An interesting case of simultaneous bilateral Adie’s tonic pupil

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Abstract:
A tonic pupil is usually caused due to parasympathetic denervation of the iris sphincter muscles. It’s usually seen in females and in 80% cases it’s unilateral, and 4% of these cases progress to bilateral involvement. Adie’s tonic pupil has been reported in literature to have multiple syndromic associations. Simultaneous bilateral occurrence of Adie’s tonic pupil in an asymptomatic healthy adult male is sparsely reported in literature. The diagnostic dilemma in the setting of a busy clinical practice is henceforth discussed.

Keywords:
Adie’s, bilateral, tonic

Introduction

Patients with tonic pupils have accommodative symptoms or photophobia which may get misdiagnosed in a busy clinical practice. Before 1924, there had been forty case reports of tonic pupils in literature, and twenty of the patients were women. In subsequent years, Moore, Holmes, and Adie added 46 cases to literature and 42 of these were women. Various case reports assembled from literature showed this disorder to be predominant in females (a ratio of 2.2/1). In 20% of the 220 cases from literature, both eyes were involved.

Case Report

A 41-year-old male presented with progressively worsening glare, visual discomfort, especially in sunlight, and difficulty in doing near work for the past 6 months. Medical history was insignificant, except that he had been diagnosed with impaired glucose metabolism 2 years ago and was on lifestyle modification regime for the same. There was no other relevant positive clinical or family history.

On examination, he was found to have 20/20 vision and N5 vision (with effort) in both eyes. However, light-near dissociation was noted. The pupils were dilated at presentation, 7.5 mm (OU), in dimension [Figure 1a], and showed a vermiform segmental contraction, in the superior quadrants, reducing to only 7 mm, to bright light. Pilocarpine (0.1%) was instilled in both eyes every 5 min for 45 min, following which pupillary constriction to 3 mm (OU), was noted [Figure 1b]. Rest of the clinical examination was unremarkable. Extensive lab investigations were done, including serology for HIV and syphilis, all of which were within normal limits. The patient was prescribed a refractive correction of +1.00 diopter sphere for his accommodative paresis leading to alleviation of his symptoms to some extent. Since the patient presented with spontaneous onset of the condition with no contributory etiology, he was diagnosed as to have idiopathic bilateral Adie’s tonic pupil.

Discussion

Adie’s syndrome is a disease usually seen in young adults. Most patients with Adie’s
 syndrome have an accommodative paresis in the affected eye at the onset of the condition and this is the chief source of their symptoms.

“Vermiform contractions” was a phrase first used by Sattler in 1911. After Adie had popularized the syndrome which bears his name, it was soon noticed that these patients often showed “vermiform movements” and irregularities of the pupillary margin, and a readily recognizable drawing of an Adie’s iris was reproduced in Thiel’s Atlas. It is believed that “vermiform movements” of the sphincter are actually a state of physiologic pupillary unrest (“hippus”) of those sectors of the sphincter which are still wired up to the light reflex; and that the real abnormality to be seen in the iris sphincter in Adie’s syndrome is the segmental paralysis due to the loss of some of the postganglionic nerve fibers either in the ciliary ganglion or in the short ciliary nerves.

Drouet et al. and Millar et al.,[6,7] in different case reports described bilateral Adie’s pupil in a patient during an attack of migraine. The authors discussed that the transient mydriasis occurred due to a postganglionic dysfunction affecting the iris sphincter. Our patient neither had any history of migraine nor was the mydriasis transient.

Jivraj and Johnson[1] described a case of acute unilateral tonic pupil in a young girl, which after 2 months of presentation became bilateral. The authors identified a rare etiological association of neurosyphilis with the bilateral tonic pupils. Our patient showed simultaneous bilateral involvement and was seronegative.

Ross syndrome,[10] described by Alexander Ross in 1958, is a syndromic presentation of bilateral tonic pupils, segmental anhidrosis, and absent or diminished deep tendon reflexes. Baran et al. and Mayer[2,3] in separate case reports have reported patients with bilateral tonic pupils as a part of the syndromic presentation of Ross syndrome. A rare presentation of bilateral Adie’s pupil as a part of Vogt–Koyanagi–Harada has been reported in literature by Kim et al. and Narang et al.[4,5] However, our patient did not have any significant drug history, neither did he have any other neurological impairments nor features suggestive of anhidrosis. According to literature, mydriasis and light-near dissociation are seen in 50% of patients with Fisher syndrome due to the involvement of the pupillomotor fibers. Nitta et al.,[11] in their study where they analyzed pupillary responses in patients with Fisher syndrome, concluded that the denervated iris sphincter muscles, which show super sensitivity to cholinergic agents, may be responsible for the mydriasis and light-near dissociation seen in patients with Fisher syndrome. Our patient showed sensitivity to the cholinergic transmitter (0.1% solution of pilocarpine) but did not have any other features of Fisher’s syndrome.

Autonomic dysfunction and absent tendon reflexes[9] have been reported in cases with Bilateral Adie’s pupil. Holmes G et al.,[8] described bilateral Holmes Adie pupil in 3 patients who on evaluation were found to have autonomic dysfunction. Our Patient had no evidence of Autonomic dysfunction and had normal deep tendon reflexes.

Bilateral cases of Adie’s tonic pupil are usually reported to occur non-simultaneously, i.e., the second pupil is involved after a few weeks or more commonly, a few months. Our case is unique in that our patient presented with simultaneous bilateral tonic pupils but with minimal visual symptoms. Moreover, the bilaterally reported cases usually occur in females in a younger age group, but our patient was a middle-aged male, which is atypical. The complaint of difficulty with near work could have been passed off as presbyopia without a high index of suspicion and a careful slit-lamp examination. He also did not have any systemic predisposing condition for the same which was evidenced by the negative laboratory work. A thorough workup toward a differential diagnosis helped in proper evaluation and management of our patient.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.
Conflicts of interest
There are no conflicts of interest.

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