

A 34-Year-Old Woman with an Unusual Rash

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CASE PRESENTATION

Initial Presentation and History

A 34-year-old woman presented to the clinic with a complaint of an unsightly rash on her posterior thighs (Figure). She had first presented with the rash to another primary care physician 1 month ago. At that time, the physician discontinued her only medication, hydrochlorothiazide, which she had been taking for hypertension for many years, as he thought the rash may have been related to the drug. He also prescribed a topical medium-potency steroid cream, triamcinolone 0.1%. She used the cream as directed, but the rash did not improve. Frustrated over the lack of improvement, she sought a second opinion. On presentation, she denied itch, tenderness, or pain as well as any history of irritant/allergen exposure. The patient had no other skin changes, lesions, or ulcers. On further questioning, the patient stated that as the weather had been cold she had made a practice of standing next to her wood stove several hours nightly for warmth before going to bed. She had been doing this for several months.

Physical Examination

The general physical examination was unremarkable. Skin examination revealed areas of reticulated, mildly raised plaques with erythematous to dark brown pigmentation on the skin of the posterior thighs bilaterally. The pigmentation on the left thigh appeared to be more pronounced than that on the right. Scattered within the pigmentation were areas of red superficial ulcerations. There was no pain, tenderness, or discomfort associated with the lesions. Palpation demonstrated normal subcutaneous tissue texture.

Laboratory Testing

Laboratory studies showed normal complete blood count, electrolytes, and renal, hepatic, and thyroid func-



Figure. Rash observed in case patient.

tion. Serologic tests for antineutrophil antibody and rheumatoid factor as well as the erythrocyte sedimentation rate were normal. The patient refused biopsy.

WHAT IS YOUR DIAGNOSIS?

- (A) Erythema ab igne
- (B) Ichthyosis
- (C) Livedo reticularis
- (D) Psoriasis

Dr. Tran is an attending physician, and Ms. Shannon is a family nurse practitioner; both are in the Department of Medicine, Kyle Health Center, Kyle, SD.

ANSWER

The correct answer is (A), erythema ab igne (EAI).

DISCUSSION

This patient's hyperpigmented skin discoloration coupled with the clinical history of heat exposure is consistent with a diagnosis of EAI. EAI is an uncommon skin condition that results from chronic repeated exposure to heat or infrared radiation.¹ EAI is characterized by a lacy reticulated pattern of erythema or pigmentation that is light to deep brown in appearance; hence, the name *erythema ab igne*, which literally means redness from heat. Livedo reticularis is a netlike mottled violaceous skin discoloration of the extremities or torso. While livedo reticularis may look similar to EAI due to its netlike skin pattern, it lacks the essential clinical history of heat exposure and the brown hyperpigmentation seen in EAI. Unlike EAI, which has no known association with a systemic condition, livedo reticularis is often observed as a physiologic skin manifestation of systemic clinical significance. It can result from cold temperature exposure or from skin reaction to a systemic vascular condition. Livedo reticularis has been associated with hypercoagulability (ie, antiphospholipid antibody syndrome, factor V Leiden mutation, protein C deficiency),^{2,3} livedoid vasculopathy,⁴ metabolic disorder (ie, hyperhomocysteinemia),⁵ hepatitis C virus infection,^{6,7} and cryoglobulinemia.⁶⁻⁸

Ichthyosis is characterized by varying hyperkeratotic changes of the skin with the appearance of fishlike scales and is often pronounced in the lower extremities. It is frequently associated with atopy. Psoriasis, a dermatosis histologically characterized by excessive epidermal cell growth, is usually found as itchy erythematous plaques with thick whitish scale. The elbows, knees, scalp, and intertriginous areas are the favored sites.

ERYTHEMA AB IGNE

Clinical History and Presentation

Because EAI results from chronic repeated exposure to heat or infrared radiation, the practice of sitting near hearth fires or coal/wood stoves during cold weather is a notable predisposing factor for developing this condition, as seen in the case patient. EAI was more frequently encountered when coal and wood stoves were widely used as a means of household heating;⁹ the incidence of EAI has decreased since the advent of central heating.¹⁰⁻¹²

Presently, use of hot water bottles, infrared lamps, heating pads, and heated recliners for therapeutic relief of chronic pain from osteoarthritis, back problems,

and pain from malignancy have been associated with EAI.^{10,11} Frequent hot bathing,¹³ prolonged exposure to heat generated from a laptop computer,¹⁴ and the use of a car heater have also been reported to induce the development of EAI.¹¹ A patient with altered mental status in an intensive care setting developed EAI iatrogenically due to a malfunctioning heating/cooling blanket.¹⁵ Another patient with chronic arthritis pain of the right wrist and knee developed EAI after applying microwave-heated popcorn kernels to the affected joints for 30 minutes per treatment for a period of more than 4 months.¹⁶ Although the use of heat for musculoskeletal pain relief of extremities is common, its use for pain relief of abdominal or back pain should alert clinicians to evaluate for underlying systemic pathology. For example, cases of patients who developed EAI after chronic use of heat therapy for relief of severe pain from acute or chronic pancreatitis or pancreatic carcinoma have been reported.^{17,18}

The distribution of the skin changes seen in EAI depends on the location of the heat source. Bakers or cooks who work with open fire often exhibit EAI on the face or palms.^{10,19} In most cases, EAI has no symptoms other than an unsightly skin discoloration. Patients may complain of mild pruritus and a burning sensation,¹⁰ particularly those who have superficial ulcerations that may be due to mild burns.⁹

Histopathology

The majority of reported cases of EAI are benign,¹² but this condition has been associated with cases of epidermal hyperkeratotic nodules with atypia resembling actinic keratosis or Bowen's disease,²⁰ squamous cell carcinoma in situ,²¹ squamous cell carcinoma,^{20,22} endothelial atypia,²³ and neuroendocrine (Merkel cell) carcinoma with squamous carcinoma.^{24,25} The usual early histopathology of EAI shows hyperpigmented epidermal atrophy and flattening with focal regressive changes of basal keratinocytes.²⁶ Electron microscopy shows functional activation of melanocytes with increased dendritic processes.²⁶ The dermoepidermal junction is flattened.²⁷ In the dermis, there is an abundance of melanophages and sporadic elastic fiber changes.²⁶ The dermis is thin, with moderate edema, connective tissue disruption, and accumulation of elastic staining material (elastosis),²⁷ and is also infiltrated with mixed cells.²⁸ Additionally, hemosiderin and melanin granules can be found within the dermis.²⁷ The dermal capillaries and venules are dilated.²⁸ The late histopathology shows varying collections of elastotic fibers, hyperkeratosis, necrotic keratinocytes, dyskeratosis, and/or atypia.²⁸

Diagnosis

The diagnosis of EAI is made by clinical history and physical examination findings. Patients with the characteristic EAI rash almost always have a history of chronic heat or infrared radiation exposure, the duration of which can range from weeks to months and even years. Although skin biopsy of the EAI rash adds additional histologic information, it is often reserved for patients with chronic painful ulcerations, excoriations, or a history of prolonged periods of heat exposure as these findings may raise suspicion for dermal dysplasia or neoplasm (eg, squamous cell carcinoma or Merkel cell neuroendocrine carcinoma).^{24,25}

Management

The primary treatment of EAI is to avoid further exposure to heat sources. The skin discoloration has been reported to gradually lighten over time.⁹ Short-term improvement of the skin hyperpigmentation can be achieved with photothermolysis with various types of laser (Nd:YAG, ruby).¹⁰ Keratinocytic dyskeratosis has been treated with 5-fluorouracil cream.²⁹ Patients with EAI should be followed periodically for early detection of suspicious growths such as squamous cell and other cell changes, as previously discussed.

CLINICAL COURSE OF CASE PATIENT

The patient was told to avoid further heat exposure and to follow-up in several months for reevaluation. However, she did not return for follow-up. **HP**

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