Paroxysmal Sympathetic Hyperactivity in a child with High Grade Glioma post resection surgery

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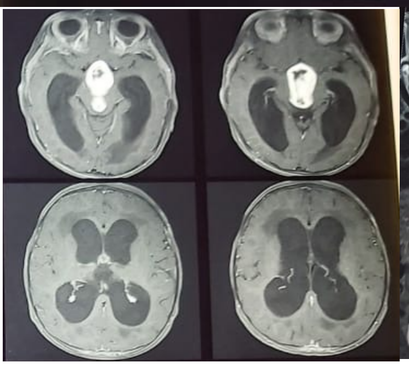
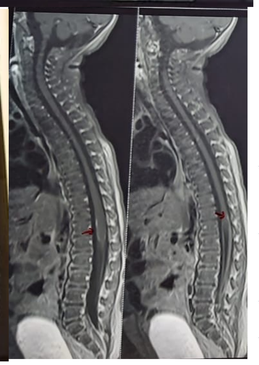
ICNC 2022
17th INTERNATIONAL CHILD NEUROLOGY CONGRESS
ANTALYA, TURKEY | OCTOBER 3-7, 2022

INTRODUCTION

- ✓ An 18-month-old infant was diagnosed with ICSOL & underwent resection for a suspected glioma .
- ✓ The child developed continuous unexplained fever post neurosurgery procedure and despite all investigations & high end antibiotics did not improve.
- ✓ A new onset worsening intermittent dystonia with exaggerated sympathetic response (tachycardia, hypertension) led to suspicion of Paroxysmal sympathetic hyperactivity (PSH).
- ✓ PSH term coined in 2010 however cases have been described since last six decades

NEUROIMAGING PREOPERATIVE





- ➢ Well defined heterogeneously intensely enhancing lobulated intraventricular mass lesion 50 x 24 x 39 mm epicentred in 3rd ventricle extending to Aqueduct & 4th ventricle
- ➢ Moderate hydrocephalus and periventricular CSF seepage, Small enhancing foci seen along surface of spinal cord

MATERIALS & METHODS

- ✓ Uunderwent Endoscopic biopsy + Left anterior transcortical approach with excision of tumor and septostomy
- ✓ Postop pyrexia was noticed from Day 3
- ✓ new onset intermittent dystonic episodes
- ✓ extreme irritability, inconsolable cry, tachycardia and hypertension.
- ✓ Sepsis workup non-contributory.
  - ✓ inflammatory markers,
  - ✓ Cultures
  - ✓ haematological indices were
- ✓ The child was managed with IV broad spectrum antibiotics, IV steroid and supportive management initially.
- ✓ Neuroimaging revealed postop status with no features of raised ICP and any worsening status.
- ✓ Keeping a possibility of PSH
  - ✓ Parenteral Morphine
  - ✓ Oral Propanolol were initiated
- ✓ Symptoms were scored on PSH Assessment Measure (PSH-AM) : Moderate severity

RESULTS

- ✓ Dramatic response in the episodic symptoms over 12-24 hours with child becoming afebrile.
- ✓ Gradually symptoms decreased in frequency as well as intensity
- ✓ Remained event free after 7 days of Morphine
- ✓ Gradually weaned off drugs over 2 weeks.

- ✓ The final histological diagnosis turned out to be Pediatric high grade glioma (WHO-Grade III).

FOLLOW UP

- ✓ On follow up after 6 months child was having disease progression despite adjuvant chemotherapy cycles and radiotherapy.

DISCUSSION

- ✓ PSH is a clinical syndrome characterised by autonomic storm with Intermittent episodes of intense sympathetic drive with hypertension, tachycardia, tachypnoea, posturing
- ✓ Paroxysms due to peripheral catecholamine release manifested as allodynic responses to non-noxious /mildly noxious stimuli
- ✓ Described mostly after severe Traumatic & Ischemic brain injury.
- ✓ PSH due to non TBI are described a few, encephalitis (TBM)
- ✓ Underecognised entity and usually treated as sepsis because of fever in post op cases.
- ✓ Pathophysiology: disconnection of cortical inhibitory centers (insula & cingulate cortex) with the caudal excitatory centers.
- ✓ Goals of therapy
  - ✓ Avoid triggers if identified
  - ✓ Mitigate excessive symoathetic outflow
  - ✓ Supprt vital organs
- ✓ Opioids
  - ✓ Morphine first line Rx
  - ✓ Fentanyl patch
- ✓ A2 agonists
  - ✓ Clonidine
- ✓ B blockers
  - ✓ Propranolol
  - ✓ Labetalol
- ✓ Neuromudulators: baclofen/gabapentin
- ✓ IV Anesthetics(Propofol)

- ✓ The episodic nature of this sympathetic hyperactivity clubbed with unexplained fever led to a suspicion of PSH in the index case
- ✓ The child was managed conservatively initially.
- ✓ IV Morphine and Oral Propranolol resulted in dramatic therapeutic response and stoppage of unnecessary high end antibiotics.

FINAL DIAGNOSIS

PEDIATRIC HIGH GRADE GLIOMA (WHO III)

POST OPERATIVE STATUS WITH

PARAOXYSMAL SYMPATHETIC ACTIVITY

CONCLUSIONS

- ✓ An unexplained fever in absence of infective aetiology in a child with TBI / post neurosurgical status should raise a suspicion of PSH.
- ✓ Intermittent episodes of intense sympathetic drive with hypertension, tachycardia, tachypnoea, posturing and dystonia in absence of raised ICP also may add to the cluster of symptoms of PSH.
- ✓ Early recognition and timely intervention with IV Morphine /Clonidine /Propranolol may result in favourable outcome and can help avoiding unnecessary Antibiotics.

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