



HYPERCYANOTIC SPELL IN A CHILD WITH LEFT BRANCH OF PULMONARY ARTERY MILD STENOSIS SUPER ADDED TO RIGHT SIDED AORTIC ARCH WITH ASCENDING LEFT AORTIC ANEURYSM CAUSING COMPRESSION OF AIRWAYS

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ABSTRACT

we describe a 2 month old male child who had an episode of central and peripheral cyanosis with respiratory distress following excessive and rigorous crying after waking up .The child had left bifid thumb and 2D Echo showing mild left pulmonary artery branch stenosis with right side aortic arch with ascending left aortic aneurysm causing compression of airways. Child had an episode of hyper cyanotic spell following excessive crying due to combined factors of left pulmonary artery mild stenosis (pressure gradient =20mmhg) and compression of upper airways i.e, trachea, due to left ascending aorta aneurysm. Child was managed for hyper cyanotic spell, and corrective cardiac surgery was advised and planned.

KEYWORDS : Hyper cyanotic spells, Upper airways, Aortic aneurysm

INTRODUCTION

A hyper cyanotic spell is a common presentation in cardiac lesions presenting with significant right ventricular outflow tract (RVOT) obstruction¹. Hypoxic spells are also known by the name of cyanotic spells, hyper cyanotic spells or tet spells². The hyper cyanotic spells are not seen as frequently now as children diagnosed with Tetralogy Of Fallot (TOF) receive corrective surgery more judiciously¹. Paediatricians should immediately be able to recognise Tet spells because prolonged hypoxia because of cyanotic spells can lead to serious hypoxic injury to brain¹.

Cyanotic spells are characterised by tachypnea, difficulty in breathing. Irritability, and prolonged and excessive crying, decreasing oxygen saturation levels and reduction in intensity of the heart murmur². Cyanotic spells present in infancy, more commonly between 2 to 4 months of age group and occur after excessive and prolonged crying, vigorous feeding or defecation.^{1,2}

A severe spell may lead to loss of tone of body, seizures, hypoxic central nervous system injury or even death¹. The time duration of cyanotic spells is between minutes to hours³. There is further decrease in pulmonary blood flow which if prolonged leads to decreased oxygen saturation of systemic blood and thus metabolic acidosis². Cyanotic spell more frequently occurs in children who have oxygen saturation at rest better than those who are more cyanotic at rest¹. This is because the latter group develops homeostatic mechanisms like polycythemia which acts as a preventive factor for a cyanotic spell to occur².

Aortic arch on right side is uncommon cardiac congenital anomaly³. An Aortic arch on right side is asymptomatic in majority of children³. However when accompanied with a vascular ring, aortic aneurysm causes symptoms due to compression of trachea or esophagus³.

PATIENT AND OBSERVATION

Two month old male child was born preterm (36weeks), Low birth weight, appropriate for gestational age. Child had a neonatal intensive care unit stay of 10 days for mild respiratory distress managed medically with oxygen via nasal cannula.

Child presented to paediatric emergency department with

history of bluish discoloration of lips, tongue and nails while crying excessively after just waking up.

On examination, a sick looking child with other positive findings on examination were SPO₂ 40% on room air, Central and peripheral cyanosis present, mild stridor present, left bifid thumb (FIG.1). Child was irritable with respiratory rate of 70/minute, heart rate of 140/min, sub costal, intercostals chest retractions were present. Chest was clear, cardiovascular system no significant finding and abdominal examination showed soft liver 2.5cms below right costal margin with span of 5cms.



Fig. 1 Bifid Thumb

Investigations showed metabolic acidosis in arterial blood gas (ABG), Chest x ray was normal showing no significant oligemia or plethora in lung fields (FIG.2). Electrocardiogram was normal (FIG.3). Other routine investigations were normal. 2 D echocardiography showed: Ascending left aortic aneurysm with right aortic arch with mild left pulmonary arterial branch stenosis with pressure gradient of 20mmhg, left branch of pulmonary artery Ostium: 4.7mm; right branch of pulmonary artery ostium: 7.1mm

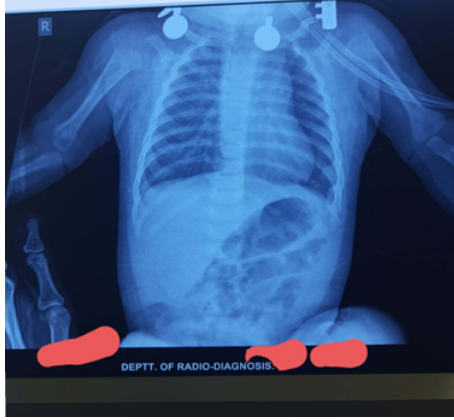


Fig. 2 CXR Of Child

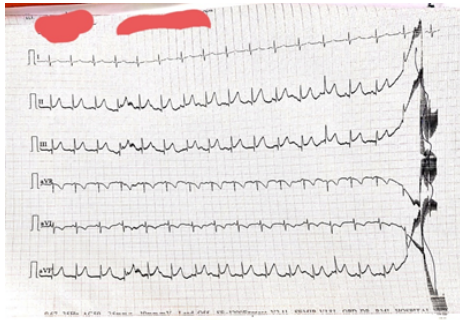


Fig.3 ECG Of Child

Child was planned for cardiac CT, was investigated for any chromosomal disorder or any congenital syndrome giving rise to multitude of congenital anomalies mentioned above. Child was managed medically with high flow oxygen (FIG.4 &5), knee chest position, intravenous fluids, IV morphine, IV soda bicarbonate and IV propranolol.



Fig. 4 Child Being Managed During Tet Spell



Fig. 5 Calm Child After Tet Spell Was Over

DISCUSSION

Hyper cyanotic spells occur usually in infants in cyanotic heart diseases more commonly in TOF due to reduction in already compromised pulmonary blood flow because of significant RVOT obstruction¹. There is combined central and peripheral cyanosis². Irritability and hypoxic seizures may occur¹. Cyanotic spells usually occur in morning after prolonged and excessive crying or vigorous feeding or defecation³.

In our 2mon old child, the RVOT obstruction was not present, only left branch of pulmonary artery was mildly stenotic. Hence cyanotic spell was least likely presentation. But due to combined lesions of aortic arch being on right side and aneurysm of left ascending aorta causing compression of trachea plus mild left pulmonary artery branch stenosis and decreased blood flow across left pulmonary artery as child was crying excessively and rigorously, child presented with cyanotic spell.

Child was managed medically with oxygen, Intravenous fluids, IV morphine, IV propranolol , IV soda bicarbonate and kept in knee chest position.

This is a rare case of hyper cyanotic spell developing due to upper airway compression combined with decrease in already compromised pulmonary blood flow in our case due to mild stenosis of left branch of pulmonary artery.

CONCLUSION

A rare case of 2 month old male infant presenting with hyper cyanotic spell due to combined lesions of upper airway compression, trachea, due to ascending left aortic arch aneurysm with right sided aortic arch and mild stenosis of left branch of pulmonary artery.

Competing Interests: None

Authors Contributions: All authors contributed in realisation of this case study .All authors have read and agreed to the final manuscript.

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