Therapeutic challenge of a paediatric case of Graves' disease with severe ophthalmopathy

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DESCRIPTION

An otherwise healthy girl aged 4 years was observed because of a 6-month history of tachycardia, polyphagia, chronic cough and agitation. Physical examination revealed hiperhydrosis, exuberant exophthalmia (figure 1) and a palpable, elastic and painless goitre. Laboratory work up showed an increased FT4, low thyroid-stimulating hormone (TSH) and positive thyrotropin receptor antibodies (TRABs) (figure 2), and thyroid ultrasonography demonstrated a diffusely heterogeneous, enlarged gland with no nodules, compatible with Graves' disease. The girl was initiated immediately to treatment with tiamazole (MTZ) and propranolol, and after few months, an euthyroid state was achieved and maintained until 2 years (figure 2). After this period, MTZ treatment was suspended and 1 month later, she returned with restless sleep and daytime agitation. Laboratory work up showed a TSH<0.005 µU/mL, T4L of 0.98 ng/mL and a positive TRABs and MTZ treatment (figure 2) was reintroduced.

At 7 years, she presented with worsening exophthalmia, eyelid retraction, lagophthalmos (figure 1)

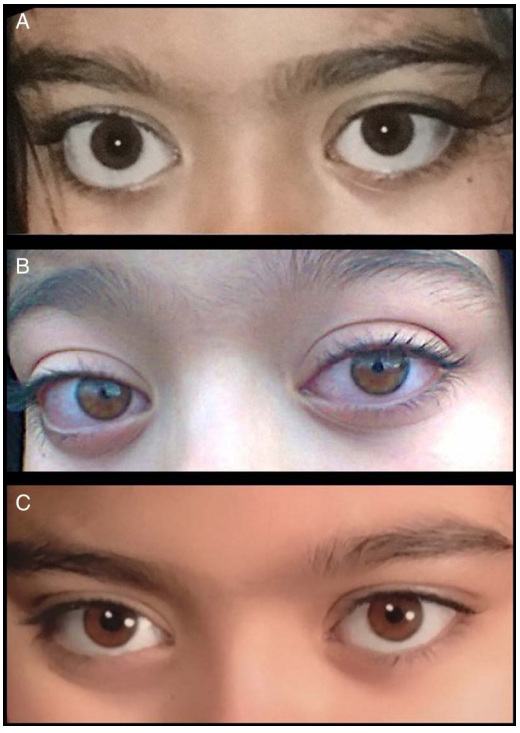

Figure 1 Exophthalmia observed on physical examination at diagnosis time (A). Exophthalmia, eyelid retraction and lagoftalmos at 7 years (B). Actually: no exophthalmia (C).

Learning points

- A characteristic feature of Graves' disease is ophthalmopathy. In fact, its presence should raise immediate suspicion of Graves' disease. Its cause remains uncertain, but a shared antigen in the orbit and thyroid gland, such as the thyrotropin receptor, seems to be a probable cause.
- Ophthalmopathy treatment depends on the phase and severity of the disease. The majority of patients only require conservative measures, although sometimes corticotherapy is needed.
- This case demonstrates a therapeutic challenge of a paediatric case of Graves' disease, where the difficulties of assisting remission with medical treatment, prompted to a definitive treatment. In this case, surgery was preferred because of the large thyroid tissue volume and iodine^{131} risks of exacerbate ophthalmopathy.

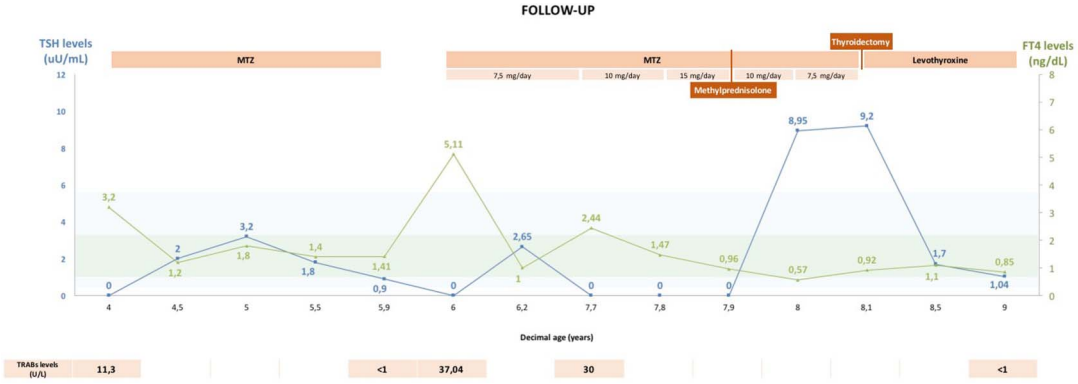

Figure 2 Longitudinal evaluation of thyroid function (FT4 and thyroid-stimulating hormone), thyrotropin receptor antibodies levels and treatment performed from the diagnosis moment to present time.

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and a bigger goitre (left lobe 17×50×19 mm, right lobe 20×55×22 mm), with worsening of hyperthyroidism and although MTZ dosage was adjusted (figure 2), the girl developed asymptomatic bilateral papilloedema. Orbit CT scan showed compressive optic neuropathy and she was started immediately on intravenous methylprednisolone with favourable clinical response. Two months later, she was submitted to a total thyroidectomy and was started on levothyroxine. One year later, she remains well, without exophthalmos (figure 1).

Graves' disease is a rare, but a leading cause of paediatric hyperthyroidism.[1] A typical finding is Graves' ophthalmopathy, often mild and self-limited, but in 3–5% of cases is associated with risk of vision loss.[2 3] The first-line treatment is synthetic antithyroid but when remission is not achieved, definitive treatment should be considered. The choice between surgery and iodine^{131} is individualised, considering the patient and the disease characteristics.[1]

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Patient consent Obtained.

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