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Heavy eye syndrome – case report

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Abstract

This paper aims to report clinical features, differential diagnosis and successful surgical outcome of a patient with myopic strabismus fixus, also known as Heavy Eye syndrome (HES). We present a case of a 47-year-old man who presented to the Ophthalmology Department with diplopia and poor vision. In the past, he had been diagnosed with Graves' disease, high myopia, and secondary open-angle glaucoma. He had undergone orbital decompression and cataract surgery of both eyes. On examination, the patient had esotropia and hypotropia with limited abduction and elevation. The patient was ultimately diagnosed with HES. Yokoyama's surgery combined with a medial rectus muscle recession in the right eye, were performed. The procedure reduced esotropia, hypotropia and improved ductions.

Running title: Heavy eye syndrome

Keywords: heavy eye syndrome, high myopia, myopic strabismus fixus

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Introduction

Heavy Eye syndrome is a rare type of strabismus resulting from high axial myopia. HES is characterized by increased axial length, large-angle esotropia, and hypotropia. Eye movements are restricted especially abduction and elevation [1,2]. We present a case of a 47-year-old male with high myopia, esotropia and hyoptropia who has been diagnosed with Heavy Eye syndrome (HES).

Case report

A 47 – year old man presented to the Ophthalmology Department with horizontal diplopia and poor vision. In the past, he had been diagnosed with Graves' disease, high myopia (-34.00 diopters), and secondary open-angle glaucoma. He had undergone orbital decompression and cataract surgery of both eyes. He also had a YAG capsulotomy of the left eye. After cataract surgery, he developed convergent strabismus. Examination revealed esotropia of +30 PD (prism diopters), hypotropia, limited abduction, and elevation of both eyes, more pronounced in the right eye (**Fig. 1**). Binocular vision was impaired. Slit-lamp examination revealed chorioretinal atrophy, foveal scarring and optic disc pallor in both eyes. The intraocular pressure was normal. Images obtained from ocular ultrasound displayed posterior staphyloma present in each eye. MRI revealed in both eyes downward displacement of lateral rectus muscle, nasal displacement of superior rectus muscle (**Fig. 2A,B**) and pseudopropto-



FIGURE 1 Photographs before the surgical treatment, showing limitations of abduction and elevation of both eyes, especially of the right eye

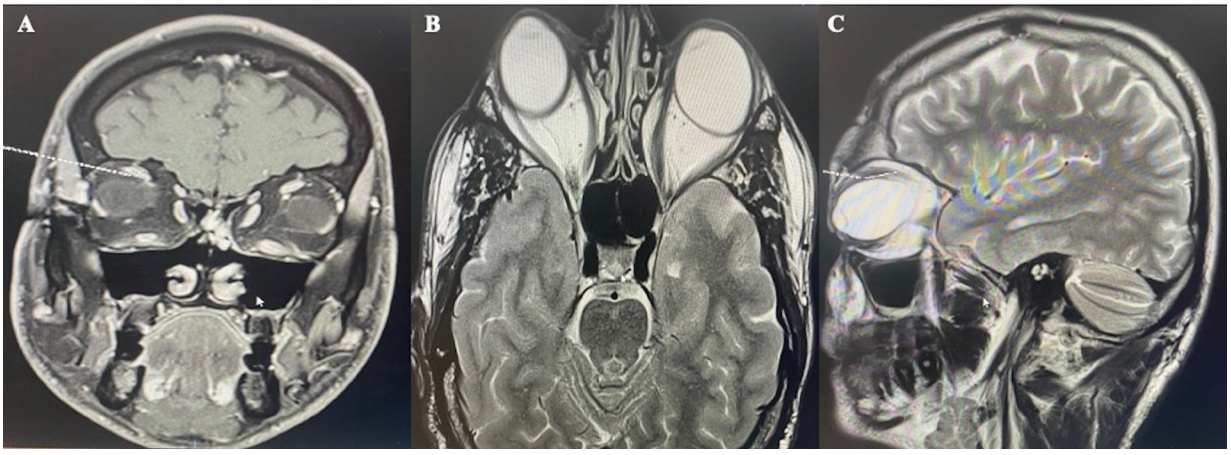


FIGURE 2 MRI scan of the patient showing: displacement of lateral rectus muscle downward and superior rectus muscle medially in both eyes (A), elongated and dislocated eyeballs (B), pseudoproptosis resulting from elongated eyeballs (C)

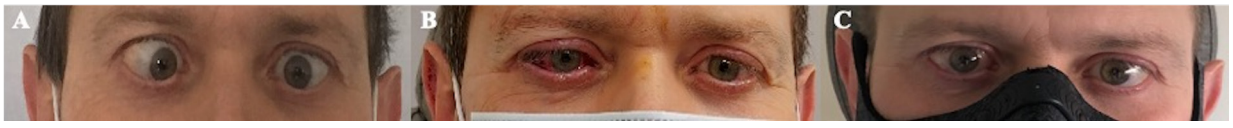


FIGURE 3 Photographs showing patient before (A), 24 hours after (B) and 5 weeks after (C) Yokoyama's surgery and medial rectus recession in the right eye



FIGURE 4 Photographs, five weeks after surgery, showing reduced esotropia, hypotropia and improvement of abduction of the right eye

sis (**Fig. 2C**). The axial length of the right eye was 31 mm and of the left one 33 mm. The patient was diagnosed with Heavy Eye syndrome (HES). Yokoyama's surgery (union between total muscle bellies of superior rectus and lateral rectus muscles) and the medial rectus muscle recession of 6 mm in the right eye were performed. The procedure reduced hypotropia and esotropia to +10 PD (**Fig. 3).** Ductions improved, (**Fig. 4**). The patient did not consent to the operation of the left eye, as he was fully satisfied with the outcome. His self-esteem and comfort of life improved significantly.

Discussion

Heavy eye syndrome develops in eyes with high axial myopia. The patients usually present with esotropia and hypotropia. Additionally, it can be complicated by pseudoproptosis, limited abduction and elevation [1,2]. Yamaguchi and coworkers proposed the theory that explains the etiology of HES. According to them, the posterior part of the elongated eyeball moves, superotemporally, out of the muscle cone. Dislocated part of the globe "squeezes'' between the superior rectus (SR) and lateral rectus (LR) muscles. This action displaces the SR nasally and LR inferiorly [2,3]. MRI is essential to identify this abnormality [1,4]. The effective treatment of HES is surgery. The surgical options are: Yokoyama's (union between total muscle bellies of SR and LR muscles with a non-absorbable suture around 14 mm posteriorly from the muscle insertion), Yamadas's (hemitransposition of the SR and LR together with large MR recession), and partial Jensen's surgery (SR and LR are split in a half and only halves of the muscles are joined). The surgery may be expanded by the recession of the medial rectus muscle (MR). In case of our patient, it was decided to carry out Yokoyama's surgery combined with a 6 mm recession of MR muscle.

Heavy eye syndrome should always be distinguished from other etiologies of acquired strabismus. The differential diagnosis includes; sagging eye syndrome, Graves orbitopathy, ocular myasthenia gravis, cranial nerve palsies, and acquired distance esotropia associated with myopia [1]. Sagging Eye syndrome (SES) is found in non-myopic elderly patients. It is caused by degeneration of connective tissues and can coexist with bilateral blepharoptosis [5]. Patients with SES present esotropia, hypotropia, limited elevation and contrary to HES - normal abduction [1,5]. MRI is crucial to differ heavy eye syndrome from sagging eye syndrome (SES). In HES globes are elongated, usually with staphylomas, while in SES typically globes appear normal. MRI in SES shows inferior dislocation of the lateral rectus muscle and LR-SR band degeneration. The mean angle between the LR and SR muscles is significantly smaller in SES than in HES [1]. In both, HES and SES inferior rectus muscle is dislocated. However, the outcome of the study conducted by Kinori and coworkers revealed that in HES the inferior rectus muscle is more nasally displaced from midline comparing to patients with SES [2,6]. Another differential diagnosis is Graves' orbitopathy, in which patients present upper eyelid retraction, edema and congestion of periorbital tissues and conjunctiva [7]. Extraocular muscles and adipose tissue are swollen [8]. To distinguish it from HES, it is important to perform MRI and assess the level of thyroid hormones. HES is more probable, when MRI images present elongated globes and endocrine tests indicate euthyreosis [2,9]. Our patient has been diagnosed with Grave's disease in the past. He had undergone bilateral orbital decompression (in 2016 and 2017 – we have no MRI images from that period). Patient has been in the euthyreosis since 2018, at a later time he developed convergent strabismus. Diagnosis of

HES was made in our department in 2020. Ocular myasthenia gravis can manifest as any comitant or incomitant strabismus [10]. The disease usually manifests with diplopia together with ptosis and saccadic fatigue [11]. Myasthenia gravis can be diagnosed by serological findings (presence of antibodies to some synaptic molecules; AChR- Ab, MuSK-Ab, LRP4-Ab), electrodiagnostic testing (repetitive nerve stimulation, single-fiber electromyography) or pharmacological tests (neostigmine, edrophonium) [12]. There is no single test to identify patients with nerve palsies. It is important to collect information on current symptoms and past medical history. Patients usually report binocular diplopia and present ocular deviations. Isolated third nerve palsy is characterised by ptosis, impaired adduction, elevation and depression of the eye and sometimes dilated, poorly responsive to light, pupil. The disorder is idiopathic or caused by head trauma, aneurysm (at the junction of the internal carotid and), vascular disease due to systemic hypertension or diabetes mellitus [13,14]. Fourth cranial nerve palsy affects function of the superior oblique muscle. The patient manifests hypertropia (higher eye is on the affected side). It can be congenital or caused by trauma or ischemia. Aneurysm is never a source [14]. Isolated sixth cranial nerve palsy typically results from ischemia (as a consequence of cardiovascular diseases, hypertension, hyperlipidemia, diabetes etc.) [15]. Another differential diagnosis is an acquired distance esotropia associated with myopia in young adults. The etiology has not yet been discovered. However, it has been observed that lateral rectus muscle weakens from prolonged near work which leads to slow, progressive esotropia. Contrary to heavy eye syndrome, in acquired distance esotropia eye movements are normal and myopia is not so severe (In the reported cases, of patients with acquired distance esotropia, the mean spherical equivalent was 5.50) [2,16].

Conclusions

Heavy eye syndrome is a rare type of acquired strabismus, nevertheless it should always be considered when diagnosing patients with high myopia, esotropia, and exophthalmos. The orbital imaging is crucial for diagnosis and for planning treatment, especially in patients with complex medical history.

Ethical approval

The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration. As all of the data presented were collected during routine clinical practice without any additional clinical procedures or inclusion of patient sensitive data, no Ethical Committee approval is needed for this study.

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Not applicable.

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Conflict of interest

The authors declare they have no conflict of interest.

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