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Case Report

Thyroid acropachy: A rare skeletal manifestation of autoimmune thyroid disease

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ABSTRACT

Thyroid acropachy is a rare complication of autoimmune thyroid disease with characteristic imaging findings. Clinically, it presents as nail clubbing, swelling of digits and toes, almost always in association with thyroid ophthalmopathy and dermopathy. On radiographs, it manifests as prominent irregular and spiculated periosteal new bone formation in the hands and feet. We present a 52-year-old man with history of Graves' disease who presented with swelling of the hands and feet. Radiographs of the hands and feet revealed marked diffuse soft tissue edema and characteristic periosteal new bone formation most consistent with thyroid acropachy.

Introduction

Thyroid acropachy is a rare extra-thyroid manifestation of autoimmune thyroid disease. It affects about 0.3% of patients with Graves' disease and often occurs in euthyroid and hypothyroid patients within weeks to many years after treatment of original thyrotoxicosis [1,2]. It is almost always associated with thyroid ophthalmopathy and dermopathy [3]. It manifests as soft tissue swelling with digital clubbing, and periosteal reaction of the extremities.

We present a 52-year old male with history of Graves' disease status-post total thyroidectomy on thyroid hormone replacement, with imaging findings most consistent with thyroid acropachy.

Case report

A 52-year old male with 5-year history of Graves' disease status-post total thyroidectomy on thyroid hormone replacement therapy for 1 year, progressive thyroid ophthalmopathy status-post bilateral orbital decompression and medial rectus recession procedures who presented with progressive swelling of his hands and feet of 8 months duration. Physical exam demonstrated bilateral exophthalmos and significant myxedematous changes to the bilateral hands, fingers, shins, feet, and legs. Laboratory work-up corresponded to a euthyroid state. Imaging of the hands demonstrated significant soft tissue swelling bilaterally, digit clubbing, and irregular periosteal bone formation involving the fifth metacarpals,

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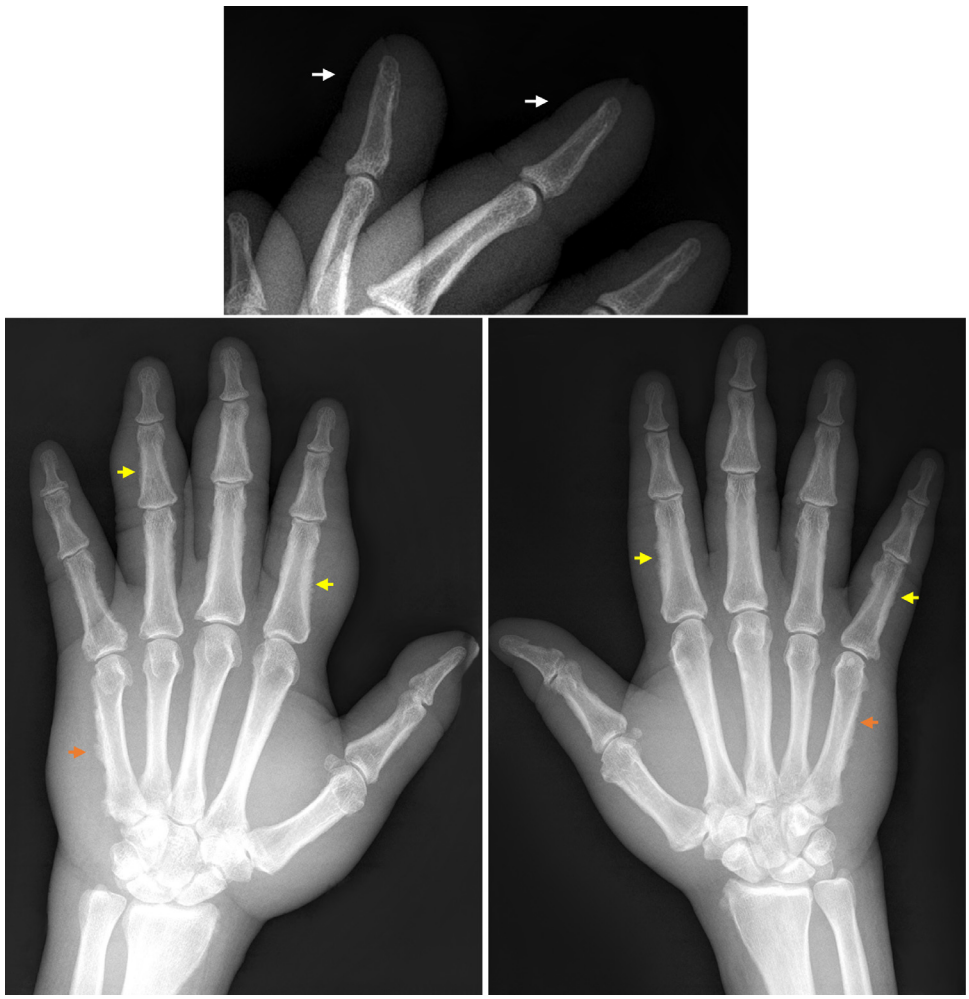

Fig. 1 – Radiographs of the hands demonstrating marked diffuse soft tissue edema, digit clubbing (white arrows), and irregular new periosteal bone formation of on the ulnar aspect of bilateral fifth metacarpals (orange arrows), proximal phalanges and middle phalanges (yellow arrows). (Color version of figure is available online.)

proximal phalanges and to a lesser extent the middle phalanges. The periosteal bone formation was most prominent involving the radial aspects of the proximal second phalanges and ulnar aspect of the fifth metacarpal shafts. Imaging of the feet demonstrated marked diffuse swelling bilaterally. Overall, these findings were most consistent with thyroid dermopathy and accompanying acropachy (Figs. 1 and 2).

Discussion

Thyroid acropachy is the least common extra-thyroid manifestation of autoimmune thyroid disease, with reported incidence of 0.3% in patients with Graves' disease [1]. It can occur in hyperthyroid patients, although it most commonly affects euthyroid or hypothyroid patients after the treatment of original thyrotoxicosis [2,3]. In almost all cases, it occurs concurrently with thyroid ophthalmopathy and dermopathy. In fact, the chronological sequence of extrathyroid manifestations of autoimmune thyroid disease is such that thyroid dysfunction develops first, followed by ophthalmopathy, then dermopathy, and finally, acropachy [3]. The exact etiology is unknown although is thought to be caused by stimulating auto-antibodies to TSH and IGF-1 receptors that are implicated in the pathophysiology of Graves' thyrotoxicosis and ophthalmopathy [4,5].

On imaging, thyroid acropachy exhibits periosteal new bone formation that is highly specific to this syndrome. The periosteal new bone is irregular and spiculated in appearance. Furthermore, these periosteal reactions demonstrate a unique distribution. As such, they tend to be asymmetrical, predominantly involving the hands and feet and only rarely long bones of the forearms and lower legs. The new bone formation tends to be diaphyseal, most pronounced in the midportion of the diaphysis. When the first through fourth metacarpals are involved, changes tend to occur on the radial aspect and when the fifth metacarpal is involved the changes tend to occur on the ulnar aspect of the affected bones [2].

Although therapy consisting of glucocorticoids, radiotherapy, immunosuppressive drugs, and surgery may be beneficial in the treatment of ophthalmopathy and dermopathy, no effective treatment exists for acropachy [1–3].

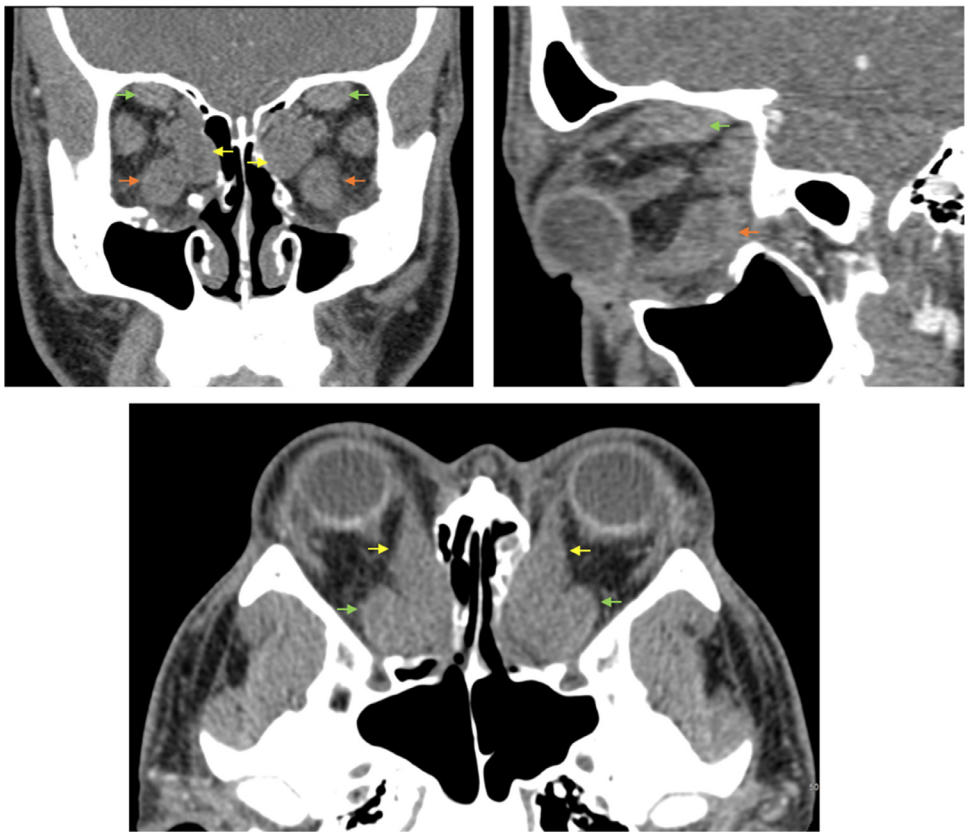

Fig. 2 – CT of the orbits demonstrating bilateral and symmetrical enlargement of superior (green arrows), medial (yellow arrows), and inferior (orange arrow) rectus muscles with sparing of the tendinous insertion as well as increased retrobulbar fat and proptosis in keeping with patient's history of thyroid ophthalmopathy. (Color version of figure is available online.)

Fortunately, thyroid acropachy is a relatively benign condition that is often asymptomatic. The importance of recognizing this entity lies in its accurate diagnosis, as the presence of clubbing and swelling of the extremities may prompt an unnecessary search for pulmonary, cardiac, or hepatic disease.

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