

Ocular neuromyotonia after radiotherapy for nasopharyngeal carcinoma: the first reported case in Hong Kong

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Abstract

Double vision is a common symptom. Usually, the cause of diplopia is due to cranial nerve palsies, such as third, fourth, or sixth nerves palsies. Sometimes, the cause may be due to a rare neuromuscular disorder. This report is of a Chinese woman with a history of radiation therapy for nasopharyngeal carcinoma who subsequently developed double vision and was found to have intermittent left-sided lateral rectus muscle spasm. The patient underwent complete neuro-ophthalmic assessment, including investigation of the eyelid position, ocular motility, force duction test, alignment, appropriate blood tests, and imaging. Her condition was finally diagnosed as ocular neuromyotonia secondary to radiation therapy. To the authors' knowledge, this is the first patient from Hong Kong to be reported.

Key words: Oculomotor nerve diseases, Nasopharyngeal neoplasms, Radiotherapy

Introduction

Ocular neuromyotonia is a rare neuromuscular disorder characterized by paroxysmal involuntary contraction of 1 or more ocular motor muscles resulting in intermittent diplopia and strabismus.¹ The underlying pathology is spontaneous firing of the unstable ocular motor nerve membranes, which causes muscle spasms. Radiotherapy is a common cause.

Membrane-stabilizing agents such as carbamazepine and phenytoin are effective for treating this disease.

Case report

A 70-year-old Chinese woman presented with a 3-month history of episodic diplopia. She reported episodes of horizontal binocular double vision that occurred spontaneously several times a day. The episodes usually lasted for 30 to 60 seconds and were worst on right gaze. She was asymptomatic between episodes. She had a history of nasopharyngeal carcinoma (NPC) treated with radiotherapy 20 years previously. She had been followed up by the oncologists and had had no recurrence of NPC.

Complete ophthalmic assessment was performed. Her visual acuity was 6/12 in both eyes. No ptosis was noted at external examination, and no fatigability or Cogan's lid twitch was demonstrated. Her pupils were equal and reactive to light. Ocular motility was full, except for a slight limitation on abduction of the left eye. Slit-lamp examination showed a quiet anterior chamber, normal intraocular pressure, and immature cataract in both eyes. Fundus examination with indirect ophthalmoscopy showed normal optic discs with sharp disc margins, flat retinas, and normal vasculature. Systemic examination, including cranial nerve and neurological examination, was unremarkable. Orthoptic assessment showed small esophoria of 2 PD (**Figure 1**), and limited left eye abduction with slight lateral rectus underaction was noted by Hess chart.

Diplopic paroxysms occurred after sustained left gaze. During these episodes, the left eye gradually deviated outwards



Figure 1. Force duction test showed no eye deviation during the quiescent phase.



Figure 2. Force duction test showed that the left eye was deviated outwards during the active phase.

and resulted in exotropia of 40 PD (**Figure 2**). Eventually, spasm of the left lateral rectus occurred as the globe was sustained in the abducted position with loss of motility in all other directions. The attacks usually lasted for 30 to 60 seconds and returned to normal gradually.

Force duction test was performed during both the quiescent and active phases. The test was negative during the quiescent phase but positive during the active phase. Laboratory investigations of thyroid function test and anti-acetylcholine receptor antibody were normal. Computed tomography scan of the brain and orbits showed no recurrence or metastasis of the malignancy. There were no features suggestive of thyroid ophthalmopathy. Clinically, the most likely diagnosis was ocular neuromyotonia.

The patient was treated with carbamazepine 200 mg twice per day. She reported improvement, with reduction in the frequency of the episodes. However she had poor tolerance to the drug and it was stopped. After cessation of the drug, her symptoms recurred, with increased frequency of the episodes.

Discussion

Ocular neuromyotonia is a rare paroxysmal involuntary contraction of one or more ocular motor muscles, resulting in intermittent diplopia and strabismus.¹⁻³ The condition was first described in 1970.⁴ To the authors' knowledge, the condition has not been reported in Hong Kong to date. The most common cause is radiotherapy to the sellar or parasellar region.² However, ocular neuromyotonia could also occur in association with a chronic nerve palsy,⁵ result from compressive lesions such as Graves' disease, vascular compression or arachnoiditis, or stroke,³ or be idiopathic in origin.²

Ocular neuromyotonia is characterized by brief episodic contractions of the ocular muscles. The condition may occur spontaneously, triggered by eye movements and/or sustained gaze in a particular direction.¹ Patients usually have normal extraocular movements and ocular alignment between episodes. Electromyographic study has shown that it is a neurogenic rather than a myogenic disorder. Spontaneous firing of the unstable ocular motor nerve membranes has been shown to cause the muscle spasms.³ It is suggested that radiation therapy or ocular motor nerve compression causes segmental demyelination and results in abnormal excitability of the peripheral motor axons.² This abnormal motor nerve activity belongs to the syndrome of continuous motor unit activity, which is the hallmark of ocular neuromyotonia. However, there is no apparent relationship between radiation therapy and time of onset. Episodes can occur from 2 months to 18 years after radiotherapy.⁶ As the condition is due to abnormal excitability of the neuron, membrane-stabilizing agents such as carbamazepine or phenytoin are used to control the abnormal muscle spasms.^{2,3}

This report is of the first patient with ocular neuromyotonia in Hong Kong. This was a rare cause of intermittent diplopia. The patient developed the disease 20 years after radiotherapy for NPC. Her symptoms were relieved by the membrane-stabilizing agent carbamazepine, although the patient did not tolerate the drug well.

References

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