply arising from aorta represented by large Major Aortopulmonary Collateral Arteries (MAPCAs) associated with a right sided aortic arch is an uncommon anomaly. Most of the patients succumb to severe respiratory compromise or congestive cardiac failure very early. Around 9% of adults with congenital heart disease likely to have moderate to severely impaired renal function and as a result have an additional adjusted 3 fold increased mortality risk. Here we are reporting a case with PA-VSD with glomerulonephritis, a rarely reported entity.

[*J Indian Med Assoc* 2021; **119(1):** 67-9]

***Key words*: Pseudotruncus arteriosus, Pulmonary Atresia, Ventricular Septal Defect, Mesangioproliferative Glomerulonephritis.**

Truncus Arteriosus is characterized by a single great artery with a single semilunar valve that leaves the base of the heart and gives rise to coronary, pulmonary and systemic circulations. In 1949, Collett and Edwards classified this anomaly into four types. Type IV ie, Biventricular aorta with an atretic pulmonary valve, is now considered as ultimate expression of severity in Fallot's tetralogy[1]. However considering the similarities with Truncus Arteriosus it is also sometimes called Pseudotruncus.

Cyanotic Nephropathy (CN) is often accompanied by congenital cyanotic heart diseases. Hyperviscocity due to polycythemia can be the underlying cause. In addition, failure of a compensatory mechanism to respond to reduced Renal Plasma Flow may be responsible[2].

We are presenting a case of 30 years old patient, with Pseudotruncus and Cyanotic Nephropathy.

Case Report

Our patient is a 30 year old, non diabetic, non hypertensive, non hypothyroid male patient who presented to us with facial puffiness and pedal swelling for two weeks duration. The swelling started from face and later involved his feet. It was associated with diminished urine output although no history of frothy or cola coloured urine was present. He had no history of preceding fever, sore throat, skin infection, joint pain or any drug intake. He also didn't complaint of any shortness of breath, cough, palpitation or jaundice. Although his past history was significant with bluish discolouration of body since 6th day of life and also there was history of recurrent chest infection (8-9 episodes each year since the age of 6 years). But there was no history suggestive of cyanotic spell. After admission on examination he was found to have ruddy conjunctiva, cyanosis, grade 3 clubbing and pedal oedema. His Jugular venous pressure (JVP) was engorged. His respiratory rate was 20/min, pulse rate was 90/min. He had no palpable neck gland. His Cardiovascular Examination findings were as follows: Apex was found to be in 6th intercostal space, outside midclavicular line with a palpable diastolic thrill. Grade 3 parasternal impulse and Epigastric pulsation was present. On Auscultation, S1 was soft, S2 soft, single, S3 was present. A grade 4 mid-diastolic murmur was heard at the apex, a grade 3 continuous murmur was heard over the lower left sternal area and a grade 3 early diastolic murmur was heard best in the neo-aortic area. ECG suggested Right axis deviation, tall peaked P waves and tall R waves in V1. Chest X-Ray (Fig 1) was done, which showed, marked

Editor's Comment :

- **Congenital Cyanotic Heart Diseases are systemic diseases that may present with symptoms even beyond cardiovascular system.**
- **Although rare, complex cyanotic congenital heart diseases can present very late.**
- **With well-developed MAPCAs, patient may have no symptoms from cardiovascular point of view.**
- **Cyanotic Nephropathy can be associated with long standing cyanotic heart disease.**
- **Management of these complex congenital heart diseases with their complications is still a matter of discussion.**

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Received on : 12/02/2020
Accepted on : 25/08/2020

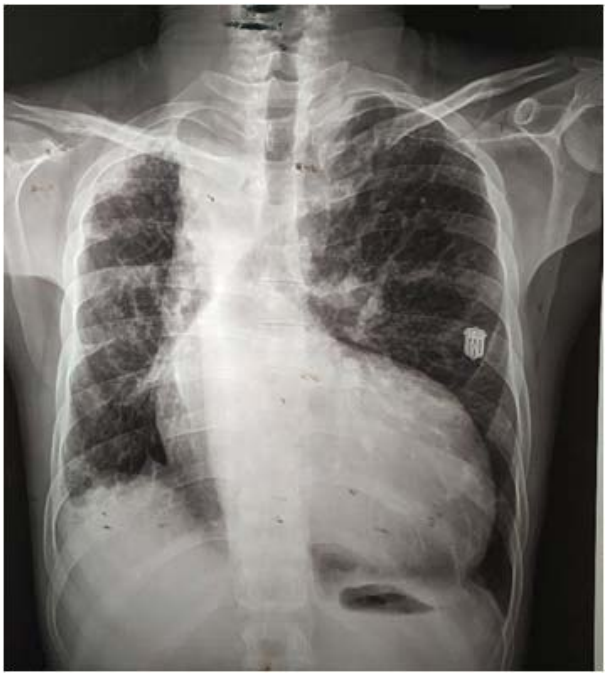

Fig 1 — Chest X Ray Showing Right lung hypoplasia and increased vascularity on Left Side. Cardiomegaly is present

Table showing the various blood parameters during hospital stay

Hb(gm/dl)	20.5
PCV(%)	63
TLC	9,100
Platelet	1.09L
Ur/Cr(mg/dl)	64/1.7
Na/K (m/mol)	136/4.4
Ca	7.6
ALP/ALT/AST	79/22/29
TB	0.8
TP/Alb	5.2/2.8
Chol/TG/HDL	281/266/65
C3/C4 (mg/dl)	111/35.2
dsDNA (IU/ml)	<10
PT/INR	13.8/0.97

dilatation of Pulmonary trunk and increased flow on the left lung, the Right lung seemed to be hypoplastic. Cardiomegaly was appreciated. We had a suspicion that we might be dealing with a case of Truncus Arteriosus, so we wanted to have a cardiological evaluation along with workup for renal parameters as the history was suggestive of renal pathology. Talking about his blood parameters, Haemoglobin and Haematocrit both were persistently raised, Creatinine was marginally elevated, deranged lipid profile with raised serum

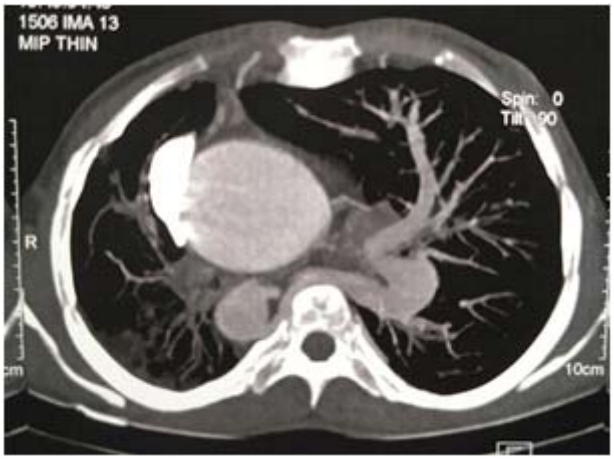

Fig 2 — Renal Biopsy showing (Upper left) Subcapsular Global Sclerosis, (Upper right) Glomerular hypercellularity, (Lower left) Tubular atrophy and Interstitial fibrosis, (Lower left) Similar changes in Silver methanamine stain

cholesterol and triglyceride. Urinalysis suggested albuminuria 2+, 24 hours quantification was done, which came out to be 2.7g. Renal biopsy was done which showed Mesangio-proliferative Glomerulonephritis with focal subcapsular scarring (Congenital heart disease associated glomerulopathy) (Fig 2). Regarding cardiological evaluation, 2D Echo with Colour Doppler suggested, moderate to severe

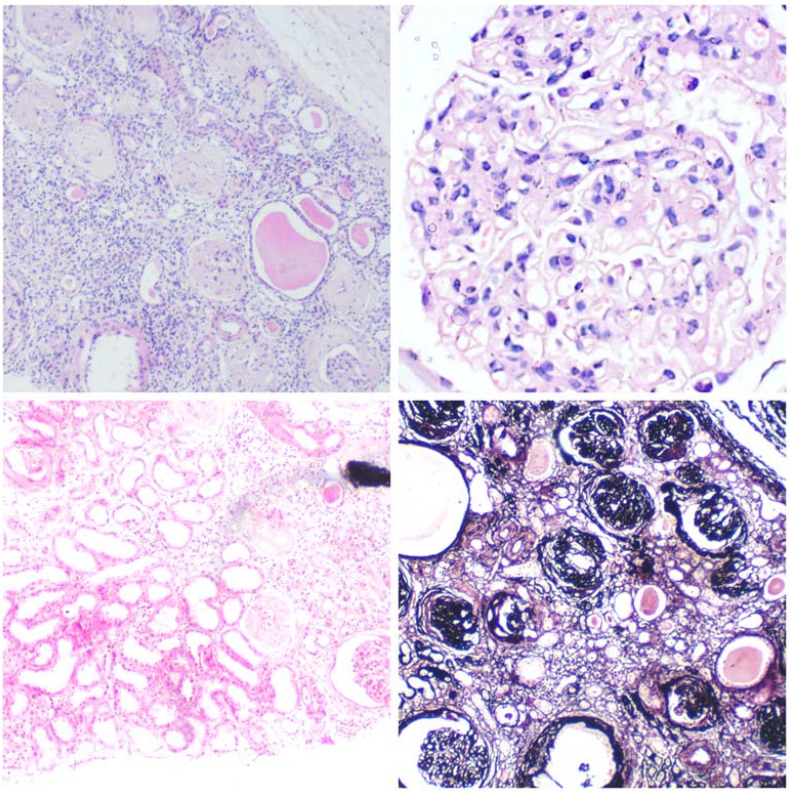

Fig 3 — CT Angiography showing that MAPCA going to Left Lung, whereas in Right lung vascularity is compromised

Augmented Reality, doubly committed aorta (<50%) with subaortic Ventricular Septal Defect (VSD) and Major aortopulmonary collateral arteries (MAPCA). Cardiac MRI was suggested, which suggested Truncus arteriosus and CT Cardio-Pulmonary Angiography (Fig 3) was done for confirmation. It showed, multiple non-confluent MAPCAs bilaterally with hypoplastic Right pulmonary artery, Right sided Aortic arch, large subaortic VSD (16 mm) with overriding of aorta. Pulmonary artery was not visualized. The features were suggestive of Pseudotruncus arteriosus.

After proper discussion with respective departments, it was concluded that neither there is any scope of surgical intervention, nor it is necessary. The patient was started on Renin Angiotensin Aldosterone System (RAAS) blocking agent for proteinuria and Statin for dyslipidemia and discharged for follow up.

Discussion

Pseudotruncus Arteriosus is indeed a very rare disease reported in the literature. In a study, 21 such cases were reviewed and the oldest survivor reported was 36 years old.[3] Thus for those survivors, treatment goals and management of complications are not well delineated. But congenital heart disease are systemic diseases with consequences reaching far beyond the heart. Cyanotic Nephropathy is one such complication. It is related to duration of cyanosis and extent to which the haematocrit is elevated.[4] In a study the renal changes observed were Glomerulomegaly, Glumerulosclerosis, Periglomerular fibrosis, Hyperplastic arteriosclerosis and Interstitial fibrosis.[5] Regarding management of nephropathy, a study using Enalapril for 12 months showed reduction in proteinuria in 80% of patients, although no change was demonstrated in Glomerular filtration rate (GFR), Renal plasma flow or Filtration fraction.[6] In this patient also, RAAS Blocker therapy has been initiated and the outcome needs to be evaluated during follow up.

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1. "Nature Makes Penicillin; I just found it."

2. "One sometimes finds what one is not looking for. When I woke up just after dawn on Sept. 28, 1928, I certainly didn't plan to revolutionize all medicine by discovering the world's first antibiotic, or bacteria killer. But I guess that was exactly what I did."

— Sir Alexander Flemming