

Rethinking the Treatment Paradigm of Greyscale: A Case Report

Dr. Samwell Tarly*

102 San Salvador Ln, Santa Fe, Edinburgh, United Kingdom

***Corresponding Author:** Samwell Tarly, 102 San Salvador Ln, Santa Fe, Edinburgh, United Kingdom.

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Abstract

Greyscale is a rare, highly contagious, disfiguring, and ultimately fatal disease that traditionally has led to patient quarantine. We present a case report of a patient who was successfully cured with aggressive surgical debridement.

Keywords: Greyscale; Dermatology; Plastic surgery

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Introduction

Colloquially known as “Prince Garin’s Curse” and “the grey death,” greyscale is an extremely rare (incidence 1.2/100,000), highly contagious skin condition that has a high mortality (5-year survival of 23%) [1]. While greyscale can affect people of any age, it is most common in the pediatric population, and has a higher incidence in cold, damp climates [1]. While the underlying pathogen has never been isolated, it is known to be transmitted via skin-skin contact with an afflicted individual. There is even a case report of a neonate contracting the disease after coming into contact with a doll owned by someone with the disease [2].

Classically, greyscale is considered to have no cure. Aggressive and timely surgical debridement has had some success in the past, but the high rate of surgeons contracting the disease has since led to quarantine of these individuals [3]. This disfiguring skin condition may take years to reach terminal state. During this time, the lesions commonly spread to cover the entire body, leading some to refer to these patients as “stone men.” Eventually, involvement of the internal organs (including cardiac and pulmonary involvement) leads to death. [4] Brain involvement is very common, with many reports of psychosis and aggressive behavior. The erratic behavior of affected patients has directly lead to others contracting the disease [3]. The following is a noteworthy case of a patient with greyscale who was successfully cured of the disease. The surgeon successfully employed barrier technique and maintained sterility during the debridement, avoiding contracting the disease himself. We hope that this case may lead to more treatment and research for these often-neglected patients.

Case Report

JM is a 57-year-old previously healthy male who presented to Citadel Community Hospital (Oldtown, Westeros) for a second opinion. Three weeks prior to his presentation at the hospital, his left forearm was exposed to a non-quarantined individual, whom he believed to have the extremely rare and highly contagious condition known as “greyscale.” Skin changes appeared around the inoculation

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site on his forearm within 24 hours of exposure, despite the absence of a penetrating wound. Over the next few days, the skin became cracked and flaky, eventually developing a grey colour. JM sought medical attention as lesion began to spread to the rest of his left arm. An outside specialist made a clinical diagnosis of greyscale, informed the patient that there was no cure, and recommended quarantine.

Upon presentation to CCH, the patient was immediately placed in quarantine and examined by a physician using precautionary barrier protection. Vital signs were normal. Physical examination demonstrated that the hyperkeratotic condition had spread to involve the entire left arm and much of the torso (Figure 1), and purulent material could be expressed from beneath scaled areas by the patient.

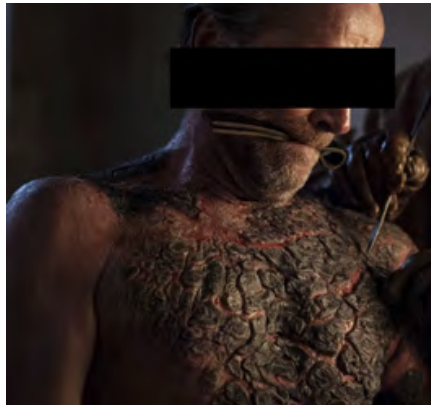


Figure 1: Greyscale is shown covering the patient's trunk.

The patient did not have pain sensation during the pinprick test in affected areas. The remainder of the physical examination was normal. The patient scored 30 points on a mini-mental state examination. Laboratory examination included CBC (WBC 14.5, Hgb 13.8, Plts 557), CMP (Na 142, Cl 100, K 4, CO₂ 22, BUN 20, Cr 1.1, Glucose 104, TP 7.4, Albumin 4.2, Ca 9.1, Tbili 1.1, AST 25, ALT 32, Alk Phos 102). ESR was elevated to 56. Blood cultures were negative. Ultrasound did not demonstrate a drainable fluid collection or abscess.

The patient underwent punch biopsy. Microscopic evaluation showed predominantly necrosis with extensive calcification superficially, with granulomatous reaction deep to this. Within the subcutaneous tissues, there was inflammatory infiltrate and abundant neutrophils. The diagnosis of greyscale was confirmed due to the classic presentation and characteristic pathologic findings.

After treatment planning conference, CCH Chief of Plastic Surgery Dr. Tarly made the decision to pursue aggressive surgical debridement. This was despite the conventional wisdom that the risk of disease transmission to the surgeon was too great, and that the prognosis for the patient was poor, even with treatment. The patient was taken to the OR, and the affected areas of the left upper extremity and trunk were successfully debrided. The area was then coated using an experimental treatment previously described by Pylos., *et al.* [5] using an ointment with extracts from pine resin, green bark from elder twigs, white beeswax, and olive oil.

The patient recovered in the hospital over the next few days with supportive care, without complication. He was subsequently discharged and remained disease-free until his subsequent death due to penetrating trauma. The surgeon did not contract the disease despite close contact during the surgical debridement.

Discussion

The successful treatment of JM has led to a rethinking of the treatment of patients with greyscale. JM was cured of the disease despite extensive involvement of the left upper extremity and trunk. While this treatment cannot be undertaken for patients with disease involving a greater body surface area or central nervous system involvement, many patients present earlier in the disease course, and may benefit from this approach. Palliative treatment, which typically includes limes, mustard poultices, and hot baths, have been largely ineffective.

The greatest hurdle to treatment for these patients will be a shortage of physicians willing to do the procedure. Contraction of the disease by surgeons attempting this was common in the past, however with improved sterile technique and barrier protection we believe this risk can be minimized. Further research into this condition should include re-evaluation of the surgical techniques, and attempts should be made to elucidate the pathogen causing the disease with an eye toward developing a vaccine.

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