

# Clinical Course of a Pediatric Graves' Extraocular Myopathy Patient Followed-up by Magnetic Resonance Imaging

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## Abstract

A 14-year-old male patient with Graves' orbitopathy presented with a downward gaze restriction in the left eye. Magnetic resonance imaging (MRI) revealed an edematous left superior rectus muscle. Retrobulbar injection of triamcinolone acetonide (20 mg) was administered in the left orbit. However, edema was still evident in the left superior rectus muscle on MRI, 3 months after the injection, and new inflammation was detected in bilateral inferior rectus muscles. The patient then underwent three cycles of steroid pulse therapy (1 cycle: methylprednisolone 10 mg/kg/day × 3 days). One week after the steroid pulse therapy, eye movement was improved and the inflammation in the left superior rectus muscle and the bilateral inferior rectus muscles subsided on MRI. However, the patient noticed diplopia during upward gaze 2 months later, and MRI showed recurrence of edematous changes in bilateral inferior rectus muscles. The patient was treated with the same protocol of steroid pulse therapy. One month after the second steroid pulse therapy, ocular motility was improved and the inflammation in both inferior rectus muscles had almost resolved. This case illustrates the detailed clinical course of edematous extraocular myopathy in a pediatric Graves' orbitopathy patient, followed-up by successive MRI.

**Keywords:** Extraocular myopathy; Pediatric Graves' orbitopathy; Edematous change; Magnetic resonance imaging; Interventional clinical course

## Introduction

Extraocular myopathy in pediatric Graves' orbitopathy (GO) is a rare manifestation [1], and its clinical course remains unclear. Only one study using magnetic resonance imaging (MRI) [2] has reported on the clinical course of this entity. However, this previous study only assessed the disease courses by examination of the extraocular muscle diameters, but edematous change in the muscles was not recorded. However, muscular enlargement is also detectable during stable disease, and does not always reflect inflammation of the extraocular muscles [3].

In the present study, we report an interventional clinical course of edematous extraocular myopathy in a 14-year-old GO patient, followed-up by successive MRI.

## Case Report

A 14-year-old male patient had noticed proptosis in both eyes 1 month before referral to our clinic. He had a 1-month history of hyperthyroidism with positive thyroid-stimulating hormone receptor antibody (5.5 IU/L; normal range: <1.0 IU/L), and was receiving thiamazole (15 mg/day).

On initial examination, his visual acuity and intraocular pressure were normal. Hess chart examination showed mild restriction of downward gaze in the left eye (Figure 1A). His binocular single vision field [4] was defective in the upper-right, left, and inferior areas (Figure 1B). Exophthalmometry showed 18 mm OD and 17 mm OS (base: 104 mm). He had periorbital inflammatory signs in both sides, including eyelid swelling, eyelid redness, conjunctival chemosis, conjunctival injection, and orbital discomfort (clinical activity score [CAS] [5]: 5 points). MRI revealed an edematous superior rectus muscle in the left orbit (Figure 2A and Figure 2B).

Retrobulbar injection of triamcinolone acetonide (20 mg) was administered into the left orbit 1 week after the initial examination. One month after the injection, the CAS in the left side improved to 2

points. However, 3 months after the injection, upward gaze limitation developed in the right eye (Figure 1C and Figure 1D), although the CAS in the right side was 2 points. The CAS in the left side was also 2 points. MRI revealed that the left superior rectus muscle was still inflamed and new edematous changes were detected in the bilateral inferior rectus muscles (Figure 2C and Figure 2D). The patient was treated with three cycles of steroid pulse therapy (1 cycle: methylprednisolone 10 mg/kg/day × 3 days). One week after the therapy, the bilateral CAS decreased to 0 points. Eye movement was improved (Figure 1E and Figure 1F) and the inflammatory changes in the left superior rectus muscle and the bilateral inferior rectus muscles had almost subsided on MRI (Figure 2E and Figure 2F).

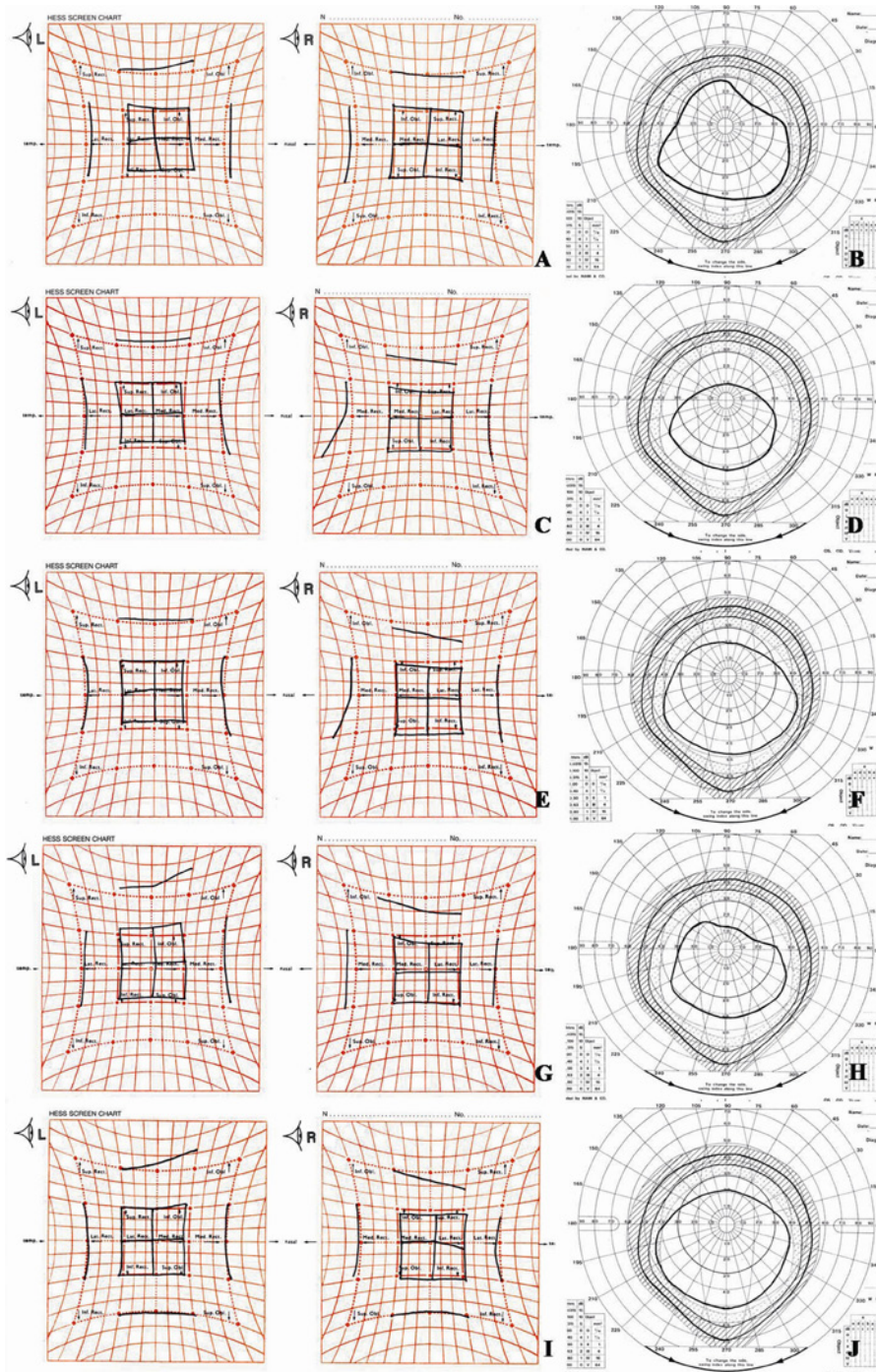
However, 2 months later, the patient noticed diplopia during upward gaze. At this time, the patient was maintained in a euthyroid state by thiamazole administration (5 mg every other day). The CAS had increased to 3 points bilaterally. Ocular motility examination revealed an upward gaze restriction in the right eye (Figure 1G and Figure 1H). MRI showed bilateral edematous inferior rectus muscles (Figure 2G and Figure 2H). The patient was treated again with the same protocol of steroid pulse therapy. One month after the second steroid pulse therapy, the CAS had decreased to 0 points in both sides. Ocular motility improved (Figure 1I and Figure 1J) and the inflammation in the bilateral inferior rectus muscles had almost resolved on MRI (Figure 2I and Figure 2J).

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**Figure 1.** A series of Hess chart and binocular single-vision field results. The diagonal line area in the Goldmann chart represents the normal limit of Japanese binocular single vision field. Downward gaze at 30 degrees is not detected on the Hess chart, except in Figure 1I.

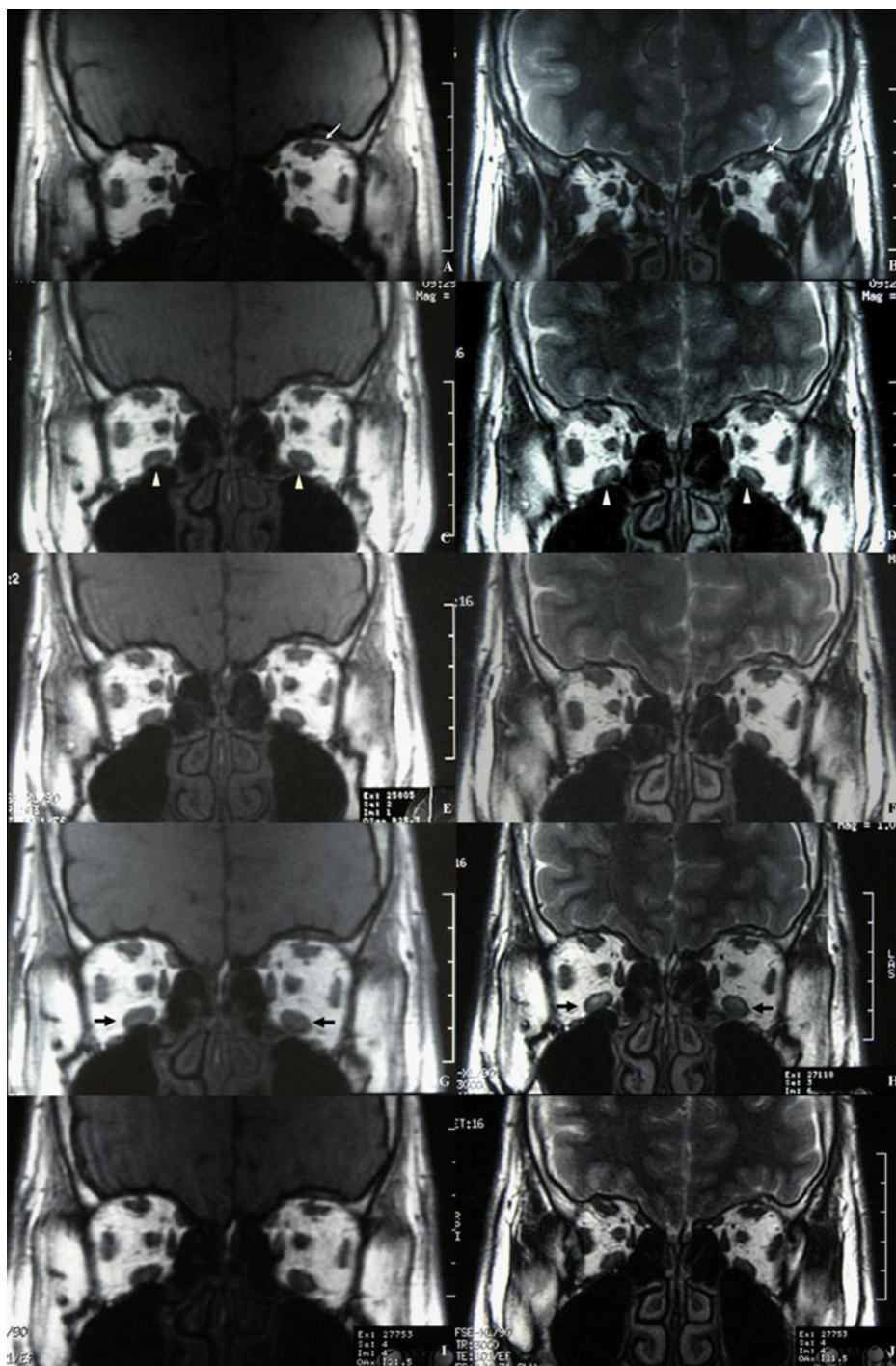
**A, B.** Examination at first visit. The Hess chart shows mild restriction of downward gaze in the left eye. The upper-right, left, and inferior areas of the binocular single-vision field are defective.

**C, D.** Examination 3 months after triamcinolone acetonide injection. The Hess chart shows a new restriction of upward gaze in the right eye. The binocular single-vision field has narrowed in the upper area.

**E, F.** Examination 1 week after the initial steroid pulse therapy. Although the Hess chart shows little change, the binocular single-vision field has widened in the upper area.

**G, H.** Examination 2 months after initial steroid pulse therapy. Upward gaze in the right eye is restricted in the Hess chart, which corresponds to the upper-area defect of the binocular single-vision field.

**I, J.** Examination 1 month after second steroid pulse therapy. Upward gaze restriction in the right eye has improved in the Hess chart, and the binocular single-vision field has widened.



**Figure 2.** Coronal sections of magnetic resonance imaging.

**A, B.** Examination at first visit. **A.** T1-weighted imaging. **B.** T2-weighted imaging. The left superior rectus muscle is swollen with low intensity area on T1-weighted imaging and high intensity area on T2-weighted imaging (white arrows).

**C, D.** Examination 3 months after triamcinolone acetate injection. **C.** T1-weighted imaging. **D.** T2-weighted imaging. The inferior rectus muscles on both sides are enlarged with low intensity areas on T1-weighted imaging and high intensity areas on T2-weighted imaging (white arrow heads).

**E, F.** Examination 1 week after initial steroid pulse therapy. **E.** T1-weighted imaging. **F.** T2-weighted imaging. The high intensity areas in the left superior rectus muscle and the bilateral inferior rectus muscles on T2-weighted imaging have almost disappeared.

**G, H.** Examination 2 months after initial steroid pulse therapy. **G.** T1-weighted imaging. **H.** T2-weighted imaging. Low intensity areas on T1-weighted imaging and high intensity areas on T2-weighted imaging are seen in the inferior rectus muscles on both sides (black arrows).

**I, J.** Examination 1 month after second steroid pulse therapy. **I.** T1-weighted imaging. **J.** T2-weighted imaging. The high intensity areas in the bilateral inferior rectus muscles on T2-weighted imaging have almost disappeared.

## Comment

The present report illustrates a detailed interventional clinical course of edematous extraocular myopathy in a pediatric GO patient, monitored by successive MRI.

Although the CAS improved after triamcinolone acetonide injection in the left side, MRI demonstrated inflammation in the left superior rectus muscle and new edema development in the bilateral inferior rectus muscles. Although the CAS generally correlates with the MRI findings of edematous changes in the extraocular muscles [3], the present case suggested a possible discrepancy between the CAS and the MRI findings.

Ocular involvement in pediatric GO is generally mild and commonly improves during a euthyroid state even if steroid therapy is not performed [1,6]. Thus stabilizing the patient's thyroid function is a significant therapeutic concern in pediatric GO [6]. However, our patient developed recurrent inferior rectus myopathy even under a euthyroid state, following initial steroid pulse therapy. This case suggests that pediatric GO patients with extraocular myopathy, i.e., with severe ocular symptoms, have a potential for disease progression, irrespective of normal thyroid function. Ophthalmologists and endocrinologists need to carefully monitor the ocular motility changes in such patients.

In conclusion, the present case illustrates an interventional clinical course of extraocular myopathy in a pediatric GO patient, followed-up by successive MRI. This case indicates that discrepancies between the CAS and the MRI findings may occur, and that pediatric GO patients with severe ocular symptoms have the potential for disease progression, irrespective of normal thyroid function.

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