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Mid aortic syndrome: A rare cause of hypertension in children

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Abstract

Case report: A 6-year-old Hispanic female, with no dysmorphic features. She was on treatment for Lichen sclerosis etatrophicus and was found to have episodic symptomatic hypertension. She had multiple episodes of severe headache; vomiting and vision disturbance that required ER visits.
No significant PMH or inflammatory symptoms and signs.
FSH was significant for migraine in siblings and mother. Father was deported to Mexico after the initial clinic visits. Physical examination was within normal limit. Peripheral pulses were well felt and no differential blood pressure between the limbs.
Investigations: CXR did not show abnormalities. EKG showed multiple wide complexes. Holter monitoring showed ventricular ectopy. Labs showed high renin activity: 6.41 ng/mL/h (0.25-5.82). CMP, ESR, ANA, cortisol, thyroid function test (TFT), Angiotensin converting enzyme (ACE), total cathecolamines, vanillylmandelic acid (VMA), homovanillic acid (HVA) and were all within normal limits. Scleroderma and SLE screen were negative. Renal Doppler US was within normal limit. Echocardiography showed no evidence of coarctation of the aorta. Cardiologist and nephrologist were consulted and she was cleared as situational hypertension. Subsequent follow-up chest auscultation revealed *paravertebral continuous bruit* in the inter-scapular region. Chest and abdominal CT angiogram showed abnormal tapered stenosis of the subdiaphragmatic descending thoracic aorta with distal kinking. There is greater than 50% luminal narrowing. No definitive evidence of arteritis. There was significant decrease in luminal diameter which maybe resulting in bilateral renal hypoperfusion and renin dependent hypertension.
Treatment: Cardiologist re-consulted and she was started on Labetalol. Angiography and catheterization were done to determine the extent of abnormalities and calculate the gradient prior to surgery. She is awaiting surgery to be done in San Antonio.
Conclusion: Mid Aortic Syndrome (Coarctation) is a rare (0.5 to 2%) cause of hypertension in children. The diagnosis requires high index of suspicion. Inter-scapular auscultation should always be part of routine cardiac examination in children.

Biography

Assistant Professor of Pediatrics, TTUHSC, SOM, El Paso.Medical education at College of Medicine, of the University of Lagos, Nigeria.
Completed Pediatrics Residency at Lagos University Teaching Hospital, Nigeria.
In the USA, Pediatric Residency at TTUHSC El Paso Texas.I had a Mini fellowship / Elective Course in Child Maltreatment, University of Texas, San Antonio, Child Abuse Division in Center for Miracles. Board certified in General Pediatrics. Bachelor of Medicine and Bachelor of Surgery (MBBS) Lagos.Fellowship of the Nigerian Postgraduate Medical College in Pediatrics (FMCPead) Nigeria.Membership of the Royal College of Physicians (MRCP) United Kingdom.American Board of Pediatrics.Fellow of the American Academy of Pediatrics.