Regarding the True Aetiology of the Skin-Lightening Syndrome in "The Adventure of the Blanched Soldier"

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From Wednesday, January 7 through Monday, January 12, in the year 1903, Mr. Sherlock Holmes investigated the mysterious malady that seems to have afflicted Mr. Godfrey Emsworth, and which is the subject of this essay.(1) Since Mr. James M. Dodd, Mr. Holmes' client and the former army comrade-in-arms of the afflicted man, had not had any communications from his friend for six months, he visited the home of Mr. Emsworth to find him to be hiding on the estate but mysteriously changed in physical appearance. Through a series of observations and scientific deductions, prior to and during the visit of Messrs. Holmes and Dodd to the Emsworth habitation, Mr. Holmes reached the tentative hypothesis that Mr. Emsworth may have contracted leprosy in South Africa, and was hiding this disastrous news from everyone except his family, trusted servants, and personal physician. It was this latter gentleman, a Mr. Kent, the primary care physician, who diagnosed the condition as leprosy without calling in an expensive consultant to confirm his findings. Deprived of the services of John H. Watson, M.D., who usually filled the role of physician-advisor to the detective but who had deserted him for a wife, Mr. Holmes was required to call in a favor and ask the noted dermatologist and tropical medicine specialist Sir James Saunders to provide a long-needed second opinion in this matter. The diagnosis of "pseudo-leprosy" or ichthyosis was gratefully received by all in attendance at the denouement of this adventure.

As is often the case in the field of medical literature, some controversy has arisen regarding the true aetiology of the skin condition that afflicted Mr. Godfrey Emsworth. Was it really leprosy as Mr. Kent had initially diagnosed it? Was it really "pseudo-leprosy" or ichthyosis, as stated by the expert Sir James Saunders? Was it something else again, as discussed in more recent medical literature(2,3)? It is unfortunate that Dr. Watson had not the opportunity to present this account to us. I am certain that the clarity of his medical observations would have obviated all of the obscurity associated with this narrative by a scientifically brilliant though medically inexpert detective. Thus, we must make do with the evidence that is available to us through the less than medically expert descriptions provided by Mr. Sherlock Holmes' account to explore the several possible alternative diagnoses(1).

Medical diagnoses are generally based on considerations of patient history, presenting symptoms, and the results of laboratory tests, and on occasion, surgical intervention. In this case, neither laboratory analyses nor diagnostic surgical procedures were performed. Thus, we are forced to rely exclusively on the patient's case history and symptomology, and the diagnostic skills and personal experience of the physician.

Mr. Godfrey Emsworth had seen military action in South Africa during the Boer War(1). According to his comrade, Mr. James M. Dodd, "They took the rough and smooth together for a year of hard fighting. Then he was hit with a bullet from an elephant gun in the action near Diamond Hill outside Pretoria. I got one letter from the hospital in Cape Town and one from Southamton. Since then not a word-not one word, Mr. Holmes for six months and more, and he my closest pal." Then, we continue with Mr. Emsworth's own account that after he was wounded, he spent the night collapsed in a leper's bed in the Leper Hospital, and during this time that he was in a weakened condition, he had a brief but close encounter with one of the infected inhabitants of the facility.

Let us first examine the foregoing, while the klews are fresh in our minds, before discussing the clinical signs. For approximately one year, Mr. Emsworth was subjected to the exigencies of warfare. There must have been many occasions during which he was malnourished, poorly protected from the environment, and lacking in proper hygiene. All of these influences may have reduced his resistance to infection. However, there is no evidence that anything was amiss for the six-month interval between his being wounded and the time that he arrived in Southampton, prior to returning home. Then, for some reason, probably the rapid visibility of horrendous symptoms, he dropped out of sight, thinking that he was a victim of leprosy, and afraid of the terrible social consequences associated with this misunderstood affliction. Thus, whatever disease revealed itself did so very quickly and completely during the time Mr. Emsworth was disembarking from Southampton or just as he arrived home. It was not a long-standing disease that can be traced back to childhood nor one that would take a long time for symptoms to reveal themselves.

Now let us turn our attention to the signs and symptoms[1]. It should be noted that Mr. Dodd's visit, that resulted in his observations of Mr. Emsworth, were made some weeks after the last communique, thus more than six months after the events that ended the military career of Mr. Emsworth. Consequently, the symptoms described by Mr. Dodd, Mr. Holmes, and Sir James are not those of the initial stages of the disease. As viewed in the window, illuminated by lamplight, Mr. Dodd provided the following description: "He was deadly pale - never have I seen a man so white." And further: "...that ghastly face glimmering as white as cheese in the darkness." And again: "His face was - how shall I describe it? - it was of a fish-belly whiteness. It was bleached." In response to Sherlock Holmes' query, it appeared that the face was not "equally pale all over." Then, we have Mr. Holmes' own written description: "One could see that he indeed had been a handsome man with clear-cut features sunburned by an African sun, but mottled in patches over this darker surface were curious whitish patches which had bleached his skin." Finally, the words of the great dermatologist/tropical disease expert Sir James Saunders: "A well marked-case of pseudo-leprosy or ichthyosis, a scale-like affection of the skin, unsightly, obstinate, but possibly curable, and certainly non-infective."

There we have it. First, the disease had a rapid progress from the time it was not evident until the time that it manifested itself with significantly marked symptoms. Secondly, the lesions on the face were patchy, very white, and scaly. And thirdly, an eminent dermatologist/tropical disease specialist had ruled out leprosy.

The subject under discussion has not been completely ignored by medical scholars[2,3]. Dr. Herman Beerman, in several landmark papers, discussed several diseases that appeared to him, based on his extensive experience as a dermatologist, as the agent responsible for Mr. Emsworth's discomfort[3]. He concluded that the disease was more likely the affliction vitiligo or the fungal infection tinea versicolor, an ubiquitous disease produced by *Pityrosporon orbiculare* (formerly *Malassezia furfur*), rather than ichthyosis. He said: "Unless Godfrey Emsworth had had scaling skin lesions since infancy he certainly did not have ichthyosis." In considering vitiligo he stated: "...what is the differential diagnosis of scaly, hypopigmented facial patches occurring in adulthood during wartime or post-war conditions? Vitiligo is one diagnosis." However, there were no evidences of preexisting conditions such as hyperthyroidism, hypoadrenalism, or Addison's disease, and "furthermore, vitiligo is not scaly." On the other hand, "Tinea versicolor is found in "equal frequency in temperate and tropical zones, and one which an unbathed soldier might easily have contracted." I can state from personal experience, subsequent to short time exposure to my sister's cat, that tinea versicolor develops rapidly after contact producing significant clinical manifestations.

With these few facts and hypotheses in mind, let us now turn our attention to each of the possible aetiologies of Mr. Emsworth's disease: leprosy, ichthyosis, vitiligo, tinea versicolor, and to add a new entry, xeroderma.

The fact that the family physician Mr. Kent misdiagnosed the affliction as leprosy should come as no surprise for several reasons[4,5], although according to some, "early clinical indications of skin lesions and muscular and neurologic deficiency are usually significantly diagnostic in patients from endemic areas"[6]. Even today, there are many cases in which a wide variety of illnesses have been attributed to leprosy, especially to the inexperienced observer. According to Mr. Keith Skillicorn, in his experience a review of other sources there are at least 30 diseases that may be wrongly attributed to the leprosy bacillus[4]. He states that "there are four Cardinal Signs of leprosy, AT LEAST TWO of which must be seen in a patient before we can safely diagnose that person as having leprosy: - (1) Hypopigmented, localized skin patches, - (2) Anaesthesia or sensory deficit, particularly of touch and temperature, - (3) Thickened nerves, particularly peripheral nerves, - (4) Non-cultivatable, acid-fast bacilli present in skin lesions and/or nasal mucosa. In addition, leprosy produces anhidrosis or absence of or deficiency of sweating of the skin, is not highly contagious requiring continuous close contact for transmission, and has an unusually long - 6 months to 30 years - incubation period to manifest itself; only about 5% of contacts acquire the disease, and it rarely appears as hairless hypopigmented patches until later stages of the disease[5,6]. Although Sir James Saunders did not appear have used the services of a laboratory, he was no doubt sufficiently experienced to make the diagnosis on clinical grounds and the brief history of exposure and short time to onset.

Let us now turn our attention to Sir James's diagnosis was "pseudo-leprosy or ichthyosis." According to Beerman, Sir James' use of the term pseudo leprosy or ichthyosis was not a definitive diagnosis, but that "it is more likely that the specialist was more sure of what the condition wasn't than what it was. In other words, he used these terms with the primary intention of reassuring young Emsworth that he did not have the disease he so feared."[3] Ichthyosis is an inherited "disease characterized by excessive accumulation of scale on the skin surface."[5] Further, the age of onset is from birth to early childhood. As Dr. Beerman said: "Unless Godfrey Emsworth had had scaling skin lesions since infancy he certainly did not have ichthyosis."[3] A similarly appearing disease, xeroderma, might be implicated. However, this affliction "usually occurs on the lower legs of middle-aged or older patients most often in cold weather and in patients who bathe frequently."[5] This also does not match the pattern of history, patient age, and disease site that were described previously for the patient.

Vitiligo is another condition that merits our attention. After all, several statements in the narrative refer to a very white or pale facial appearance- "white as cheese" and fish-belly whiteness."[1] Who can forget the remarkable transformation that this disease has made on the facial appearance of a Mr. Michael Jackson, a singer of some fame? This disease is often misinterpreted as leprosy by inexperienced practitioners, and a recent poignant account by a dermatologist stresses the importance of differentiating these two disorders which can have very profound social consequences in certain societies.[7] There is no loss of sensation in vitiligo as there is in leprosy, and no loss of sweating so that the patch is not warmer as in leprosy.[4] As pointed out by Dr. Balin, the lesions develop slowly over time, not precipitously as described in our story.[7] In addition, the lesions are subject to sunburn.[5] and thus could not have been so prominent in the lamp light. Also, there is no evidence that Godfrey Emsworth suffered from any of the preexisting associated conditions: pernicious anemia, hypothyroidism, hypoadrenalism, pernicious anemia, or Addison's disease, "And, furthermore, vitiligo is not scaly."[3]

That leaves us with Dr. Beerman's favorite diagnosis - the fungal infection tinea versicolor, an ubiquitous disease produced by *Pityrosporon orbiculare* (formerly named *Malassezia furfur*). Unlike vitiligo, "Tinea versicolor is found in equal frequency in temperate and tropical zones, and one which an unbathed soldier might easily have contracted." I can state from personal experience, subsequent to short time exposure to my sister's cat, that tinea versicolor develops rapidly after contact producing significant clinical manifestations. Any microbiologist can very easily ascertain the evidence for tinea versicolor "by finding groups of yeast and short plump hyphae on microscopic examination of scrapings from the lesions."[5] The

lesions have been described as "tan, brown, or white, very slightly scaling, which tend to coalesce, are seen on the chest, neck, and abdomen and occasionally on the face.[5] However, although the descriptions of the clinical signs encountered in this story under discussion may somewhat match those listed above, this disease is relatively widespread and should be easily recognized by a competent physician, even one who is not an expert in dermatology and tropical medicine. On the other hand, tinea versicolor is a disease that may be confused with leprosy. Differentiation is accomplished by microscopic examination of skin scrapings and noting that there is no loss of sensation at the site [4]. It is almost certain that this clearly recognizable disorder would be named as such by the dermatologist Sir James Saunders, rather than using the term "pseudo-leprosy." However, this is mere speculation, and we cannot definitively rule out tinea versicolor on these grounds.

In conclusion, we have analyzed the paltry available data for the aetiology of the condition that afflicted Mr. Godfrey Emsworth, as recounted to us by Mr. Sherlock Holmes in his narrative "The Adventure of the Blanched Soldier."[1] Although have reviewed klews in an attempt to define which of several conditions might fit the few pieces of information available to us, and the discussions of a highly qualified expert[3], we can draw no final conclusions, except the very felicitous conclusion that it is not leprosy. Perhaps the main message to come out of this adventure is the unfortunate fact that, as pointed out by Mr. Skillicorn and Drs. Balin, even in today's sophisticated medical climate many diseases are still misidentified as leprosy, [4,7] resulting in horrendous psychological and social consequences to the innocent victim of this medical error.

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