Interesting Case of Pediatric Hypertension

DR. Naga jyothi - Dnb Pediatrics - Resident
Dr. S. Ramkumar MD, DNB(Endo), DM(Endo) AIIMS

Dept. of Pediatric Endocrinology & Diabetology
Apollo Children’s Hospital
Case history

- 13 year old boy (Twin) from Erode, who was apparently normal 1 month before had fever for 3 days, for which his parents consulted a general pediatrician.
- There incidentally boy was detected to have had hypertension.
- Blood pressure recorded was 180/130 mm hg in the rt upper limb.
## Causes of Pediatric Hypertension

<table>
<thead>
<tr>
<th>Age</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>One to six years</td>
<td>Renal parenchymal disease; renal vascular disease; endocrine causes; coarctation of the aorta; essential hypertension</td>
</tr>
<tr>
<td>Six to 12 years</td>
<td>Renal parenchymal disease; essential hypertension; renal vascular disease; endocrine causes; coarctation of the aorta; iatrogenic illness</td>
</tr>
<tr>
<td>12 to 18 years</td>
<td>Essential hypertension; iatrogenic illness; renal parenchymal disease; renal vascular disease; endocrine causes; coarctation of the aorta</td>
</tr>
</tbody>
</table>
History relevant to HTN

- No history of abdominal pain, no history of oliguria/hematuria, no history of increased frequency of urine.
- No history of chest pain, paroxysms of palpitations, breathlessness, headache, sweating
- No history of arthritis, no history of rashes
- No history suggestive of weakness, muscle cramps, polyruia
- No history of drug intake
- No weight gain since 6 months, inspite of good appetite.
History (Cont..)

- His twin sibling is normal.
- No similar complaints in the past
- No family history of hypertension.
- Paternal grandfather died of some chest tumor.
On examination

- He got admitted in Apollo Hospital
- At admission,
  - Weight: 41kg, Height: 165cm, normal BMI
  - BP: upper limbs, supine – 140/90 mmHg, 108/min
    standing – 130/80 mmHg, 114/min,
    no radioradial or radiofemoral delay
  - BP: lower limbs, 150/90mmHg
- Skin: no café au lait spots, no neurofibromas
- Systemic examination normal
- Medications:
  - Tab. Minipress XL 5mg OD
  - Tab. Amlodipine 5mg OD
### Pointers to hypertension

<table>
<thead>
<tr>
<th>Physical findings</th>
<th>Potential relevance</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>General</strong></td>
<td></td>
</tr>
<tr>
<td>Pale mucous membranes, edema, growth retardation</td>
<td>Chronic renal disease</td>
</tr>
<tr>
<td>Elfin facies, growth retardation</td>
<td>Williams syndrome</td>
</tr>
<tr>
<td>Webbing of neck, wide spaced nipples, wide carrying angle, low hair line</td>
<td>Turner syndrome</td>
</tr>
<tr>
<td>Moon facies, buffalo hump, hirsuitism, trunkal obesity, striae</td>
<td>Cushing syndrome</td>
</tr>
<tr>
<td><strong>Habitus</strong></td>
<td></td>
</tr>
<tr>
<td>Thinness</td>
<td>Pheochromocytoma, renal disease, hyperthyroidism</td>
</tr>
<tr>
<td>Virilization</td>
<td>Congenital adrenal hyperplasia</td>
</tr>
<tr>
<td>Rickets</td>
<td>Chronic renal disease</td>
</tr>
</tbody>
</table>
### Pointers to hypertension

<table>
<thead>
<tr>
<th><strong>Skin</strong></th>
<th><strong>Eyes</strong></th>
<th><strong>Head and neck</strong></th>
<th><strong>Neurological signs</strong></th>
<th><strong>Neurologic deficits</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Café au lait spots, neurofibromas</td>
<td>Proptosis</td>
<td>Goitre</td>
<td>Neurofibromatosis, pheochromocytoma</td>
<td>Chronic or severe acute hypertension with stroke</td>
</tr>
<tr>
<td>Tubers, ash leaf spots</td>
<td></td>
<td></td>
<td>Tuberous sclerosis</td>
<td></td>
</tr>
<tr>
<td>Rashes</td>
<td></td>
<td></td>
<td>SLE, vasculitis</td>
<td></td>
</tr>
<tr>
<td>Palor, flushing, sweating</td>
<td></td>
<td></td>
<td>Pheochromocytoma</td>
<td></td>
</tr>
<tr>
<td>Needle tracks</td>
<td></td>
<td></td>
<td>Illicit drug use</td>
<td></td>
</tr>
<tr>
<td>Eyes</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
## Pointers to hypertension – systemic examination

<table>
<thead>
<tr>
<th>Cardio vascular signs</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Absence or diminished femoral pulses, low leg pressure relative to arm</td>
<td>Aortic coarctation</td>
</tr>
<tr>
<td>Respiratory difficulty, hepatomegaly</td>
<td>Congestive heart failure</td>
</tr>
<tr>
<td>Bruit over great vessels</td>
<td>Arteritis or arteriopathy</td>
</tr>
</tbody>
</table>

**Pulmonary signs**

| Pulmonary edema                                             | Congestive heart failure |
| Picture of Broncho pulmonary dysplasia                      | BPD associated hypertension |
Pointers to hypertension – systemic examination

<table>
<thead>
<tr>
<th>Abdomen</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Epigastric bruit</td>
<td>Primary renovascular disease or in association with williams syndrome, neurofibromatosis, fibromuscular dysplasia, arteritis.</td>
</tr>
<tr>
<td>Abdominal masses</td>
<td>Wilms tumor, neuroblastoma, pheochromocytoma, polycystic kidney, hydro nephrosis</td>
</tr>
<tr>
<td>Genitalia</td>
<td></td>
</tr>
<tr>
<td>Ambiguous, virilized</td>
<td>Congenital adrenal hyperplasia.</td>
</tr>
</tbody>
</table>
History till now

- 13 yr
- Asymptomatic severe hypertension
- Normal BMI, no acanthosis nigricans
- No pointers to renal cause – history/clinical examination
- Normal difference between limbs (COA/aortoarteritis)
- No drug history
# Causes of Pediatric Hypertension

## Causes of Childhood Hypertension According to Age Group

<table>
<thead>
<tr>
<th>Age</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>One to six years</td>
<td>Renal parenchymal disease; renal vascular disease; endocrine causes; coarctation of the aorta; essential hypertension</td>
</tr>
<tr>
<td>Six to 12 years</td>
<td>Renal parenchymal disease; essential hypertension; renal vascular disease; endocrine causes; coarctation of the aorta; iatrogenic illness</td>
</tr>
<tr>
<td>12 to 18 years</td>
<td>Essential hypertension; iatrogenic illness; renal parenchymal disease; renal vascular disease; endocrine causes; coarctation of the aorta</td>
</tr>
</tbody>
</table>
Investigations recommended

- Urine analysis
- Urine culture
- Serum creatinine
- Serum electrolytes
- Blood counts
- X ray chest and abdomen
- USG abdomen
- Intra venous urogram
- Voiding cystourethrogram
Investigations recommended

- Radio nuclide imaging
- Selective renal angiography
- CT kidneys
- Doppler flow ultrasound
- MRI arteriogram
- CT angiography
- Renal vein assay
- Peripheral plasma renin activity/Plasma aldosterone
- Serum cortisol and 24 hour urine 17 hydroxy corticosterone
- Urinary and plasma catecholamine levels
- Meta iodo benzyl guanidine (MIBG) scan
Our boy - In Erode

- Evaluated by Cardiologist in Erode
- Started on anti-hypertensive medications
- Chest X-ray showed a mediastinal lesion
- CT scan done in Erode: para-aortic mass – suspected as paraganglioma
- 24 hour urinary VMA was done – normal

- Referred to Chennai for Surgery

- Medications at admission:
  Tab. Minipress XL 5mg OD
  Tab. Amlodipine 5mg OD
Investigations done for our boy

- Urine routine examination normal
- Serum creatinine normal
- Serum electrolytes normal
Chest X-ray

- Well defined soft tissue lesion seen in the left retrocardiac region, paravertebral location
CT Thorax

- 5 x 3 x 3 cm well circumscribed significantly enhancing solid mass in the para-aortic region
- D8-D10 levels
Mediastinal masses

ANTERIOR
- Thymoma
- Lymphoma
- Germ cell tumors
- Mesenchymal tumors
- Thyroid and parathyroid masses
- Thymic cyst

MIDDLE
- Cysts (pleuropericardial, neuroenteric)
- Vascular aneurysm
- Lymphoma
- Mesenchymal tumors

POSTERIOR
- Neurogenic tumors
- Meningoceles
- Gastroenteric cyst
- Mesenchymal tumors
- Pheochromacytoma
- Lymphoma
Provisional Diagnosis

- Extra adrenal mediastinal pheochromocytoma (paraganglioma) with Hypertension
Further Evaluation

- 24 hours urinary catecholamines

- I-131 MIBG Scan
Catecholamines

• 24 hours urinary
  – Metanephrines: 180 mcg/24 hours (39 - 242)
  – Normetanephrines: 3784 mcg/24 hours (53 - 290)
I-131 MIBG scan

• Must if
  1. Suspected malignancy to know metastasis
  2. Bilateral pheochromocytoma
  3. Paraganglioma
  4. Youth with pheochromocytoma
I-131 MIBG scan

Focal abnormal increased I-131 MIBG uptake noted in the left paravertebral region in the lower thorax.
48 HRS IMAGING

I-131 MIBG WHOLEBODY 48 HRS

ANTERIOR
72 HRS

POSTERIOR

ANTERIOR

POSTERIOR
I-131 MIBG/SPECT-CT

Focal uptake in the well defined soft tissue mass in left para-vertebral region at the D8-D10 level
SPECT
Pre-operative preparation

- Volume expansion –
- Fluids
- Added Salt
- Control of BP/HR

First – alpha blockade
Do not use XL
After adequate alpha-blockade, beta-blockers to be added
At least of beta-blockers to be given for 3 days before surgery

CCB – Amlodipine can be added for better BP control
Pre-operative preparation

- Encouraged adequate fluids up to 3L/day
- Added salt 10gm per day

- Minipress XL stopped and changed to plain Tab.prazosin 1.0mg TDS
- Tab.Atenolol 25mg once daily added

- Prior to Sx,
- Had BP controlled 110/74, 80/min, 100/70, 84/min
VATS excision of mediastinal tumor

VATS – video assisted Thoracoscopic surgery

By

• Dr. Rajiv Santosham
• Dr. Rajan Santosham
Follow-up

- Postoperatively child bp is normalised in one day
- Biopsy report: specimen consistent with paraganglioma, posterior mediastinum.
Post-op

- BP normal after 2 weeks of discharge

Further plan

- Repeat urine catecholamines after 1 month

- Screening of twin sibling

- Genetic analysis for familial paraganglioma- SDH gene, VHL gene planned

- Life long follow up yearly
Thoracoscopic resection of functional posterior mediastinal paraganglioma: a case report.

A 48-year-old man with posterior mediastinal mass was diagnosed as functional mediastinal paraganglioma during surgical exploration via open thoracotomy in another hospital. The operation was terminated because of severe hypertension when touching the tumor. He was transferred to our center later. After systemic evaluation, the patient was medicated with oral alpha- and beta-blockades, as well as intravenous fluid resuscitation for two weeks. His blood pressure became stable and a second operation was planned. The tumor was removed completely via the thoracoscopic approach, and was finally confirmed as functional paraganglioma by immunohistochemistry. The patient recovered uneventfully after surgery, with no recurrence during one year follow-up visit.

KEYWORDS: Thoracoscopic resection; case report; functional mediastinal paraganglioma
• First time – VATS to remove paraganglioma in a young age
Take home message

• Importance of routine BP measurement even in younger age group

• In Pheochromoctyoma/Paraganglioma
  1. Urine catecholamines - For confirming diagnosis
  2. MIBG Scan - For detecting other lesions
  3. Adequate alpha and beta blockade prior to surgery - For avoiding intra-operative crisis
Acknowledgements

• Dr. Rajiv Santosham – Thoracic Surgeon
• Dr. Rochita – Radiologist
• Dr. Indrani – Nuclear Medicine Consultant
• Dr. Anruradha/Dr. Prabhu – Anaesthetist
• ICU Team

• But Who contributed the most for this Boy?
Who contributed the most for this Boy?

- The pediatrician who checked BP in a child with fever
Thank You!