MID AORTIC SYNDROME- A RARE CA(U)SE OF HYPERTENSION IN CHILDREN.

BOOPATHY
Department of Paediatrics, MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract: Mid aortic Syndrome (or) Middle aortic Syndrome (MAS) is a rare disease with only 200 reported case. MAS may present clinically as Uncontrollable Hypertension, abdominal claudication or Lower limb claudication. We present another case of this rare entity, diagnosed and treated at Institute of Child Health, Chennai.

Keyword : Mid aortic Syndrome, Hypertension, Abdominal aortic co-arctation.

CASE REPORT:
A 11 year old male child admitted with Complaints of sudden onset weakness of right upper limb and lower limb along with deviation of angle of mouth to the left side. The child had no other complaints like headache, vomiting, seizures. For the above mentioned complaints, the child was admitted in a private hospital and diagnosed to have hypertension. C.T. Brain was taken which showed intracranial haemonhage and the child was referred to ICH. At ICH, the child was examined and found to have blood pressures of right upper limb-160/120 mm Hg, left upper limb- 164/122 mm Hg, right Lower limb-120/80 mm Hg, left Lower limb- 120/100 mmHg. Upper limb pulses were well felt. Lower limb pulses were feeble. Abdominal bruit was present on the left side of the umbilicus. Examination of central nervous system revealed right sided hemiparesis with upper motor neuron type of right sided facial nerve palsy with higher functions and sensory system being normal. Examination of other Systems were also normal. Investigations revealed normal CBC and ESR. Renal and liver function tests and coagulation profile were normal. CRP, ANA were negative. USG Abdomen showed nonvisualisation of the left kidney with grade I echoes in the right kidney. Echocardiography showed mild concentric hypertrophy of the left ventricle and discrete obstruction in the abdominal aorta at the level of superior mesenteric artery (SMA) and renal rtery. C.T. Brain & M.R.I. Brain revealed left gangliocapsular hemorrhage. Renal Doppler showed discrete obstruction in the abdominal aorta at the level of SMA. C.T. Angiogram showed narrowing of abdominal aorta & thinning of left renal artery with hypoplastic left kidney and other vessels were normal.

DISCUSSION:
Mid aortic Syndrome or Middle Aortic Syndrome (MAS) is a rare entity affecting abdominal aorta in children and young adults. It is characterized by constriction of distal thoracic and/or abdominal aorta and its branches, therefore it is also known as “Abdominal aortic co-arctation”. MAS can present as hypertension or lower limb claudication or abdominal angina. MAS is characterized radiologically by severe narrowing of abdominal aorta and its branches. Most of these patients usually die due to progressive severe hypertension before the age of 35-40, if left untreated.
Acquired MAS is associated with neurofibromatosis, William’s syndrome, Alagille syndrome, fibromuscular dysplasia, retroperitoneal fibrosis, mucopolysaccharidosis, Takayasu arteritis, and temporal arteritis. The most common anatomic form in congenital or acquired MAS is inter renal (19-52%), followed by supra renal (11-40%), infra renal (19-25%), and diffuse (12%). Stenosis of the renal arteries is common (60-90%), then celiac and SMA (20-40%) lastly inferior mesenteric artery.

It usually present as hypertension in young age which is unresponsive to medical therapy. Rarely it presents as lower limp claudication or abdominal angina due to proximal stenosis (hypertension) and distal stenosis causing hypotension. The life expectancy of patients with untreated MAS is 30-40 years. The main cause of death is cardiovascular complications of progressive hypertension including cerebrovascular accidents, left heart failure, coronary artery disease. Medical treatment may be helpful in mild to moderate hypertension. Open surgery is the primary treatment in renovascular hypertension and visceral artery stenosis.

CONCLUSION:
MAS is a rare cause of uncontrolled hypertension with poor outcome if left untreated. MAS may present as hypertension in both upper limbs and hypotension in both lower limbs and feeble peripheral pulses in the lower limb. And hemiparesis due to intracerebral hemorrhage as in this case.

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