Generalized essential telangiectasia

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Abstract

The pathophysiology of generalized essential telangiectasia is not well understood. Generalized essential telangiectasia is an uncommon disorder in which widespread telangiectasias of unknown cause develop without associated systemic or antecedent dermatologic disease. We report a case of generalized essential telangiectasia in an otherwise healthy 49-year-old man.

Keywords: generalized essential telangiectasia, telangiectasias

Introduction

Generalized essential telangiectasia is characterized by symmetric blanchable telangiectasias that tend to progress proximally. Dilated capillaries and veins are noted without antecedent skin lesions. The eruption is generally asymptomatic and persists indefinitely without treatment.

Case Synopsis

A 49-year-old man presented to the faculty group practice of The Ronald O. Perelman Department of Dermatology initially in 2014 for the evaluation of discoloration on the lower extremities that had been present for the preceding five years. At the time of presentation, the patient complained of muscle weakness of his calves and denied pruritus, recurrent nose bleeds, fevers, chills, calf pain, claudication, or other systemic symptoms. One year after initial presentation, the eruption spontaneously and dramatically improved, but then recurred within months. Two years after the patient's initial presentation, he also started noticing discoloration on his upper extremities. The patient continued to deny pruritus, recurrent nose bleeds, fevers, chills, calf pain, claudication, or other systemic symptoms.

On the anterior aspect of his shins and the posterior aspect of his calves, there were diffuse, mostly blanchable telangiectasias and finely reticulated erythema in a background of xerotic scale (**Figure 1**). Purpuric and tan patches with many small varicosities were seen on the dorsal aspect of his feet (**Figure 2**). No leg edema was present and 2+ dorsalis pedis and posterior tibialis pulses were palpated bilaterally. A complete blood count, complete metabolic panel, erythrocyte sedimentation rate, C reactive protein level, and vitamin C level were all within normal limits. Tests for antinuclear antibody by indirect fluorescent antibody and anti-Jo-1 antibody were negative. A creatinine kinase level and aldolase level were mildly elevated. Vascular endothelial growth factor levels were low.

Figure 1. *On the anterior aspect of his shins and the posterior aspect of his calves, there were diffuse, mostly blanchable telangiectasias and finely reticulated erythema in a background of xerotic scale.*

Figure 2. *Purpuric and tan patches with many small varicosities were seen on the dorsal aspect of his feet.*

A venous doppler ultrasound was negative for thrombus. A computed tomography scan of the chest/abdomen/pelvis was unremarkable with the exception of incidentally noted pulmonary nodules. The patient is expected to have magnetic resonance imaging studies to further evaluate his muscle weakness.

A 4mm punch biopsy was obtained from the right lateral lower leg. In the papillary dermis there are numerous dilated thin-walled blood vessels (**Figure 3**).

Case Discussion

Generalized essential telangiectasia (GET) is an uncommon disorder in which widespread telangiectasias of unknown cause develop without associated systemic or antecedent dermatologic disease. Distribution of lesions can be generalized over the entire body or localized to a large anatomic area such as the lower extremities, upper extremities, and trunk. Limbs, usually the lower extremities, are initially affected [1, 2]. Rarely, conjunctival involvement has been reported [3, 4].

The lesions are generally asymptomatic and persist indefinitely without treatment. Some patients report associated numbness, tingling, or burning sensations. No associated systemic diseases have been reported [5]. Rarely, associated gastrointestinal tract bleeding has been reported [6].

Generalized essential telangiectasia primarily affects adult women but has been reported as early as childhood. It has been reported to be more common in Caucasian individuals [7]. The pathophysiology of generalized essential telangiectasia is not well understood.

The differential diagnosis for GET includes hereditary hemorrhagic telangiectasia and cutaneous collagenous vasculopathy (CCV). CCV is another disorder with an unknown etiology. GET and CCV differ in that CCV does not have a female predominance, does not exhibit caudal progression, and often affects only the trunk and/or proximal extremities. Histologic findings in CCV that are not demonstrated in GET include ectatic superficial blood vessels with hyalinized and laminated, concentric concretions around vessel basement membranes [8].

Treatment of individual areas of GET can be pursued with lasers, including both the 585-nm pulsed dye and long-pulse Nd:YAG lasers [5, 9, 10]. Other reported treatments include tetracycline and ketoconazole [11-12]. Whether it is an anti-inflammatory or antibacterial mechanism through which tetracycline may be effective has been debated in the literature [11]. The authors of the original report regarding the use of ketoconazole postulated that microbially-induced focal intravascular coagulation played a role in the pathogenesis of GET, which, at that time, was termed progressive ascending telangiectasia.

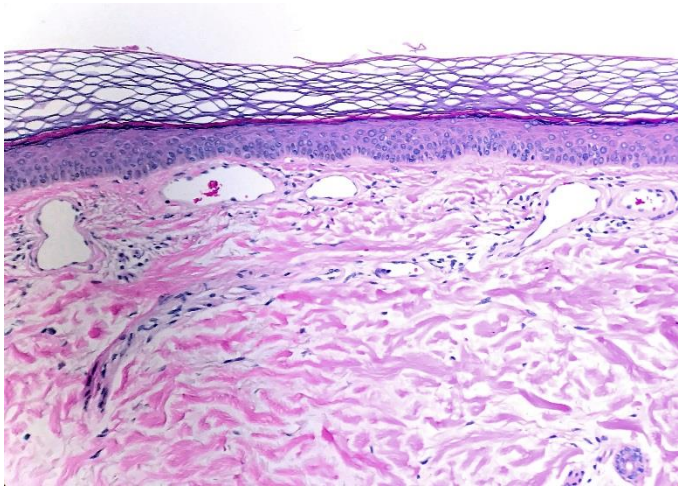

Figure 3. *A 4mm punch biopsy was obtained from the right lateral lower leg. In the papillary dermis there are numerous dilated thin-walled blood vessels. H&E, 10×.*

Given that our patient's eruption initially improved but then recurred without treatment, the possibility of an inflammatory component was raised, although the biopsy was not supportive. Our patient initially deferred treatment as the eruption was asymptomatic. Given the progression of the eruption, topical rapamycin was trialed without benefit. More recently, he began an empiric trial of oral doxycycline with some benefit. He is now considering pursuing laser treatment, which he initially deferred.

Conclusion

Generalized essential telangiectasia is an uncommon disorder in which widespread telangiectasias of unknown cause develop without associated systemic or antecedent dermatologic disease. The pathophysiology of generalized essential telangiectasia is not well understood. We report a case of generalized essential telangiectasia in an otherwise healthy 49-year-old man.

References

1. Rothe MJ, Grant-Kels JM. Nomenclature of the primary telangiectasias. *Int J Dermatol*. 1992;31:320. [PMID: 1587658].
2. McGrae JDJr, Winkelmann RK. Generalized essential telangiectasia: report of a clinical and histochemical study of 13 patients with acquired cutaneous lesions. *J Am Med Assoc*. 1963;185:909-913. [PMID: 14044221].
3. Ali MM, et al. Generalized essential telangiectasia with conjunctival involvement. *Clin Exp Dermatol* 2006; 31:781. [PMID: 17040263].
4. Mannis MJ, et al. Ocular manifestations of generalized essential telangiectasia. *Cornea* 1999; 18:731. [PMID: 10571307].
5. Bolognia J, Jorizzo JL, Schaffer JV. Dermatology. 3rd ed. Philadelphia: Elsevier Saunders; 2012.
6. Checketts SR, et al. Generalized essential telangiectasia in the presence of gastrointestinal bleeding. *J Am Acad Dermatol* 1997; 37:321. [PMID: 9270538].
7. McGrae JD Jr, Winkelmann RK. Generalized essential telangiectasia: report of a clinical and histochemical study of 13 patients with acquired cutaneous lesions. *J Am Med Assoc* 1963; 185:909. [PMID: 14044221].
8. James Wd, Berger TG, Elston DM. Andrews' Diseases of the Skin: Clinical Dermatology, 12th edition. Elsevier Saunders; 2012.
9. Buscaglia DA, Conte ET. Successful treatment of generalized essential telangiectasia with the 585-nm flashlamp-pumped pulsed dye laser. *Cutis* 2001; 67:107. [PMID: 11236217].
10. Gambichler T, et al. Generalized essential telangiectasia successfully treated with high-energy, long-pulse, frequency-doubled Nd:YAG laser. *Derm Surg* 2001; 27:355. [PMID: 11298706].
11. Shelley WB, Fierer JA. Focal intravascular coagulation in progressive ascending telangiectasia: ultrastructural studies of ketoconazole-induced involution of vessels. *J Am Acad Dermatol* 1984; 10:876. [PMID: 6327785].
12. Shelley WB. Essential progressive telangiectasia: successful treatment with tetracycline. *J Am Med Assoc* 1971; 216:1343. [PMID: 5108457].