A Case Report: A Battle With An Angioinvasive Fungus

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Introduction

Mucormycosis is an angioinvasive fungal infection characterized by various clinical syndromes, with the most common being rhino-orbito-cerebral infections. It is particularly seen in immunocompromised hosts, including patients with diabetes mellitus, hematologic cancers, and solid organ or hematopoietic stem cell transplants.[1] With an estimated incidence of 3 cases per million in the United States of America, its rarity has limited the ability to perform large, randomized clinical trials, making actual case reports a necessity for research.[2] Its diagnosis triggers a medical and surgical emergency, with delay in treatment increasing morbidity and mortality. Thus, education on its presentation and management is crucial to improve patient outcomes.

Case Description

- 58-year-old relatively healthy male presented to ORL inpatient floor as a direct transfer from an outside hospital for worsening left orbital swelling.
- Over 4 days, symptoms progressed to left-sided ptosis, unilateral orbital drainage, vision loss, and epistaxis.
- Had taken oral steroids and IM Methylprednisolone for weeks prior, to avoid adverse effects from his COVID-19 vaccination.
- Labs showed leukocytosis with WBC of 34.9 with left shift and severe hyperglycemia with glucose of 337.
- Urgent radiology readings of outside CT scans were requested.
- Ophthalmology and Neurology services were consulted.
- Empirically placed on Unasyn and Vancomycin.

Physical Examination

- Vitals: 37.4C, BP 159/110, P 98, R 16, SpO2 99% on room air
- General: A&Ox3, sitting comfortably and in no acute distress.
- HEENT:
  - Left eye: frozen and proptotic, without EOMs in all directions, eyelid edematous and occludes eye at rest. No direct or accommodation pupillary reflex. Right eye: unremarkable.
  - Nasal Cavity: Severe septal and turbinate crusting, with evidence of prior epistaxis and dryness. No mass, lesion, pus or discharge.
- Respiratory: Clear to auscultation bilaterally.
- Neurology: Complete vision loss of left orbit with CN 3, 4, 6 deficits. Mild V1 CN 5 sensation deficit, due to forehead swelling. Terminal branches of CN 7 intact.

Workup

CT imaging showed left medial and superior infraorbital inflammation with proptosis and left ethmoid and maxillary sinusitis.

MRI Brain/Face with contrast demonstrated:
- Left optic nerve infarct/ischemia
- Left preseptal and orbital cellulitis with associated exophthalmos
- Mucosal disease involving left ethmoid and left maxillary sinus
- Hypoenhancement of left inferior turbinate consistent with invasive fungal sinusitis

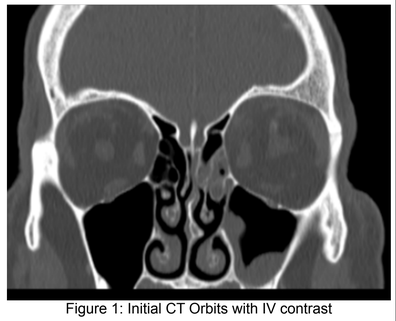

Figure 1: Initial CT Orbits with IV contrast

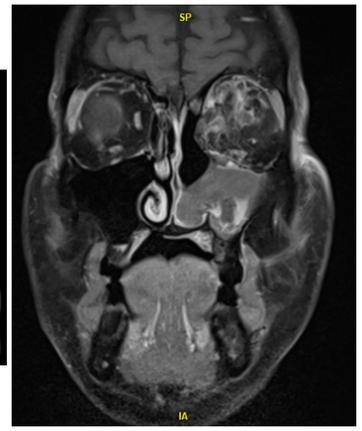

Figure 2: Initial MR face with IV contrast

Taken to OR for endoscopic sinus surgery and debridement
- Intraoperative findings show grossly evident fungal hyphae.
- Frozen tissue pathology confirmed angioinvasive fungal sinusitis secondary to *Rhizopus* mucormycosis.

Hospital Course

- Initiated on IV lipid Amphotericin B, Meropenem, and Vancomycin. Posaconazole later added as salvage therapy.
- Hyperglycemia controlled with sliding scale and long-acting insulin.
- Course complicated by:
  - Cavernous sinus thrombosis leading to left orbital exenteration
  - Acute mentation changes with right hemiparesis due to SAH
  - Left internal carotid artery occlusion status post left extra-cranial to intracranial carotid bypass surgery
  - Multiple infections, brain infarcts, cerebral vasospasms, and hydrocephalus

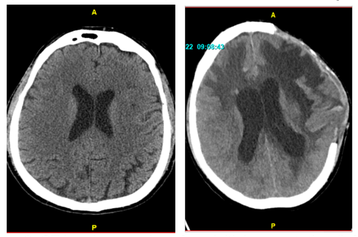

Figure 3: CT head comparison from initial presentation (left) to progression of hydrocephalus and intraparenchymal hemorrhages (right)

Conclusion

It was suspected that the cause of his condition was likely related to his weeks of steroid use prior to presentation, making him quite immunocompromised. Given the patient's poor prognosis, his family transitioned him to comfort care and elected for him to be discharged to hospice.

Discussion

- Mucormycosis is a serious but rare fungal infection caused by a group of molds called mucormycetes.[4]
- Nearly half of cases present as Rhino-Orbital-Cerebral Mucormycosis (ROCM).[4]
- Clinical presentation includes non-specific symptoms, which may appear like those of sinusitis.[2]
- Due to its angioinvasive characteristics, can progress to vessel wall infiltration leading to thrombosis and nerve dysfunction within hours to days.[2]
- Serum studies, CT and MR imaging are diagnostically useful
  - Tissue biopsy required for definitive diagnosis.[1]
- Treatment: early anti-fungal administration, surgical debridement, and rapid reversal of immune-suppressive factors.[3]
  - 1st line: IV Amphotericin (liposomal) 5-10 mg/kg/day for 3-4 weeks.
  - IV or oral Posaconazole or Isavuconazole used for step-down therapy and/or with Amphotericin.
- Frequently a life-threatening infection - mortality is high (>30-50%).[1]
- Given its poor prognosis, a suspicion or diagnosis of mucormycosis requires quick intervention to improve overall outcome and survival.

References

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