Benıgn Episodic Unilateral Mydriasis

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Abstract: Benign unilateral episodic mydriasis, is a related entity in which intermittent episodes of pupillary asymmetry mydriasis occur in young adults, usually women. Each episode is self-limited, and the situation has not been associated with any systemic or neurologic disease. Usually related to migraine, some authors classify it as a limited form of ophthalmoplegic migraine. Selective serotonin reuptake inhibitors are effective in the treatment of depression, generalized anxiety disorder and a recurrently regarded as the pharmacotheraphy of choice. Probabl central inhibitory effects of SSRI extend to dilation-correlated neurons that inhibit the E.W. nucleus, the administration of SSRI would also suppress BUEM. The underlying physiopathology is not clear and may involve either parasympathetic deficiency or sympathetic hyperactivity affecting the iris. We consider that intermittent inhibition of dilation-corelated neurons in the Edinger West pal nucleus are responsible for the pupillary dilatations seen in BUEM.

Keywords: Episodic, unilateral and mydriasis.

1. INTRODUCTİON

Benig unilateral episodic mydriasis, is a related entity in which intermittent episodes of pupillary asymmetry mydriasis occur in young adults, usually women. Each episode is self-limited, and the situation has not been associated with any systemic or neurologic disease. Usually related to migraine, some authors classify it as a limited form of ophthalmoplegic migraine, although some cases have been described with no accompanying headache [1, 3]. The underlying physiopathology is not clear and may involve either parasympathetic deficiency or sympathetic hyper-activity affecting the iris. We consider that intermittent inhibition of dilation-corelated neurons in the Edinger Westpal nucleus are responsible for the pupillary dilatations seen in BUEM [2].

2. CASE REPORT

A thirty nine year old female patient presented us with intermittent enlargement in her left pupil for the last five months, explaining that this condition could last between 15-20 minutes. She has been described with no accompaying headache. Also she complained of anxiety, irritability and insomnia recently. Her neuro-logical and physical examination was unremarkable. Her complete blood count, metabolic profile, and thyroid function test were unremarkable. She had normal head and neck magnetic resonance imaging and angiography, electroencephalography, and visual evoked potential. She was started on essitalopram, initially 20mg daily, which led to significant improvement of her pupil disturbance.

3. DİSCUSSİON

Benign unilateral episodic mydriasis, is a descriptive term for recurrent episodes of isolated unilateral mydriasis occurring in young adults, usually women. The mydriasis typically appears in the same eye but can alternate sides. Mydriasis usually lasts for several hours but may persist for days [4, 5]. Benign episodic mydriasis probably represents a heterogeneous group of disorders, including migraine-associated anisocoria and physiologic anisocoria, with different mechanisms that result in transient and episodic anisocoria [6]. The pupil is innervated by brain structures involved in both cognitive and emotional processing. Inhibition of the pupillary constrictor muscle occurs through parasympathetic innervation of the Edinger-Westphal nucleus, which receives extensive inputs from cortical and limbic regions. Edinger Westphal nucleus is the part of the oculomotor complex that is the source of the parasympathetic preganglionic motor neuron input to the ciliary ganglion, through which it controls pupil constriction and lens accommodation [9, 10]. EW has two anatomically and function would be designated the Edinger-Westphal preganglionic (EWpg) population and the peptidergic neurons with consumption and stress-related functions would be designated as the Edinger-Westphal centrally projecting (EWcp) population. EW neurons are spontaneously active pacemaker cells that, when devoid of synaptic input, have a high intrinsic firing rate, resulting in pupillary constriction [11, 12]. It has been demonstrated that short axon neurons with bursting firing patterns are present in the periaqueductal gray matter (PAG) that are inhibitory to E.W. nucleus. These dilation-correlated neurons produce pupillary dilation when stimulated electrically. While the exact mechanism of opioid induced miosis is not completely known, there is

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evidence that opioids increase the activity of the E.W. nucleus by exerting a depressant effect on inhibitory neurons that project into this nucleus [12, 13]. Such disinhibition results in miosis, due to the high intrinsic firing rate of the E.W. neurons. We consider that intermittent inhibition of dilation-correlated neurons in the E.W. nucleus are responsible for the pupillary dilatations seen in BUEM [14, 15]. The mechanism of unilateral episodic mydriasis might be related to ipsilateral activation of the autonomic network in periaqueductal gray matter. The periaqueductal gray (PAG), a midbrain structure which regulates stress or anxiety-related behavior has robust 5-HT fibers and reciprocal connections with the hypothalamic–pituitary–adrenal axis [16]. Serotonin (5-HT) modulates pain and anxiety from within the midbrain periaqueductal gray matter. The pupil is innervated by brain structures involved in both cognitive and emotional processing. Inhibition of the pupillary constrictor muscle occurs through parasympathetic innervation of the Edinger-Westphal nucleus, which receives extensive inputs from cortical and limbic regions [17, 18]. Depressed adults exhibit altered neural and physiological responses to emotional material, particularly greater activation in limbic regions, such as the amygdala, and dysfunction in prefrontal cortical systems that modulate limbic activity [19]. There is growing evidence that depressed adults exhibit greater pupil dilation to negative emotional words compared with never depressed adults. With regard to anxiety one recent study found that anxious youth exhibited increased sustained pupil dilation in response to fearful faces compared with nonanxious youth [20]. Serotonergic (5HT) drugs are widely used in the clinical management of mood and anxiety disorders. If the central inhibitory effects of SSRI extend to dilation correlated neurons that inhibit the E.W. nucleus, the administration of SSRI would also suppress BUEM [21, 22]. These findings links activation of the PAG and EW nucleus involved in inhibition to a parasym-pathetically dominated autonomic response profile that is characteristic of pupil dilation. Selective serotonin reuptake inhibitors are effective in the treatment of depression ,generalized anxiety disorder and are currently regarded as the pharmacotherapy of choice [23, 24]. Finally, our findings may provide new directions for research into the pathophysiology of BUEM.

CONCLUSİON

Benign episodic mydriasis probably represents a heterogeneous group of disorders, including migraine-associated anisocoria and physiologic anisocoria, with different mechanisms that result in transient and episodic anisocoria [7, 8]. The pupil is innervated by brainstructures involved in both cognitive and emotional processing. Inhibition of the pupillary constrictor muscle occurs through parasympathetic innervation of the Edinger-Westphal nucleus, which receives extensive inputs from cortical and limbic regions [24, 25]. We consider that intermittent inhibition of dilation-correlated neurons in the E.W. nucleus are responsible for the pupillary dilatations seen in BUEM. The mechanism of unilateral episodic mydriasis might be related to ipsilateral activation of the autonomic network in periaqueductal gray matter. The periaqueductal gray (PAG), a midbrain structure which regulates stress or anxiety-related behavior has robust 5-HT fibers and reciprocal connections with the hypothalamic–pituitary–adrenal axis [26]. Serotonin (5-HT) modulates pain and anxiety from within the midbrain periaqueductal gray matter. Activation of the PAG and EW nucleus involved in inhibition to a parasympathetically dominated autonomic response profile that is characteristic of pupil dilation [27, 28].

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