Episodic tonic pupil with aneurysm located on the same side

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Tonic pupil (TP) characterized by unilateral or bilateral dilatation of the pupil and lack of light reaction due to postganglionic injury of the parasympathetic oculomotor nerve was firstly described in 1931.[1] While sphincter pupillae denervation produces mydriasis, ciliary muscle denervation causes accommodation impairment. It is often accompanied with peripheral nerve dysfunctions.[2] It is predominantly seen in females between the ages of 20 and 40. Although this condition is of unknown etiology, most of the TP cases are caused by viral infection. Trauma, tumors, vascular lesion with ischemia, temporal arteritis, diabetes mellitus or autonomic neuropathies are the other causes damaging the ciliary ganglion.[3-5] In this paper, we report an episodic TP case with aneurysm located posteriorly.

A healthy 32-year-old female presented to the emergency room of our Faculty with blurred vision and enlargement of the right pupil. She had a history of nausea without vomiting, sharp, severe headache in the temporal region approximately 3 days before pupil enlargement was noticed. There was no deterioration in consciousness. Her neurological examination revealed mydriasis with absent light reaction (direct and consensual) and contracted pupil during the accommodative reflex test in the right pupil. There was no ptosis. Optic disc examination was normal. No motor or sensory deficit was found. Deep tendon reflexes were normal, and the pathological reflex was not found. According to the history and neurological findings, the first considered diagnosis was compressing lesions and urgent neuro-imaging techniques were performed. Cranial and orbital MRI were normal but cranial MR angiography revealed aneurysmal dilatation in the A2 segment of right anterior cerebral artery (ACA) and distally to the right posterior cerebral artery (PCA) with left vertebral artery hypoplasia (**Figure 1**). Neurosurgical assessment was demanded to determine the need for a surgical approach, however, it was accepted that the site of the aneurysms were not found to be responsible for the non-reactive enlargement of the right pupil. After the exclusion of tumor, infection, ischemic disease, trauma, and mechanical compression, 0.1% pilocarpine test was performed in order to differentiate Adie pupil. Pilocarpine eye drops elicited brisk constriction of the right-sided TP approximately 25 minutes after the application. No abnormality of iris or sphincter was found on slit lamp exam. This positive result demonstrated the denervation hypersensitivity and confirmed the diagnosis of Adie pupil. Etiological evaluation of Adie pupil included serologic tests for neurosyphilis, Lyme disease, Sjögren disease, and sarcoidosis. The results of these tests were negative. Moreover, the histories of viral infections, trauma, vaccinations, diabetes mellitus, and giant cell arteritis of the patient were also negative. When the patient was reevaluated 10 days after the first examination, the right pupil was still mydriatic and non-reactive. In addition, information of a 3-day period with normal size was obtained from the patient. Due to this period of normalization in size, the patient was accepted as "episodic TP".

Pupil asymmetry is an important finding and should be investigated to rule out severe underlying neurological disorders such as tumors, aneurysms, and hernia. Exposure to mydriatic substance, Adie's TP, and local disorders such as closed angle glaucoma or segmental spasm of the iris dilator muscle are the other causes, which should be considered in the differential diagnosis.[5] Another cause of the pupil asymmetry is benign episodic unilateral mydriasis. The underlying physiopathology is not always clear and may involve either parasympathetic deficiency or sympathetic hyperactivity affecting the iris.[5] Patients with episodes are usually associated with benign neurological conditions, for example, migraine, and do not require further neuro-diagnostic studies.[6]

Tonic pupil is the most common problem among those mentioned above in unilateral dilatation of pupil. Mostly, the condition is idiopathic and you cannot find an acceptable etiology. The diagnosis is primarily made by slit lamp study and segmental pupillary sphincter palsies are very typical.[5] Absent or severely reduced light reaction and better accommodation reaction can be demonstrated due to regrowth of the initially damaged autonomic fibers. The pupil constricts with 0.1% pilocarpine (cholinergic hypersensitivity), but this concentration is ineffective in normal pupils.[5] Usually, no further investigation is necessary in TP cases. There was a light-near dissociation meaning a postganglionic parasympathetic lesion (TP) in our patient. She presented with intermittent episodes of pupil asymmetry with no other neurological symptoms. She did not

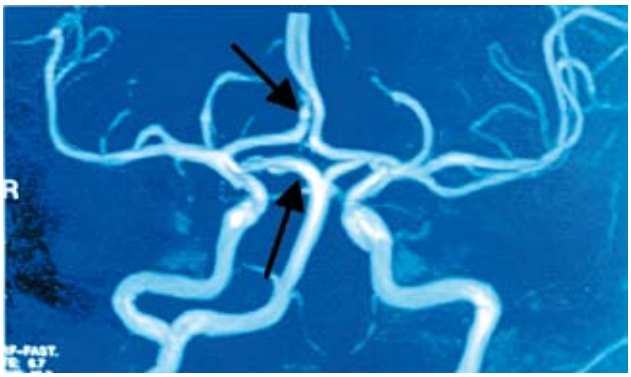

Figure 1 - Cranial MRI revealed aneurysmal dilatation in the A2 segment of right anterior cerebral artery and distally to the right posterior cerebral artery with left vertebral artery hypoplasia.

complain of any headache episodes more previously. Although no cause can be identified in anatomical locations, aneurysmal dilatation was accepted as a related condition in the patient. The question is still to be answered whether this co-existence was co-incidental or not.

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