A CASE OF SEVERE GRAVES’ OPHTHALMOPATHY

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Introduction

Graves’ ophthalmopathy typically presents with symptoms of proptosis and diplopia. It is an autoimmune condition of retro-orbital tissues. It is estimated that at least 50% of patients with Graves’ disease develop clinically evident ophthalmopathy. Thyroid eye disease (TED) may occur before, during or after resolution of the hyperthyroidism. It remains one of the most difficult diseases to treat in endocrinology. We present a case in which the management of the ophthalmopathy has been complex and required escalation to immunosuppression and consideration of biological agents.

Presentation

A 34-year-old female graphic designer presented to eye casualty with 2 weeks of diplopia. She had normal visual acuity with no past medical history or family history. She was not on any regular medication and never smoked. MRI showed bilateral extraocular muscle enlargement. Thyroid eye disease was diagnosed. She was commenced on selenium and referred to the thyroid eye clinic. Biochemistry revealed her to be thyrotoxic with values of: TSH < 0.01 U/mL, FT4 > 65 ng/dL, FT3 45 ng/dL. TSH receptor antibodies were positive at 2.9 unit/ml (NR < 0.4). She was very symptomatic and commenced on Carbimazole 60mg daily with a view to be treated with a ‘block and replace’ regimen and beta blockers.

Management of TED

Thyrotoxicosis

The patient responded very quickly to high dose Carbimazole therapy and Levothyroxine was added in a timely fashion. She was soon established on 40 mg Carbimazole and 75 mcg Levothyroxine with a detectable TSH and normal free hormone levels. TSH receptor antibody titres decreased to 1.2 unit/ml within a few months of treatment.

Ophthalmopathy

The patient was commenced on intravenous pulsed methylprednisolone (6 weeks of 500 mg weekly, followed by 6 weeks of 250 mg weekly). 10 weeks into her pulsed methylprednisolone course, and despite being biochemically euthyroid, the patient developed worsening visual acuity and colour vision. She underwent emergency bilateral orbital decompression. Post-operatively, she was commenced on oral prednisolone 50 mg daily and mycophenolate, which was uptitrated to 1.5 g twice daily after regular reviews in the thyroid eye clinic.

The patient has recovered her colour vision but she has restrictive strabismus in the left eye with visual acuity of 6/18 pinhole (6/9 unaided) and visual acuity in the right eye of 6/18 pinhole (6/12 unaided). The patient is being slowly weaned down on prednisolone, but not beyond 15mg daily as attempts at further reducing doses have led to worsening diplopia.

The patient continues to be reviewed in a multi-disciplinary thyroid eye clinic with an endocrinologist and a thyroid eye specialist and having very close monitoring of her vision. Given her clinical euthyroidism and the relative decrease in thyroid receptor antibody titres, the team has decided against thyroidectomy. Her eye limitation improved initially with anti-inflammatory treatment. However, her repeat magnetic resonance imaging showed bilateral extensive swelling of the muscles, which does not entirely explain the unilateral severe right restrictive strabismus causing intractable diplopia. She has temporarily increased her steroids again and RituXimab through two courses is planned in the immediate future. Steroid treatment needs to be weaned down, as there is now a very existing risk of adrenal suppression that needs to be considered.

Discussion

The management of Graves’ ophthalmopathy is complex. Sometimes it is difficult to predict the course of Graves’s ophthalmopathy from that of thyrotoxicosis, despite biochemical euthyroidism always being a target of treatment at the cornerstone of managing TED. Many treatments have side effects (hepatoxicity, adrenal suppression). In severe cases of ophthalmopathy, aggressive treatment is required for sight-saving measures. This case highlights the importance of the multi-disciplinary approach in managing severe cases to ensure early diagnosis and treatment escalation with immune suppression and/or biological agents.

References