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Case Report Open Access

Elephantiasis in a Patient with EMO Syndrome

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Abstract

Exophthalmos, myxedema and osteoarthropathy (EMO) comprise the triad known as EMO syndrome. Thyroid dysfunction followed by Graves' ophthalmopathy (GO) commonly was its classic manifestation. The elephantiasic variant was rarely seen. Herein, we reported an unusual case of EMO syndrome associated with severe elephantiasic subtype of pretibial myxedema.

Keywords: Thyroid; EMO Syndrome; Pretibial myxedema; Elephantiasis

Abbreviations: EMO: Exophthalmos, myxedema and osteoarthropathy; GD: Graves' disease; TSH: thyroid-stimulating hormone; GO: Graves' ophthalmopathy.

Introduction

Exophthalmos, myxedema and osteoarthropathy (EMO) comprise the triad known as EMO syndrome, which is a rare extrathyroid syndrome seen in patients suffering from hyperthyroidism, occurring in less than 1% of patients suffering from autoimmune thyroid disease [1]. In its classic presentations, manifestations of Graves' disease (GD) begins with thyroid dysfunction, followed by Graves' ophthalmopathy (GO) in 30% of patients several months later and subsequent thyroid dermopathy in 1% to 4% of patients several months to years later [2,3]. Thyroid dermopathy is a rare manifestation of GD. It occurs in less than 5% of all patients with GD [4]. Much less common is the elephantiasic variant as seen in this case, which is a rare and extreme subtype of Graves' dermopathy. As reported by the classic Mayo Clinic series, it only accounts for 1 of 150 patients with skin involvement [5]. We here describe an unusual case of EMO syndrome associated with severe elephantiasic subtype of pretibial myxedema, which occurs in less than 1% of patients with thyroid dermopathy [6].

Case Report

A 32-year old man was diagnosed with GD seven years ago, followed with radioiodine therapy as initial therapy. Approximately two years previously, the patient was presented with gradual onset of exophthalmos and thyroid dermopathy. An erythema in the right pretibial area gradually enlarged. Both legs, particularly the ankles, significantly swelled. It appeared to be remarkably thickened in deep grade and fixed to the subcutaneous tissues. In particular, these changes progressed and extended relentlessly to the bilateral feet and big toes of both. Chronically localized myxedema resulted in significant lower extremity elephantiasis. The striking abnormality and the hard, unyielding tissue made it unacceptable to use of normal footwear accompanied by compromised mobility. Even worse, in the past few months he had noticed the gradually stiffened skin of the forearm and new-onset skin rashes on the right elbow fossa. Afterwards, he was referred to our department.

Physical examination revealed a significant exophthalmos of both the eyes; swelling and congestion of bilateral eyelids and bulbar conjunctiva, associated with incomplete eyelid closure (Figure 1A) and manifest thyroid dermopathy (Figure 1B). Beyond the presence of myxedema and exophthalmos, there was mildly digital appearance of typical clubbing of thyroid acropachy the patient even did not note and complain of. Besides, hardened skin (Figure 1C) and skin rashes were also found (Figure 1D).

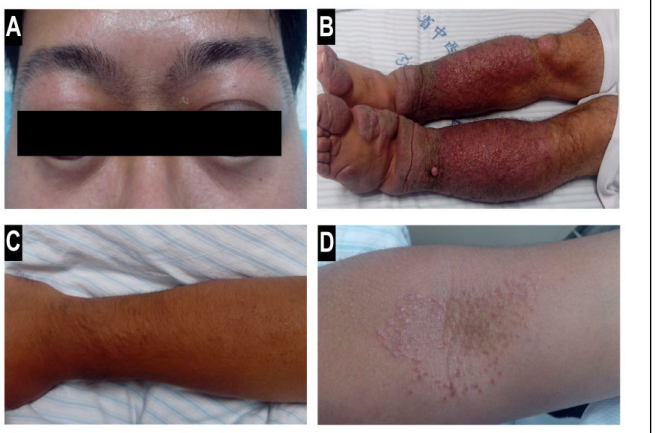

Figure 1A-1D: Clinical manifestation of EMO syndrome. A: the exophthalmos with swelling of bilateral eyelids, bulbar conjunctiva and incomplete eyelid closure; B: thyroid dermopathy on both legs; C: hardened skin; D: skin rashes.

Laboratory tests showed a mildly decreased thyroid-stimulating hormone (TSH) level at 0.528 uIU/mL (normal range, 0.55-4.78 uIU/mL), normal free triiodot-hyronine level at 5.15 pmol/L (normal range, 3.5-6.5 pg/ml), normal free thyroxine level at 15.27 pmol/L (normal range, 11.5-22.7 pmol/L), obviously elevated TSH receptor antibody at 40 mIU/mL (normal range, 0-1.58 mIU/mL), anti-thyroperoxidase antibody more than 1300 U/mL (normal range, 0-60 U/mL) and anti-thyroglobulin antibody at 13.4 U/mL (normal range, 0-60 U/mL). A magnetic resonance imaging scan of the eyes confirmed exophthalmos.

The diagnosis of EMO syndrome was made as the presence of dermopathy, Graves' exophthalmos, typical location and nature of the lesion in this patient with Graves' thyrotoxicosis. The second radioiodine therapy was given and followed with subsequent systemic corticosteroid therapy in combination with immunosuppressive treatment.

Discussion

As the most common cause of hyperthyroidism in non-iodine deficient areas, GD may be presented with extra thyroidal manifestations. It is an autoimmune condition commonly associated with thyroid dysfunction and with positive thyroid antibodies, including TSH receptor stimulating antibodies. On the other hand, pathogenesis of extrathyroidal manifestations is more complex, involving the interaction of immunologic, cellular, and mechanical processes.

Being a component of the EMO syndrome, GD is speculated to have a similar basic pathogenesis with EMO syndrome. Among a number of local factors, differentiation and proliferation of fibroblast and synthesis of glucosaminoglycan are major factors. TSH receptor is associated with pathogenesis of both ophthalmopathy and dermopathy as a common antigen in the skin and eyes tissues. An environmental factor that may contribute to development of these two conditions is tobacco use. Those patients with thyroid dermopathy or ophthalmopathy frequently use tobacco.

The classfication of myxoedema includes the following four forms: non-pitting oedema; plaque; nodular; or elephantiasis. Our case describes an elephantiasis variant, the most severe subtype [6]. Given response to topical corticosteroid therapy is poor in severe cases and dermopathy may be quitely resistant, treatment with systemic corticosteroid application and immune modulators was initiated.

As an extreme manifestation of autoimmune thyroid disease, most commonly thyroid acropachy manifests as asymptomatic digital clubbing associated with severe ophthalmopathy and symptomatic dermopathy. The main goal of treatment generally is to improve ophthalmopathy and dermopathy. No available specific and effective treatments for acropachy have been addressed. However, immunosuppressive therapy and corticosteroids may have beneficial effects on acropachy [7].

With the intention to inhibit, destroy, or remove the thyroid gland, therapeutic strategies for patients with hyperthyroidism have not changed over half a century. However, treatments for the extrathyroid syndrome were so limited and remained to be settled. Particularly, elephantiasis, the most aggressive subtype, was still refractory to conventional therapeutic strategies. Present researches are directed towards genetic and molecular approaches to clarify the pathologies of autoimmune hyperthyroidism and the causes of extrathyroidal manifestations. It may ultimately improve the existing therapies and excogitate innovative treatments.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interests with respect to the authorship and/or publication of this article.

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