Episodic motor posturing and unilateral oval pupil from cryptococcal meningitis

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Abstract

Motor posturing in acutely comatose patient is suggestive of an evolving brain herniation. In the presence of a unilateral pupillary abnormality, a metabolic or drug induced coma can be excluded. Neuroimaging studies in such cases will likely reveal underlying mass or acute ventricular obstruction. We recently encountered a patient who presented in stupor with repetitive motor posturing and a transient right oval pupil to light stimulus. Surprisingly, an urgent brain CT revealed only mild global atrophy for age. Further evaluation disclosed cryptococcus meningitis as the underlying cause.

Keywords: Meningitis, fungal infections, cryptococcal meningitis, increased intracranial pressure, oval pupil

Case Report

A 43-year-old diabetic woman presented to the emergency room after having been found unwell by her roommate. She was febrile, hypertensive, and unresponsive, requiring ventilatory support. Semirhythmic adduction-flexor posturing of upper extremities with lower extremity extension were observed for a few seconds every 5-10 minutes without an obvious trigger. The right pupil was circular, but with light stimulus constricted minimally from 4mm to an obliquely-oriented 3mm long-axis oval configuration. The left pupil constricted normally from 4 to 2mm, remaining circular. Extra ocular movements induced with head-rocking response ("doll’s head maneuver") revealed symmetric ocular excursion both vertically and horizontally. Funduscopic examination was normal without disc edema. Serum glucose was elevated at 611 mg/dL. Urgent brain CT revealed only mild global atrophy without mass effect.

Initial lumbar puncture (LP) revealed WBC of 4 /mm³, protein 77 mg/dL, and glucose 238 mg/dL but an opening pressure was not measured. Cryptococcal antigen was detected at 1:16 titer with negative India ink stain. Intravenous amphotericin and flucytosine were started. Brain MRI was unrevealing. EEG showed generalized slowing. Repeat LP on the second day revealed WBC of 57 /mm³ and normal opening pressure of 190 mmHg. Third LP on the 10th day revealed WBC count of 23 /mm³ with an elevated opening pressure of 300 mmHg. Serum serology for HIV was negative.

On the second hospitalized day, the motor posturing ceased with residual mild increased flexor tone of upper extremities. On the third hospitalized day, the right light-pupillary response normalized. A week later the patient responded to simple questions and was able to follow “show me your right thumb and move your left foot”. Motor tone normalized.

DISCUSSION

Initially, the presence of an asymmetric pupillary response made hyperglycemia unlikely as the sole cause. The combination of a fever on admission, episodic motor posturing, and an unrevealing CT scan led to an LP and the diagnosis of cryptococcal meningitis (CM).

Coma, repetitive motor posturing and asymmetric pupils localize to diencephalon or rostral brainstem, however both CT/MRI were unrevealing without mass or 3rd ventricular ballooning. Although generally described as meningitis, cryptococcal CNS involvement frequently results in secondary parenchymal or cranial nerve infiltration sometimes even resulting in a residual intracranial cryptococcoma.

Fisher was first to emphasize the significance of an oval pupil in an acute clinical setting based on the observation of 17 patients with mostly underlying cerebrovascular disorders. He hypothesized that an oval shaped pupil represented...
a transient early injury to the oculomotor nerve complex. His patients had a larger oval shaped pupil than the unaffected side. In our case, the oval pupil became obvious only in response to a light-stimulus resulting in a sluggish segmental constriction. Prior to the light stimulus, pupils were of equal size and circular in shape. Complete resolution of the aberrant pupillary response by the third day with good functional outcome suggests that light-induced sluggish oval pupil may represent a forme-fruste of the oval pupil Fisher described.

Distinctly unusual intermittent recurrent third cranial nerve palsy lasting minutes have been documented in CM. Keane reported two cases and suggested spikes of intracranial pressure elevation may be responsible if the cranial nerve is already infected. This astute observation was based on a transient response to mannitol infusion in the absence of radiologic herniation. Anatomic proof was provided by Azan et al., on yet another case of intermittent oculomotor nerve palsy, who on autopsy demonstrated parenchymal and cranial nerve infiltration by Cryptococcus. In our case there was no consistent correlation of the episodic posturing with the light-induced oval pupil on the first day, but resolution of both aberrant pupillary response and episodic posturing over several days following the initial LP and initiation of treatment suggests intracranial pressure elevation as a likely underlying contributing cause. Absent motor posturing and pupillary changes despite an elevated opening pressure at around the time of the third LP underscores the complex relationship of intracranial pressure, CM and treatment effect in eliciting these unusual clinical signs.

DISCLOSURE

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Conflict of interest: None

REFERENCES