Tonic Pupil Following the Use of Dermatoscope

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Adie's tonic pupil is a benign condition supposed to be due to a damage of the parasympathetic innervation of the eye. It may cause considerable anxiety in the patient and lead to extensive but futile investigations. Here, we describe a 34-year-old dermatologist who developed a tonic pupil just after she had started to use intensively a 6 light-emitting diode dermatoscope. We hypothesize that frequent, repetitive exposures of one eye to intense light might fatigue the pupillary reflex and trigger the appearance of a tonic pupil.

Key Words: Adie, Adie's pupil, Dermatoscopy, Tonic pupil

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A 34-year-old female dermatologist was referred to Neurology Department for a recent onset pupillary inequality. Her history was unremarkable, except for a transient (~2 hours) episode of blurred vision of the right eye few weeks before, at the end of her first day of repetitive and strenuous use of a 6 light-emitting diode (LED) illumination dermatoscope with that eye. In the following weeks, she used the same dermatoscope for her work, in the context of a screening program for neoplastic skin lesions, which required the evaluation of 30 patients in 3 hours for 2 days/week. At our observation she was completely asymptomatic. A neurological examination was normal, except for a marked anisocoria due to a midriatic right pupil with an absent reaction to light and to near vision (Fig. 1A). Blood exams, including VDRL, CA125, antinuclear, anti-extractable nuclear antigen, and anti-neuronal antibodies were unremarkable. Three Tesla cerebral and ocular MRI, and angio-MRI were negative. In spite of a preserved extraocular muscle function, these investigations were done to rule out a compression of the third nerve in the subarachnoid space, caused by an unruptured intracranial aneurysm or other lesions. A diagnosis of right tonic pupil was suspected.

After approximately one and a half years, the patient underwent a formal ophthalmological examination. Visual acuity with correction was 10/10 in both eyes and no afferent pupillary defect was detected. The intraocular pressure was 12 mm Hg in both eyes. The anisocoria, greater in light, was still present (Fig. 1B). In fact, in bright illumination the right pupil was 5 mm in diameter, while the left was 2 mm; in dim light the right pupil slowly dilated to 7 mm, while the left pupil readily dilated to 5 mm. Moreover, a sluggish reaction to near vision

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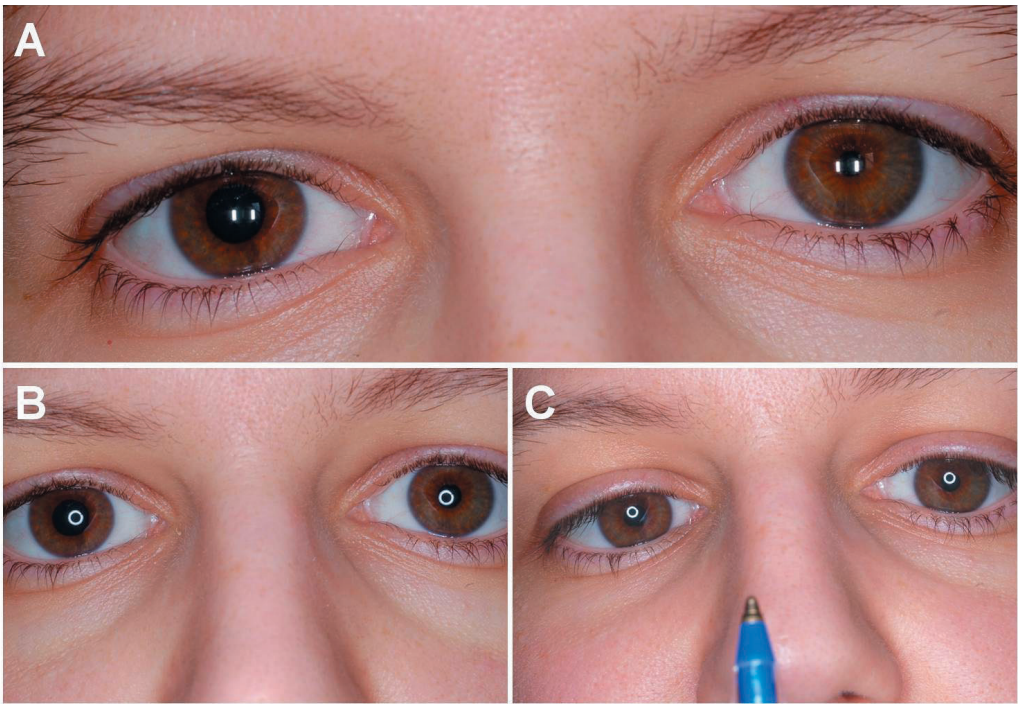

Figure 1. (A) Dilated right pupil unresponsive to light and near vision, as observed in a tonic pupil of recent onset. Indeed, according to the hypothesis of Irene E. Loewenfeld's 6,7), the pupillary response to near is restored by aberrant regeneration, which takes a couple of months after the injury to the ciliary ganglion (see figure 2 for details). At re-evaluation after one and a half year, the right pupil is still larger than the left one in bright illumination (B), but it slowly reacts to accommodation, and after at least 30 seconds anisocoria is no more detectable (C).

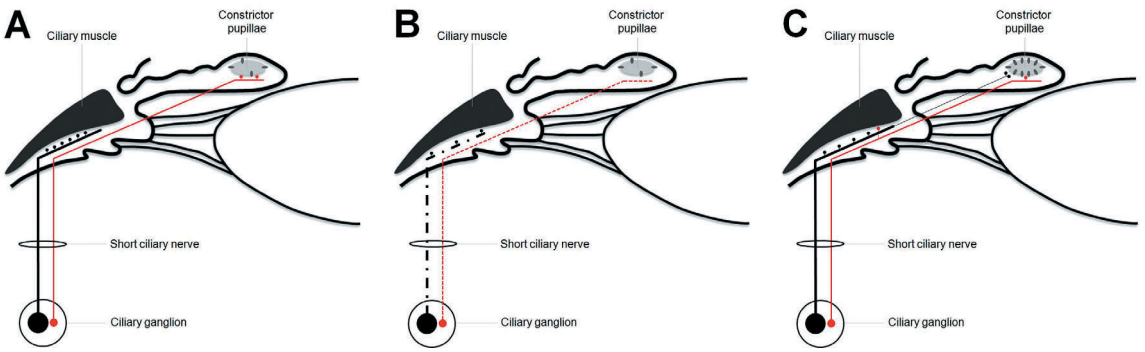

Figure 2. Cartoon illustrating the pathophysiology of the tonic pupil. (A) Around 95% of the parasympathetic fibers originating in the ciliary ganglion innervate the ciliary muscle (black), while only 5% innervate the constrictor pupillae (gray), according to their unequal masses. (B) Damage to the ciliary ganglion (CG) or to the short ciliary nerves (SCNs) results in a tonic pupil, which is initially characterized by a fixed, dilated pupil with loss of accommodation. (C) A couple of months after the injury to the CG or SCNs, surviving ganglion cells re-grow and randomly re-innervate the ciliary muscle and the constrictor pupillae. Some accommodative neurons (black) will innervate the constrictor pupillae, while some light-reaction neurons (red) will innervate the ciliary muscle. Due to the great number of fibers originally intended for the massive ciliary muscle, this aberrant regeneration will result in a pupil that constricts to near stimulation, but no accommodation will be detected with the exposure to light[6,7]. Moreover, very diluted solutions of pilocarpine, that have no effect on normal iris, will cause the constriction of the iris because of a denervation supersensitivity, due to an increased number of acetylcholine receptors (small gray oval).

(not detected in the first examination) was then observed in the right eye (Fig. 1C). A slit lamp examination of the anterior segment showed segmental constriction (Czarnecki's sign) of the iris border of the right eye, and was normal in the left eye. The pilocarpine 0.125% test demonstrated supersensitivity of the right pupil, but no effect on the left pupil. Hence, a diagnosis of Adie's tonic pupil was confirmed.

Adie's tonic pupil is a benign condition usually occurring in young women and supposed to be due to a neuron degeneration in the ciliary ganglion, which produces denervation of the sphincter muscle of the iris[1,2], also called constrictor pupillae. First described by W.J. Adie in 1932[3], its etiology still remains obscure, although it has been reported in the context of viral and autoimmune diseases, including paraneoplastic syndromes. It should be distinguished from other causes of benign unilateral mydriasis, including: 1) the "springing pupil", which is a not permanent, but episodic pupillary dilatation occurring with migraine or even in otherwise healthy young adults; 2) the transient unilateral mydriasis that may follow the use of anti-motion sickness patches or the exposure to cosmetics and perfumes, which contain agents that may affect the pupil size (e.g., scopolamine)[1]. Notably, tonic pupil may cause considerable anxiety in the patient and lead to extensive, unnecessary investigations.

Here we report a case of Adie's pupil on the right side, which developed in a young dermatologist few weeks after the beginning of her work as dermatoscopist. The dermatoscope she used had a remarkably brilliant light beam, emitted by LEDs. It is difficult to explain the mechanism, if any, which may link the frequent and intermittent exposure of one eye to intense light with a damage of the homolateral ciliary ganglion. Nevertheless, it should be considered that the pupillary responses to light may be fatigued and disappeared with repeating light stimulation several times[4,5]. Hence, we hypothesize that highly repetitive exposure of one eye to intense light beam such as that of a LED dermatoscope, fatiguing the pupillary light reflex, might have brought out an underlying, up-till-now silent, dysfunction of the efferent branch of this reflex in that eye.

The association between appearance of pupil abnormalities and professional use of dermatoscope has never been reported thus far. It may be completely coincidental, and further observations might lead support to the hypothesis that excessive use of a dermatoscope, or similar instruments, could predispose to the emergence of a tonic pupil.

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