RHINO ORBITO CEREBRAL MUCORMYCOSIS – IN A CHILD WITH DKA.
(A black sweet tragedy!!)

By,

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CASE SUMMARY

- 9 yr old, developmentally normal female child was admitted.
- C/o, Fever
  - Breathlessness
  - Abdominal pain x 2 Days
  - Vomiting
  - Polyuria
- O/E, Child was drowsy & tachypnoiec.
- Multiple furuncles over the face.
- CVS,RS,Abd-N.
- CXR-N; CBG-High.
- Urine ketones Positive.
- ABG-Severe wide anion gap metabolic acidosis.
Diagnosed as a case of New onset type I Diabetes with DKA.

Started on DKA fluid management with insulin infusion.

Anti edema measures were started due to persistent ALOC
Course of events

- Next day child sensorium improved.
- Acidosis improved but had high blood glucose.
- At 48 hrs of admission child became irritable; acidosis worsened.
- Had a dark colored bloody discharge from right nostril.
- Inj. Amphotericin B started, suspecting Mucormycosis clinically.
- ENT opinion obtained; no nasal mass & palate was normal.
- In the evening of D3, her right pupil was dilated and sluggishly reacting.
- On D4 child had Right eye proptosis, Ophthalmoplegia with dilated non reacting pupil. Fundus-Normal. Left eye-Normal.
- Black necrotic lesions developed over palate.
- Neurologist opinion obtained.
- No weakness of limbs.
- CT Brain- Normal
- CT PNS-Rt Maxillary sinusitis
- DNE planned.
- DNE done on D5;
- Necrotic eschar removed from nasal cavity.
- Septal perforation +
- Antrostomy done.
- Ethmoidectomy done.
- Characteristically *NO BLEED*.
On D6, developed right facial palsy with left hemiparesis.
Poor respiratory efforts- Child intubated.
CT Brain-frontal & cerebellar infarct.
CT Orbit- normal.
On D7, blood discharge from both nostril.

DEM absent, hypotonia, reflexes absent.

CT Brain-massive infarct of temporal, frontal & parietal infarct.

MRI could not be done.
On D9, child expired.

- Post mortem LP was done.
- Unusual color - ORANGE-RED??
- ACELLULAR
- Prot-1250; Sug-35.
HPE

- Wide angle, broad aseptate hyphae.
- Hyphae inside blood vessels.
- Culture-**RHIZOPUS**.
**Discussion**

- **Mucormycosis** is an acute, frequently fatal, opportunistic fungal disease.
- **Etiology:**
  
  Mucorales- *rhizopus* (MC), mucor, absidia, apophysomyceae, rhizomucor.
  
- **Predisposing factors:**
  
  DKA, prolonged neutropenia, HSCT, Malignancy, chronic corticosteroid, deferroxamine therapy.
Why target DKA patients?

- Chemotactic activity of neutrophils decreased.
- Ketosis diminish reactive oxygen species production.
- Acidosis disrupts transferrin binding to iron.
- As Advanced Glycosylation End products increases, transmigration of PMNs decrease.
Rhinocerebral

**Route of entry**: inhalation and deposition of spores in nose & PNS.

- Begins with facial pain, numbness followed by sinusitis, dark & bloody nasal discharge.
- Spreads from Ethmoid sinus to orbit - external ophthalmoplegia, proptosis.
- Brain invasion - meningoencephalitis / abscess
- Rapid progression - *hrs to days.*
Investigations

- Direct examination – KOH preparation.
- HPE – Hyphae-broad, thin walled, aseptate branching at rt angles; Angio invasion.
- Culture-Positive in <50% cases; Black colonies
- Imaging-CT, MRI
Treatment

- Amphotericin B deoxycholate - 1-1.5mg/kg/d.
- LAMB - 5-10mg/kg/d.
- ABLC - 5-7.5mg/kg/d.
- Echinocandin + lipid polyenes.
- Posaconazole.
- Deferasirox (20mg/kg/d) + lipid polyenes.
- Surgical debridement
Outcome & Prognosis

- High mortality (>60% with cerebral involvement; 15% without cerebral involvement).

- Poor prognosis:
  - delayed diagnosis
  - inadequate surgical debridement
  - spread beyond sinonasal cavity
  - cerebral involvement.
Conclusion

- **High index of suspicion** is critical for diagnosis of this *unusual complication of DKA*.
- Early diagnosis and a joint medical & surgical approach increases survival.
Acknowledgement

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