Pearls & Oysters: Oval pupil

Two observations

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PEARLS

- The presence of an oval pupil usually localizes the neurologic problem to the midbrain.
- Partial damage to the preganglionic parasympathetic pupillary fibers resulting in segmental relaxation of iris sphincters is a possible mechanism for oval pupil.

OY-STERS

- Oval pupil may also be due to primary ocular diseases such as trauma, acute glaucoma, and eye surgery.
- Oval pupil is most commonly seen in comatose patients following acute midbrain injury with elevated intracranial pressure and tissue shift, but it can also be observed in patients with midbrain stroke.
- Oval pupil may be unilateral, bilateral, or alternating, and can be reactive or nonreactive to light. Although typically a temporary and transitional finding, it may persist.

PATIENT 1 A 79-year-old man with an asymptomatic left carotid stenosis became comatose immediately following left carotid stenting. He had a left surgical, round, 7 mm, nonreactive pupil, and a right oval, 6 × 5 mm, nonreactive pupil (figure, A). CT showed an acute hemorrhage within the left midbrain peduncle with subarachnoid and intraventricular hemorrhage (modified Fisher scale grade 4) (figure, B). Over the next hour, he lost all brainstem reflexes except spontaneous breathing, which lasted for another 18 hours. After he became brain dead, the oval configuration stayed for 36 hours.

PATIENT 2 A 55-year-old woman with a history of neuroendocrine carcinoma of the colon with lung metastases was found unconscious. She was neurologically intact 30 minutes prior to being found unconscious. On examination she was stuporous with her right pupil dilated, nonreactive, and oval; there was right exotropia. There were no other neurologic abnormalities. CT angiogram of the head and neck was unrevealing. MRI of the

brain showed a hyperintense lesion on the diffusion-weighted imaging sequence in the right paramedian midbrain region (figure, C) and bilateral medial thalamic nuclei consistent with an acute ischemic stroke.

DISCUSSION There are a plethora of monikers for oval pupil, including ectopia pupillae,¹ corectopia, midbrain corectopia,² and tadpole pupil. To simplify the terminology, Fisher coined the term oval pupil.³ A dilated, oval pupil can also be seen secondary to ophthalmic procedures like cataract surgery, though the pupil may still react to light in that situation. Acute glaucoma may present with a fixed, mid-dilated, and oval pupil with painful red eye, blurred vision, and complaints of halos around lights.

Oval pupil is usually considered transitional in nature. We noted a persistent oval pupil in a patient who fulfilled all criteria of brain death. In one study, 13 of 15 patients had high intracranial pressure (>20 mm Hg) and pupils in 9 patients improved to normal pupil size, 4 worsened to fixed round pupil (3 large and 1 small), and 1 remained oval.⁴

We also noted MRI documentation of a midbrain infarct as a cause of oval pupil. There has been some speculation about the mechanism of an oval pupil. One of the first studies on this type of pupil described 3 patients with bilateral ectopia pupillae with midbrain lesions. It proposed midbrain injury resulting in loss of segmental contraction of pupillary dilator or segmental relaxation of pupillary constrictor as the possible mechanism for pupillary abnormality.¹ Others found bilateral lesions of the rostral tectus, periaqueductal gray lateral midbrain tegmentum, and ventral lateral midbrain on autopsy. One patient had alternating transient nonreactive oval pupil during coma. The authors used the term midbrain corectopia and their proposed mechanism was a random inhibition of portions of Edinger-Westphal nuclei and damage to preganglionic parasympathetic innervation causing segmental relaxation of the iris sphincters.²

In his oval pupil study, Fisher described 17 patients: 16 with acute neurologic injury and 1 with recovering isolated oculomotor palsy. His explanation was injury to the oculomotor nucleus or nerve at the midbrain level with partial paresis of the parasympathetic fibers

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(A, B) The right oval pupil seen in the first patient after an acute left midbrain hemorrhage associated with severe subarachnoid and intraventricular hemorrhage. (C) MRI brain, diffusion-weighted image, shows a right paramedian hyperintense lesion consistent with an acute ischemic stroke in the second patient who had an ipsilateral oval pupil. The right frontal hypodense lesion represents a metal artifact from the previous clipping of an anterior communicating artery aneurysm and associated encephalomalacia in the right frontal lobe.

leading to an unopposed sympathetic action in the paralyzed iris leading to an oval pupil.³ The MRI finding in our second patient is consistent with this localization.

AUTHOR CONTRIBUTIONS

Manoj K. Mittal: conceptualization of study, data gathering and analysis, first draft and revision of manuscript, and final approval of the manuscript. Alejandro A. Rabinstein: revision of manuscript and final approval of the manuscript. Eelco F. Wijdicks: conceptualization of study, revision of manuscript, and final approval of the manuscript.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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