TREAT THE CHILD NOT THE DIAGNOSIS

By

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2nd yr MD,
Prof. DGS Unit
A 11 yr old male child who had presented with 4 episodes of right focal seizures on 20-11-11, admitted at GVMCH, diagnosed initially as seizure disorder.

CT Scan Brain – focal ill defined hypo dense area in Right parieto occipital lobe,

MRI Brain- multiple focal edema.
DIAGNOSIS: Seizure Disorder / TBM

Child was started on antiepileptic drugs and antitubercular drugs and discharged on 28-11-11
Child readmitted at GVMCH with seizures, fever, multiple body and joint pain on 1-12-11 for which child was referred to ICH&HC for further management.
At ICH&HC, child presented with c/o Generalized myalgia, arthralgia & recent h/o seizures.

O/E: conscious and alert Tenderness present in the Lt.occiput, Lt. shoulder, arm, elbow, bilateral knee, ankle joints, calf & feet.
General examination: normal
Vitals: HR-110/min,
RR-24/min
Systems - NAD
1. Seizure disorder To r/o TBM
2. Rheumatological problem
INVESTIGATIONS

CBC: Normal,
ESR-10.30 mm/hr
RFT - Normal.
LFT - S.Bilirubin<1,
    SGOT-156, SGPT133, ALP-170 KA
Serum CPK-51
RA Factor - Negative
ASO titer - 60 IU
ANA - Negative
TB screening- negative,
Chest X-ray – N,
CT scan brain-N
CSF-Normal Study,
Urine - Alb-3+,
Urine C/S-NG
Urine spot PCR- 7,
24hr urine protein-3276 gm/day
USG Abdomen-
RK-10.5cm, Gr-II echoes, No focal collection
LK-8.3cm, echoes-N
BP was checked

<table>
<thead>
<tr>
<th></th>
<th>UL</th>
<th>LL</th>
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<tr>
<td>Right</td>
<td>Not recordable</td>
<td>160/120</td>
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<tr>
<td>Left</td>
<td>170/140</td>
<td>170/120</td>
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Rt UL brachial & radial pulses - absent, other peripheral pulses felt
No bruit heard over carotids/aortic region
no renal bruit heard
REVISED DIAGNOSIS:

Systemic Hypertension
-Probably secondary to TAKAYASU’S ARTERITIS (PULSELESS DISEASE)
-Seizures probably due to PRES (Posterior Reversible Encephalopathy Syndrome)
DOPPLER STUDY

R subclavian artery - monophasic flow
L renal artery origin - not visualised
L subclavian artery, R renal artery & aorta – Normal

IMPRESSION: Increased acceleration time, decreased acceleration index of L renal segmental artery, s/o segmental artery stenosis
CT ANGIOGRAM

- Juxta ostial occlusion of L main renal artery, L kidney size small compared to right due to vascular stenosis
- Diffuse narrowing of 2nd, 3rd part of R subclavian artery
- Other vessels normal
Cardiac Evaluation:

Echo- Concentric hypertrophy of LV, Normal biventricular function

Ophthalmological evaluation: Normal

DTPA Scan - awaited
Child was discharged with anti hypertensive's (Tab. Methyl Dopa & Tab. Nifedepin) oral prednisolone methotrexate.
• As child had uncontrolled hypertension on followup, Tab Enalapril & Tab Atenolol was added.
• Child is under follow up at rheumatology, nephrology and cardiothoracic OP
• Renal Angioplasty is being planned.
DISCUSSION
HISTORY

• Japanese ophthalmologist, Mikito Takayasu in 1908 described the first case of Takayasu’s arteritis.

• Takayasu described a peculiar "wreathlike" appearance of the blood vessels in the back of the eye (retina).
• Takayasu arteritis is a chronic inflammatory disease of the aorta and its major branches.

• Age- as early as 6 months; in adults of any age, most common is in 2nd to 3rd decade

• Sex- F:M-2:1
INCIDENCE

World wide incidence-2.6 cases/million.
More common in Asians
ICH statistics from 2001 to 2011- 3 cases
characterized by granulomatous inflammation of the aorta and its major branches, leading to stenosis, thrombosis, and aneurysm formation.
Types of Takayasu’s Arteritis

- Type I - Branches of the aortic arch
- Type IIA - Ascending aorta, aortic arch, and its branches
- Type IIB - Type IIA region plus thoracic descending aorta
- Type III - Thoracic descending aorta, abdominal aorta, renal arteries, or a combination
- Type IV - Abdominal aorta, renal arteries, or both
- Type V - Entire aorta and its branches
ETIOLOGY:

• Identical twins
• Association with A10, B5, Bw52, DR2, DR4 & B22.
• Association with tuberculosis.
CLINICAL FEATURES:

<table>
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<tr>
<th>SYMPTOMS</th>
<th>SIGNS</th>
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<tr>
<td>1) The usual presentation is pain in affected site due to claudication</td>
<td>1) Loss of pulses</td>
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<td>2) Weight loss</td>
<td>2) Hypertension</td>
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<tr>
<td>3) Arthalgia</td>
<td>3) Aortic incompetence</td>
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<tr>
<td>4) Fever</td>
<td>4) Bruit</td>
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</table>
• Skin lesions
• Eye involvement
  retinal hemorrhages
  cotton wool exudates
  micro aneurysms of peripheral retina,
  vitreous hemorrhage,
  optic atrophy
### 1990 CRITERIA OF AMERICAN COLLEGE OF RHEUMATOLOGY (3/6)

<table>
<thead>
<tr>
<th>CRITERIA</th>
<th>DEFINITION</th>
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<td>Age of onset</td>
<td>Development of symptoms or findings related to Takayasu arteritis at age &lt;40 years.</td>
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<td>Claudication of extremities</td>
<td>Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities.</td>
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<td>Decreased brachial artery pulse</td>
<td>Decreased pulsation of one or both brachial arteries</td>
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<td>BP difference &gt;10mmHg</td>
<td>Difference of &gt;10mmHg in systolic blood pressure between arms</td>
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<td>Bruit over subclavian arteries or aorta</td>
<td>Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta</td>
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<td>Arteriogram abnormality</td>
<td>Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibro-muscular dysplasia, or similar causes: changes usually focal or segmental</td>
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Ishikawa's Criteria for Takayasu's Arteritis (Modified According to Sharma et al.)

**Three major criteria:**

- Left mid subclavian artery lesion
- Right mid subclavian artery lesion
- Characteristic signs and symptoms of at least one month duration:
Ten minor criteria

• High ESR:
• Carotid artery tenderness
• Hypertension: Persistent blood pressure > 140/90 mmHg in brachial or > 160/90 mmHg in popliteal
• Aortic regurgitation or Anuloaortic ectasia
• Pulmonary artery lesion, Left mid common carotid lesion:
• Distal brachiocephalic trunk lesion:
• Descending thoracic aorta lesion:
• Abdominal aorta lesion:
• Coronary artery lesion:
• Presence of two major or one major and two minor criteria or four minor criteria suggests a high probability of Takayasu's arteritis
INVESTIGATIONS

- CBC - NN Anemia, Leucocytosis, thrombocytosis, acute phase reactants
- LFT - transaminases & hypoalbuminemia
- vwf related antigen
- Anti endothelial antibodies present
- ANA - negative
- RF +ve in 15%
- Ig G,M,A
IMAGING

- Arteriography - invasive angiography or MRA
- Duplex doppler, chest Xray, echocardiography
- CT & MRI – serial examination & diagnosis in early phase
- Gadolinium-enhanced cardiovascular MRI
- Noncontrast, T2-weighted, short inversion imaging recovery (STIR) images
- Gallium-67 radionuclide scan
• CCF due to aortic insufficiency, hypertension, myocarditis
• Aortic aneurysms, thrombus formation & rupture
• Ischemic stroke
• Myocardial infarction
• Hypertension
DIFFERENTIAL DIAGNOSIS

• Acute rheumatic fever
• Juvenile arthritis
• Behcet’s disease
• Cogan’s syndrome
• Relapsing polychondritis
• Ankylosing spondylitis
• Seropositive RA
• Juvenile temporal arteritis
Oral prednisolone 1-2mg/kg × 4-6wks till control of active disease & then tapered over 1 month
Relapse on oral steroids- iv methyl prednisolone weekly doses-30 mg/kg not exceeding 1gm/wk
Methotrexate- daily/weekly/monthly doses have been tried & found useful
Cyclophosphamide-oral/iv
OTHER DRUGS:
• Cyclosporine
• Mycophenolate mofetil
• TNF alpha inhibitors- Etanercept/infliximab
• Rituximab-anti CD20
• Tocilizumab-anti IL-6
SURGICAL THERAPY

- For stenotic lesions - percutaneous angioplasty, stenting
- In severe cases resection of the stenosed segment & placement of graft.
PROGNOSIS

• 50% Achieve remission after 1st course of therapy, but 25% of cases never achieve remission

• 5 yr mortality-35%
TAKE HOME MESSAGE

An unusual presentation of a common disease can be missed but never miss the usual presentation of an uncommon disease
Case 2

Dr. T. Vanathin
2 nd year DCH
Prof. D. G. S UNIT
2nd CASE:

A 11 yr old child admitted with c/o sudden onset weakness –Rt UL & LL with deviation of angle of mouth ×1 day
-no h/o headache, vomiting, seizures

Pondicherry Hospital- Hypertension
CT Brain- Intracranial hemorrhage

Referred to ICH
O/E: Alert & Conscious child

HR-120/min, RR-26/min

BP - UL  LL
Rt 160/120  120/90
Lt 164/122  120/100

Peripheral pulses-
UL- Present
LL- feebly present  DOMIN

• abdominal bruit present
CNS:
Higher Functions- Normal
Motor system- Rt. hemiparesis
Cranial nerve- R Facial palsy (UMN)
Sensory system- Normal

Other Systems- Normal
INVESTIGATIONS:

CBC, ESR, Normal
RFT, S.electrolytes, LFT- Normal
Coagulation Profile- Normal
CRP, ANA- negative
USG Abdomen- L kidney not visualised in Lt Renal fossa
Rt kidney – Grade 1 RPD
ECHO-
Mild concentric hypertrophy of left ventricle,
Discrete obstruction in abdominal aorta at the level of SMA and Renal artery
CT Brain & MRI Brain:
Lt gangliocapsular hemorrhage

Renal Doppler: Discrete obstr.in Abd aorta (level of SMA)
CT Angiogram:

- Narrowing of Abd aorta & thinning of Left Renal artery with Hypoplastic Lt Kidney
- Other vessels normal
DIAGNOSIS

MID AORTIC SYNDROME
MID AORTIC SYNDROME:

- Rare entity affecting abdominal aorta in children & young adults
- Constriction of distal thoracic & abdominal aorta & its branches
- Case report- 200 cases so far
Causes:

• Congenital
• Acquired:
  a) Neurofibromatosis
  b) William’s syndrome
  c) Allagille syndrome
  d) Fibromuscular dysplasia
  e) Retroperitoneal Fibrosis
f) mucopolysaccharidosis

g) foetal alchohol syndrome

h) temporal arteritis

i) takayasu arteritis
• Most common- renal artery involvement (60-90%)
• SMA (20-40%)
• Inf.mesentric artery

RENAral ARTERY:
- inter renal
- suprarenal
- intrarenal
- diffuse
CLINICAL FEATURES:

- Hypertension (MC)
- Lower limb claudication
- Abdominal angina
MANAGEMENT:

Medical management:
- control of hypertension

Surgical management:
endovascular surgery:
- minimal invasive procedure
- not involving the branches of aorta
INVESTIGATIONS:
- doppler study
- CT angiogram
Open surgery:
- involvement of branches of aorta
- aorto-aortic bypass
- patch aortoplasty
- renal and mesentric arterial reconstruction
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THANK YOU