

# Generalized Essential Telangiectasia in a Patient with Graves' Disease: Should the Spectrum of Autoimmune Diseases Associated with Generalized Telangiectasia be Expanded?

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*Generalized essential telangiectasia (GET), as originally described, is not associated with any underlying disease. Although patients with GET lack the typical periungual telangiectases associated with autoimmune collagen vascular diseases, these patients may have an underlying autoimmune process. We present a patient with a history of Graves' disease and low-titer anti-nuclear antibodies, who developed rapidly progressive generalized telangiectases. The gender and age of the majority of patients with GET fit well within the demographics of most autoimmune diseases. The documented occurrence of an autoimmune disease in several of the limited number of patients previously diagnosed with GET provides additional evidence that GET may be associated with an underlying autoimmune disease.*

**W**idespread acquired telangiectasias have been associated with both hereditary and acquired diseases. These include scleroderma, dermatomyositis, systemic lupus erythematosus, syphilis, cutaneous polyarteritis, hereditary hemor-

rhagic telangiectasia, metastatic carcinoid syndrome, angiokeratoma corporis diffusum, ataxia-telangiectasia, portal cirrhosis, and congenital dysplastic angiopathies.<sup>1</sup> In addition, an entity known as generalized essential telangiectasia (GET) has been described without associated disease.<sup>2</sup>

The majority of the patients with GET are female, with onset usually around the fourth decade of life.<sup>2</sup> In general, telangiectatic vessels first appear on the lower extremities, and over a few years to decades, there is progressively more diffuse skin involvement.<sup>2-6</sup> Although lack of an association with systemic autoimmune disease was a criterion for this diagnosis in the original report, our patient, as well as some others who carry this diagnosis, have a documented or probable underlying autoimmune disease.

## Case Report

A 42-year-old white woman with a history of Graves' disease since 1985 presented with widespread telangiectases. She was treated with radioactive iodine, and placed on the thyroid replacement therapy, levothyroxine (Synthroid®). In 1995, the patient developed telangiectases on her thighs, which quickly appeared on her calves, and within a year, on her upper extremities (Figure 1, A and B). Although the patient did develop a few lesions (less than 1 cm in diameter) on her face, she had no periungual or mucous membrane lesions. The patient exhibited no evidence of exophthalmos or pretibial myxedema. Her general health has remained excellent and she has had no complaints of joint pain, muscle weakness, or sun sensitivity.

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**FIGURE 1 A and B.** Bilateral lower extremities show telangiectatic vessels diffusely over the anterior thighs as well as the posterior lower legs.

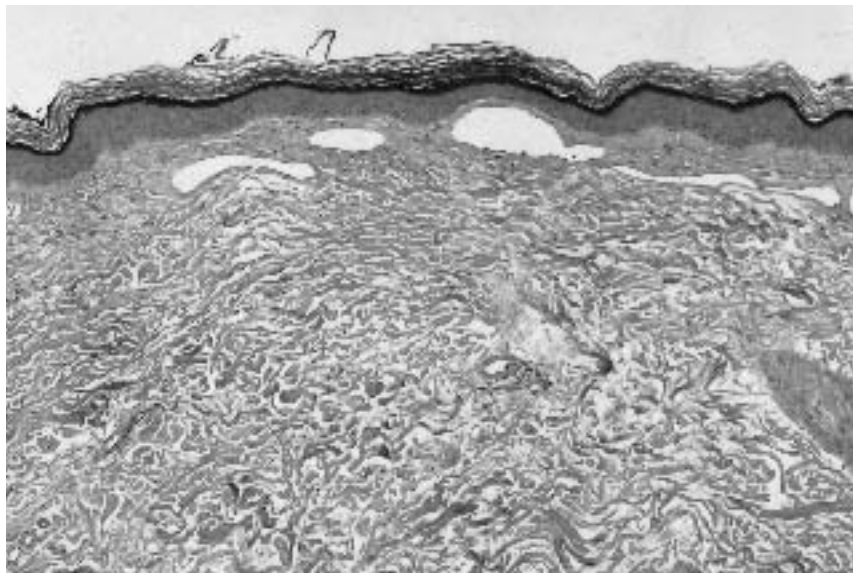
Laboratory studies, including complete blood cell count and differential, blood-urea-nitrogen, glucose, creatinine, serum glutamic oxaloacetic transaminase, lactate dehydrogenase, alkaline phosphatase, total bilirubin, and urinalysis, were within normal limits. Anti-thyroglobulin levels were elevated at 2.40 IU/ml (normal range, 0 to 2 IU/ml). All other thyroid function measurements were within normal limits, including the thyroid-stimulating hormone-receptor antibody. Antinuclear antibodies were positive at a titer of 1/320 with a diffuse pattern, and at a titer of 1/80 with a nucleolar pattern, and anti-histones were positive (H). Anti-Smith, anti-RNP, anti-SSA, anti-SSB, anti-Jo-1, and anti-Scl-70 antibodies were all negative.

Dilated vessels were present exclusively within the papillary dermis, with minimal mononuclear inflammatory infiltrate (Figure 2). An Alcian blue stain at pH 2.5 for acid mucopolysaccharides showed increased staining most marked within the papillary dermis around dilated vessels. After hyaluronidase digestion, there was no increased staining.

### Comments

Most of the patients with GET have been female, and autoimmune disease is more common in females. The usual age of onset of GET is also a relatively good fit for an autoimmune process. In our patient, and in at least one previous report of a patient with GET, there was a documented occurrence of autoimmune disease.<sup>3</sup> Our patient had Graves' disease. As with other patients with autoimmune thyroid disease, she did have low-titer, positive antinuclear antibodies, and she did not have scleroderma, dermatomyositis, or systemic lupus erythematosus, the autoimmune diseases most commonly associated with acquired telangiectasias. In addition, our patient did not have the typical periungual telangiectases commonly associated with collagen vascular diseases, and no mucous membrane lesions.

Reports of cases diagnosed as GET are rare. Although the information available on the patients in the first report of this entity was minimal, two patients, one with thyroid disease and one with diabetes, may both have had an underlying autoimmune process.



**FIGURE 2.** Biopsy specimens show diffuse dilatation of vessels within the papillary dermis (H&E; original magnification, X 150).

The markedly increased staining of hyaluronic acid within the dermis, particularly around the dilated vessels in the papillary dermis, may also be a clue to the association with autoimmune disease, since increased mucin is commonly seen in other cutaneous manifestations of autoimmune disease.<sup>7,8</sup> The patient had no clinical evidence of pretibial myxedema, and there was a slight to minimal increase in mucin in the mid- to lower dermis.

Primary vascular disease is believed to contribute to the pathogenesis of telangiectasia formation in other forms of collagen vascular disease.<sup>1,9-13</sup> The specificity of the involvement may be diverse, and may account for the variable clinical vascular lesions and pathology seen with different autoimmune processes. Although most patients with GET do not appear to have a diagnosable collagen vascular autoimmune disease, at least some of these patients may have some underlying autoimmune process.

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