Pediatric Systemic Lupus Erythematosus (SLE) Associated Orbitopathy

Samira Aggoune

Departement of pediatric’s, University Teaching Hospital, Belfort Algier’s, Algeria.

Graves’ ophthalmopathy is an organ specific autoimmune process strongly linked to Graves’ hyperthyroidism. Most children with Graves’ ophthalmopathy, have a family history of some type of autoimmune thyroid disease. and some have other autoimmune endocrine diseases, such as diabetes mellitus and Addison’s disease. Systemic lupus erythematosus, rheumatoid arthritis, myasthenia gravis, vitiligo, idiopathic thrombocytopenic purpura and pernicious anemia also have been described in these children.

We report this exceptional observation of an adolescent girl followed for systemic lupus erythematosus associated with orbitopathy.
INTRODUCTION:
Systemic lupus erythematosus (SLE) is an autoimmune disease involving chronic inflammation in numerous organs and tissues, including the thyroid gland. [1,2]

A number of studies have suggested that thyroid disease is more common in SLE than in the general population and the development of thyroiditis can occur before, during or after the diagnosis of SLE. [3,4]

Exophthalmos is subtle and often overlooked in childhood Graves’ disease, and severe proptosis is rare as is orbital disease that is sufficiently severe to compromise vision. There are no pediatric cases of optic neuropathy reported in the literature, and there are only a handful of mild strabismus cases. In addition to the hyperthyroidism, up to 40% of patients with Graves’ disease develop a manifestation localizing to the orbit, called thyroid associated orbitopathy (TAO) or Graves’ orbitopathy (GO). [5]

The most significant pathological findings in TAO include glycosaminoglycan (GAG) deposition (accompanied by swelling resulting from the hydrophilic capacity of these macromolecules), fibrosis affecting the extraocular muscles, and adipogenesis in the orbit. [6,7]

We report teenage girl with SLE who had developed a progressive bilateral exophthalmos. Her presentation was complex and posed a diagnostic challenge. This orbitopathy was subsequently linked to a re-entering thyroid involvement.

CASE
11-year-old girl followed since the age of 6, for arthralgia attributed to acute rheumatoid arthritis treated with extencillin. It was only since the onset of a hemorrhagic syndrome with thrombocytopenia, labeled as severe chronic idiopathic thrombocytopenic purpura, that the patient was referred to us for treatment. This patient came from a non-consanguineous marriage, clinical examination showed good clinical conditions and bilateral exophthalmos, greater on the left eye [Fig1]. The heart rate was 88 beats per minute with a Buchanan at 03.

In view of the serious and refractory nature of the bleeding and thrombopenia, we decided to put the child on rituximab and we took for a thyroid check-up with dosage of Thyroid Peroxidase anti bodies. The dosages were as follows FT3 5.44 pg/ml [ 2.5-3.5] TSH 0.33[0.67-4.75], anti TPO 45 UI/ml (<35). We also found that antiphospholipid (aPL) were present. The ophthalmologic examination found nothing in particular apart from this exophthalmos and we therefore decided to put our patient on synthetic anti-thyroid drugs.

At the last clinical follow-up, after eight months, thyroid function remained good, the exophthalmos was reduced.

Discussion
Thrombocytopenia occurs in about two thirds of children with infantile SLE and in 25 to 30% of pediatric SLE patients with disease onset later in life. [8].

Low platelet counts are associated with the presence of antiphospholipid (aPL) and anti-platelet autoantibodies. Prior to developing pediatric SLE, children and adults with thrombocytopenia may have carried a diagnosis of idiopathic thrombocytopenic purpura (ITP). As was the case for our sick carrier also of (aPL). [9]

In a single retrospective study of 365 Turkish children with ITP, Antinuclear Antibody (ANA) titers of 1:80 or higher were present in 9% of the children but none developed pediatric SLE during the mean followup of 3.6 years. [10]

The SLE patients with overlap syndrome had even higher risks for hypothyroidism and auto immune Thyroid disease[AITD], but not hyperthyroidism in comparison to those SLE patients without overlap syndrome. After diagnosis of SLE, thyroid disease can happen anytime during follow up. The range of interval between the diagnosis of SLE and thyroid disease was 0-10.5 years, with the mean of 3.3-3.7 years. It was about 3 years in our patient.

The incidences of thyroid diseases in SLE patients were compatible to previously reported clinical studies. [11-12]
Clustering of multiple autoimmune diseases has been reported by some studies. [13-14] Increased frequency of thyroid autoimmunity was reported among SLE, RA, SSc, SS, and mixed connective tissue disease (MCTD) patients, especially MCTD and SS. [15,16] There have been a few series of pediatric cases of Graves’ disease in the literature describing the eye manifestations. [17] Graves’ disease in children, however, has been reported to be rare and the calculated incidence was only 0.79 per 100 000 in Danish children. [18] On the other hand, a recent study in Hong Kong Chinese has documented an incidence of 6.5 per 100 000 children. [19] Graves’ ophthalmopathy is an organ specific autoimmune process strongly linked to Graves’ hyperthyroidism. [20] Childhood onset Graves’ ophthalmopathy is uncommon and occurs mostly in girls. Common aspects are edema of eyelid, lid lag, and lagophthalmos. Mild proptosis and conjunctival association occurs in about 10%. CT Scanning or ultrasound may show enlarged extraocular muscles, but significantly limitation of ocular movement is rare and visually threatening problems have not been reported [21]. Serum TSH measurement has the highest sensitivity and specificity of any single blood test used in the evaluation of suspected hyperthyroidism and should be used as an initial screening test [22]. They were highly in our patient FT3 5.44 pg/ml | 2.5-3.5] TSH 0.33 [0.67-4.75] However, when hyperthyroidism is strongly suspected, diagnostic accuracy improves when both a serum TSH and free T4 are assessed at the time of the initial evaluation. [23] Graves’ hyperthyroidism is treated by reducing TH synthesis, using ATD [Anti Thyroid Drug Therapy], or by reducing the amount of thyroid tissue with Radioiodine I-131 (RAI) treatment or total thyroidectomy [24,25]. The treatment used was synthetic antithyroid drugs in our case, ATD are indicated as a first-line treatment of GD, particularly in younger subjects, and for short-term treatment of GD before RAI therapy or thyroidectomy [26,27,28]. Graves’ ophthalmopathy is quite rare in pediatric age and the demonstration of efficacy and safety of oral corticosteroids in this uncommon complication, appear particularly interesting. Moreover, different therapeutic approaches have been considered. Oral corticosteroids, such as prednisolone or dehydrocortisone, have been reported in few cases in the literature, such as initial dose of 1 mg/kg/24 h for the first week and then tapering to 10 mg/24 h, for three months, or an initial dose of 40–60 mg/24 h and then tapering to 15 mg daily for four or six months [29]. The typical treatment of Graves’ disease in children is two years of antithyroid therapy, such as with methimazole, followed by re-evaluation and monitoring thereafter. This data from the literature is so well illustrated by our case since the response was spectacular under synthetic antithyroid drugs. [30].

CONCLUSION
Ocular manifestations are common in pediatric Graves’ disease. However, they are much milder than in adult Graves’ ophthalmopathy. Treatment plans of most children included antithyroid therapy. Children’s symptoms tend to regress after an adequate thyroid status is achieved. Considering how rare Graves’ disease is in children, and the possibility that ocular findings may elude detection by a non-ophthalmologist, we recommend routine ophthalmology consultation for all children diagnosed with Graves’ disease.
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