18 Stories of the Skin

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The Butterfly Child and Other Patients

By

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“Why, sometimes I’ve believed as many as six impossible things before breakfast.”

—The White Queen in Lewis Carroll’s,
Alice Through the Looking Glass

TABLE OF CONTENTS

ABOUT THE AUTHOR

Dr. Robert A. Norman is a board-certified dermatologist and family practitioner who has been in practice for over 25 years. He is a faculty member in several medical schools (Clinical Professor) and has been honored with numerous service and teaching awards, including Physician of the Year (2005); the Distinguished Service Award (2007) in Hillsborough County, Tampa, Florida; Tampa Bay Medical Hero Award (2008); and the Hadassah Humanitarian Award (2012). Dr. Norman has written 50 books, including *The Blue Man and Other Stories of the Skin* (University of California Press) and *Discover Magazine's Vital Signs—True Tales of Medical Mysteries, Obscure Diseases, and Life-Saving Diagnoses*. He has been the editor and contributing writer of eighteen textbooks on Geriatrics and Geriatric Dermatology and has published over 300 articles in various major media publications. He is the founding and current editor of the Springer Series on Clinical Cases in Dermatology.

Dr. Norman has a private practice and is the chief physician and owner of Dermatology Healthcare, founded in 1994, which delivers essential skincare services to nursing home patients. He delivers many national and international lectures each year; in October 2007, he was the chairman and lecturer in Geriatric Dermatology at the World Congress of Dermatology in Buenos Aires, Argentina. In 2014, he was a Keynote Speaker at the Australasian Skin Cancer Conference in Brisbane, Australia, and was a co-chairman in Geriatric Dermatology at the World Congress of Dermatology in Vancouver in 2015. He spoke at the 2016 Pan-American

Congress of Neurology and the 40th Mexican Academy of Neurology Annual Meeting in 2016 on *The Skin: Our Outer Brain*. He also has both MPH and MBA degrees and was awarded the Doctor of Humane Letters after providing the commencement address at a major university.

PREFACE

Nature has marvels and mysteries, and sometimes acts in ways that are almost beyond our imagination. Many of us are fortunate enough to have skin that reflects mostly good quality experiences, with perhaps more turbulent times on occasion. Our natural tapestry, the skin, can also be torn apart by disease, and we can suffer from horrendous physical and emotional trauma.

Over 30 years of medical care, I have looked after patients with enormous tumors erupting on their faces and chests, others with fragile blisters on their extremities, and a group with progressive pigmentary changes who result in being social and economic outcasts. We will explore a number of skin diseases, from the more common such as acne, which when severe, can rise up angrily in the teens and twenties and beyond if left untreated, to among the most rare, given the abbreviations PXE, DEB, and XP.

Think of each chapter in a cinematographic way—each person has a story that you are witnessing—and I will supply the interactive narrative. For example, I often see patients with neurodermatitis—who are weighed down by a heavy psychological influence—and I want the story to unfold with conversations that you will be in on. (For more visual input on each disease, you may want to consult the internet as you read.)

The book's organizing principle is each patient's story and the search for the fundamental humanity, compassion, and empathy for those with a wide variety of skin and systemic issues. Amid the pages will also be unforeseen happenings and problems to be solved. The book reflects the sociology of the skin—the people, the environment, and the mysteries. Any particular disease can take on a whole different gestalt in one person as opposed to another.

One central element here is the need to dignify a person's life in the face of disease—something that is often not their fault. The attempts to make a positive investment in each patient's future with an inoculation of interest and hope will be highlighted, hopefully with a natural, organic feel. Each person is part of an anthology in a series, an experience of anecdotes and events that are weaved together with each new appointment. As the book's narrator and producer, I witnessed the words and actions being born, and I am part of the team, the surgery, and the conversations. As a physician listening to personal narratives, it is impossible to stay fully outside of the action. I have the privilege to both listen to the story and shape the narrative at the same time.

I hear the accents that curve around words, the nasal New Yorkers, the machine-gun fire voices of Puerto Ricans, the haunting tone of former Russians, and the halting staccato of a Syrian woman with one eye glazed over and the other slanted toward Mecca. I see the plastic rings around the wrists of the teens and the peace signs on their ripped pants. The parchment of skin is filled with visual stories—the tattoos that stretch over multicolored skins, from alabaster white to speckled salmon. There are the ones with gold teeth, little sparkly diamond earrings, and shiny silver crosses over the cleavage. The room is filled with smells—the scent of outdoor workers in their stained shirts, the perfume of an older woman, and the stinky feet of a teenager. The anatomical variations of our patients would make Darwin's head spin. One guy had a head so small it reminded me of a shrunken head dangling from a black thread, so impossibly tiny that you could not imagine it on his eczematous broad shoulders unless you actually saw it as I did. Some have dry skin like parched earth and others have sweaty palms that drain like tiny faucets.

Not to be overly cynical, but I prefer the “give me your tired, your poor” immigrants and those at the edge of the common flow any day of the week, with their patois or street jargon, over the erudite utterances of the well-bred and highly schooled with fleeting problems the size of diatoms who perceive that their ancient Greek self-concepts have been disrupted. Although I care for all ages and demographics, I have a special urge to serve the truly indigent with major impediments to a healthy life, the ones who struggle every day and come to me after years of no insurance or medical care, and who fill my ears with stories I want to capture and woes I want to heal. As a measure of my days, I take down the stories and desires filling the exam rooms and extract each onto paper and into computers. I offer treatments and explanations for maladies and give my best wishes for safe journeys and good health.

ACKNOWLEDGMENTS

Thank you to all my patients whom I have had the privilege of caring for and who often provide me with curious stories and challenging questions that keep me going. My daughter Fionna Lillian suggested the title for the book and I dedicate it to her. Love to my family and friends, fellow physicians, students, office staff, and to all those who add a creative spark to the world and make it a better place in which to live.

Special thanks to Helen Edwards, Sophie Edminson, Jamie George, and Eleanor Moore for their kind assistance in bringing my book to publication.

The patient stories chosen for this book reflect a small but important segment of our overall practice. Names and personal data have been changed to protect patient privacy.

1

THE RED MAN

Charles, an African American man in his 60s, came to see me. He was scratching and moaning, like a bluesman with lots of trouble on his mind.

“I’m losing my skin, Doc,” he said.

He looked like he was sitting inside a covering of distress, a rainstorm of violent skin in an otherwise quiet and clear exam room. He had one of the worst cases of inflammatory skin disease I had ever witnessed, with erythema and scaling that affected nearly his entire cutaneous surface. I could only see a few islands of clear black skin anywhere on him. He was more of a red man than a black man.

“Are you out in the sun a lot?” I asked.

“Yes, sir,” he replied. He told me about his share of hard times and confessed that he had been on the streets and in the elements.

“How long have you had this going on?” I asked.

“I’ve had bad skin for a bunch of years. But lately, it has got much worse. It started with a lot of red areas and then got real scaly. In the last month, it tore me up. Finally, I got on the county insurance and came to see you.”

“We need to take a tissue sample to see why you are losing your skin,” I said.

"Whatever you have to do, Doc, do it," he said. "I'm itchy and in pain."

He did allow us to get a biopsy, and the result, although not conclusive, pointed to psoriasis. Although I would have preferred to have him hospitalized, he refused to go down that route.

Psoriasis alone would not account for the massive mutiny that his skin was undergoing. Clinically, he appeared to have erythroderma, a nasty and difficult problem. The red areas that Charles described were patches that enlarged and coalesced to form extensive areas of erythema, which eventually spread to cover all or most of the skin surface. The profuse scaling generally has its onset 2-6 days after the erythema. Charles's skin was conspicuously bright red, dry, scaly, hot, and indurated. His nails had become thick, dry, and brittle and had lost their luster. Charles's periorbital skin showed inflammation and edema and he exhibited swelling of his feet and ankles.

With most erythroderma sufferers, the basal metabolic rate increases to keep up energy production and untreated patients can slip into an irreversible hypothermia or hyperthermia, with the former resulting in ventricular bradycardia and hypotension. An increased peripheral blood flow may result in high-output cardiac failure.

Occasionally, a patient's skin disease slips past the confinements of expected parameters and erases any sense of anticipated borders. In Charles's case, it was as if a match had struck the parchment of his skin and a wildfire had spread. Over the next couple of weeks, the fire did not respond to the customary preparations to quiet the flames.

Now was the time to get down to business and corner the beast that had brought on these monstrous changes to his skin. In order to facilitate a clear plan of management, I had to look into the underlying reason for his suffering. I ordered blood tests, and no underlying HIV infection, hepatitis, or other major abnormality was detected. He had a history of hypertension but no other diseases or recent changes in medications that could account for his current predicament.

Erythroderma is also known as “Exfoliative dermatitis” and “Red man syndrome.” It is a generalized exfoliative dermatitis that involves 90% or more of the patient’s skin. Since it is a secondary process, determining its cause is essential to facilitate precise management. The most common cause of erythroderma is what Charles had—an exacerbation of an underlying skin disease such as psoriasis. Other diseases such as seborrheic dermatitis, atopic dermatitis, lichen planus, cutaneous T-cell lymphoma, HIV, pityriasis rubra pilaris, contact dermatitis, drug reactions, malnutrition, lupus erythematosus, crusted scabies, and hereditary or idiopathic reasons can bring on erythroderma. Differential diagnosis and treatment in patients with erythroderma may be quite difficult. Infection, drug ingestion, topical application of medicaments, and sun/ultraviolet light exposure can exacerbate the condition. If his disease did not improve with treatment and instead had an insidious, debilitating, progressive course, the recalcitrant nature would warrant an exploration for an underlying malignancy; this is especially true if there is an absence of a history of previous skin disorder and the disease suddenly appears.

The incidence as a function of age is usually variable, and any age group may be affected; however, affected (excluding hereditary disorders/atopic dermatitis) patients are usually older than 45 years, with an average age of onset of 55 years. Psoriasis is the most common cause of erythroderma in adults.

What makes the skin flake out on a man like Charles? The skin is always in a state of renewal. The prodigious fabric that is our skin sheds skin cells at the rate of over 1 million per hour. The pathogenesis of erythroderma is complex but appears to develop secondary to an intricate interaction of cytokines and cellular adhesion molecules that result in a dramatic increase in the epidermal turnover rate and accelerated mitotic rate. This translates into a decrease in the time for cells to mature and travel through the epidermis, and an increased loss of skin.

Treatment is dependent on the cause, so I had to first treat the condition with emollients, antihistamines, topical steroids, topical antibiotics, oral retinoids, and painkillers. I would not consider oral steroids as a first choice, because once the steroids are stopped, the person with underlying psoriasis may often undergo a significant rebound that can be even worse than the initial insult. Each case requires regular monitoring of protein, electrolyte balance, circulatory status, and body temperature. Blood urea, serum electrolyte, and fluid balance should also be monitored, and that is best done in hospital, where he refused to go. With someone like Charles, it was an enormous challenge to manage the condition, because the relentless process puts an extensive strain on an already compromised body, especially given his environmental and economic hardships. He was homeless with skin that looked like it had caught on fire.

Over time, the fire subsided. “My skin is getting better,” Charles said. “And I got me a girlfriend now with an apartment so I can sleep indoors.”

I saw Charles again a month later, and he appeared to have recovered, although he still had the chronic condition of psoriasis to contend with along with his other battles. His management would include skin moisturizing, avoiding scratching and other precipitating factors, applying his topical steroids, and treating the underlying cause and complications. But, for now, he had survived a firestorm on his skin, he had shelter for the night, and he was mostly black with scaly patches of psoriasis, the red having faded into the past.

2

WHY NOW?

A frail alabaster-skinned woman sat on my exam room table, the last patient of the day late on a Friday afternoon. The woman in the corner introduced herself as the patient's sister. The patient, whom I will call Priscilla, slowly opened her blouse. She revealed a growth about the size of a human heart, but outside the central chest instead of inside, as if her heart had somehow flipped outside like an everted starfish.

"How long have you had that?"

"I don't know."

"Approximately."

"More than four years."

I looked at the sister. "Is there a reason you all waited so long to get seen?"

"I can't force her."

Priscilla spoke quietly. "I went to another doctor, a surgeon."

"And?"

"She was a bitch."

"What did she do?"

"She took a swab from it and sent me out. Said she would not remove it."

Priscilla had medical insurance only rarely accepted by any dermatologist—Medicaid—and her choice of providers was severely limited.

We talked and I emphasized that she needed to have this taken off as soon as possible.

We checked her vital signs. Her blood pressure was elevated. Given her non-compliance with the tumor that had grown on her chest, I did not expect her to be taking care of herself, so having high blood pressure fell in line with her approach to health.

“How are you feeling?” I asked.

“I’m scared. I think this may kill me,” she said. “I know I waited too long.”

“I think you’re going to be OK,” I said. “You’re here now so let’s all do the best we can.”

In textbook medicine, I would have sent her away with instructions to lower her blood pressure, giving her all the best options to decrease her risk factors, including perhaps general surgery under anesthesia. But this was far from textbook. I knew that it was highly likely that if she left now, she would never get anything done, and this probable cancer would erode into her chest and take a fatal turn. Even now, I did not know if it had spread and would quickly be her demise.

With some persuasion from her sister, she agreed to have the operation and the proper consent forms were signed. The surgical room was prepared and she was prepped. As she lay down on her back, she had the visage of a frightened child.

I carefully explained to her what I was doing during each step of the surgical journey. “I need you to let me know if you are feeling any pain. I’ll do my best to take this away and get you home as soon as I can.”

I had to reach under the large mass and cut the pedicle that clung to her chest.

“You do what you have to doctor. I trust you.”

As I performed the procedure, one blood-soaked sponge after another was used to mop up the profuse bleeding. Finally, the giant growth gave up its hold on poor Priscilla. If this had been a fish, I would have had to toss it back in the water—it was far above the legal size limit.

Priscilla was shivering and seemed a bit paler. “You have been through a lot already,” I said. “You’re doing great. Hang in there and we’ll get you finished.” I had her sit up and put pressure on her wound so the gravitational flow of blood would not result in more bleeding. In a few minutes, the flow of blood subsided, and I began the onerous chore of sewing up the defect where the tumor had grown and spread.

After the surgery, she seemed to have a bit more color and I was hopeful that she would have a good recovery.

“God sent me to you,” Priscilla said.

“We have a way to go,” I said. “We have to see what the biopsy shows. You need an MRI (magnetic resonance imaging) scan and your blood pressure needs to be controlled. You may need to see a general surgeon or plastic surgeon.”

“I need something for the pain.”

I gave her a pain med, an oral and topical antibiotic, and strict instructions to her and her sister to head to the ER if she started having bleeding or any problems.

When I checked her before she left, she appeared as if her burden had lightened; she had shifted from a scared child to someone who at least seemed to be able to stand a little taller.

The biopsy was reported as a 314 gram, 12.5 cm x 8 cm basal cell tumor, certainly the biggest one I had ever removed. I saw her again a couple of weeks later, and her wound had started to heal. When I asked her about getting her blood pressure checked or an MRI, she replied, “I haven’t had time." In Priscilla's case, she had delayed her entry into the world of treatment for many reasons. She had perceived her insurance as being inadequate and had assumed she would not get help. In addition, she clearly had a fairly robust psychiatric overlay to her problems. When I took a more nuanced look at her life, including her unorthodox dependency on a rather reluctant and taciturn sister, I sensed that her emotional state contributed heavily to the mix.

Although the focus here is on Priscilla, a patient with skin cancer, a bigger problem unfolds with deeper observation—why now? Why do certain people wait so long to come to see us when they must be aware that they have a problem that is continuing to get worse? And then comes the great question of everyday life—what do we do next?

Here are some other examples. A man came to see me who complained about a "growth on my arm." When he was in the exam room, and my medical assistant gently pulled off his ragged home-rigged dressing, a profusely bleeding half-dollar-sized skin cancer made its appearance. I mentioned the option to surgically remove it right then, and the patient agreed. But underneath the story of this nasty growth was a whole history of events that had occurred prior, in particular an extreme lack of money. I remember a woman who had waited over ten years to see a physician and when she showed up at my office, she had a basal cell cancer that covered almost her entire right cheek. "I didn't have any money and just got insurance last month," she said.

I saw an eight-year-old girl in my office with severe atopic dermatitis. She had been out of school for three weeks and the mother had been off work due to lack of sleep and trying to care for her child during much of the three weeks. Why did the mother wait for three weeks?

And the horrendous cases often gain purchase in my mind, such as the 41-year-old man with AIDS who came to see me because he felt a bump on his buttocks and never did anything about it. When I examined him, he had a huge and irregular growth that I removed—a 12 mm deep melanoma—that had already spread internally, and despite every effort, eventually claimed his life. Or the 50-year-old man who looked at least 70 years old and had smoked two packs a day since his teenage years. He showed me a nasty growth that everted his lower lip. During his examination, I noticed a swelling on his neck, and later found out that the squamous cell cancer had spread to the cervical lymph nodes and beyond.

A 48-year-old man rolled up his pants on his right leg and showed me a huge, round growth that looked like it was about to burst. “I’ve had it 20 years,” he said. “But only in the last five years has it started to give me any pain.” The guy was really strange, with a sort of child’s pride in having kept this hidden for so long. If he had told me he had swallowed a grapefruit and it somehow got lodged in his upper thigh, I may have believed that he thought it was true.

The man covered his face with a drape while I worked on him, but his wife hovered nearby and was engrossed in the whole event. I opened the lesion with my scalpel blade and it turned out to be a large, irritated cyst which gushed ounces of a cheesy substance. The key was removing the shell of the cyst so it would not recur, and this took some time. His wife and I discussed the task at hand and he moaned with a squeamish cry every few moments.

“Any pain?” I asked.

“No,” he said. “I just don’t want to look.”

Even though I used an absorbable foam to fill the large deficit, an indentation remained when I finished the task of sewing him up. Over time, the normal contour of the leg would hopefully come back.

A 32-year-old Hispanic woman came to see me, accompanied by her parents. She asked about getting a growth taken off her lip. I asked how long it had been there, and she replied, “ten years.” I performed a history and physical and noted a raised red bump on her left upper lip, appearing as an irregular hematoma. The patient, who I will call Graciella, was mildly mentally retarded, and the family had moved to Tampa from a tough area in New York City about eight years earlier. She remembered getting some rocks thrown at her one day by rowdy neighborhood kids. “I think one of them hit my lip,” she said, pointing to the raised bump.

“Why after all this time did you want this off?” I asked.

“I don’t know,” she said. “I’m just tired of looking at it.”

I had her sign a consent form after reviewing the procedure with her and her parents. The mother appeared quite anxious, and the parents left the room prior to the procedure.

I numbed up the area, and Graciella seemed to handle everything without much distress. I used a scalpel to open the lesion and explored it with dissecting scissors. Within a short time, I probed and found a solid object that resembled a small hematoma. Using a pickup, I was ready to pluck it into the biopsy bottle when I noticed something shiny. While my nurse put pressure on the wound, I pressed on the object with my fingertips. The bloody covering slipped off, revealing a tiny copper ball. It was a BB! I dropped it in the bottle, and it landed with a little “kerplunk.”

“I don’t think you got hit by a rock ten years ago,” I said. “You got shot with a BB gun.”

"A BB?" she exclaimed. "Oh, my God! Momma, Pappa, come here!"

I told my nurse to have the parents come in. The mother was a bit hysterical when she heard the news.

"That's a first," I said.

I just shook my head in amazement. After ten years, the mystery had been solved.

Recently, I had a 39-year-old Cuban man come in to see me because of what he thought was a cancer on his abdomen.

"It has been there for about 16 years but now it is bothering me and it has gotten bigger."

I asked him about trauma or other history but he denied anything that may have contributed to the growth. I examined the raised area near his belly button, which appeared to be more cystic than solid, and told him I would need to take a biopsy. After he agreed, the area was prepped and I opened it up. Inside, I found a two-inch piece of suture wrapped around and twisted.

"Oh yeah," he said. "I had surgery there 16 years ago."

The dissolvable suture had never dissolved and had brought on a skin irritation that had raised itself up and made itself known.

Another patient, a woman in her early 50s, came to see me for a raised lesion in her right axilla.

"The doctor told me it was probably an abscess," she said. "And he gave me antibiotics. But it has not gone away."

With further examination and evaluation, I discovered she had late-stage breast cancer. Despite aggressive surgery and chemotherapy at a first-class treatment center, the cancer had metastasized and she was dead within a year of her visit to me.

Here were two patients—one with a lesion imagined as cancerous that was benign, and one that had been initially diagnosed as benign and carried a virulent cancer.

How has all this changed me? I have always known that health issues have their own time and trajectory. Diseases do not read the textbooks and fit into neat presentations, nor do they read the insurance plans and grow in a uniform way to be treated by all insurances. We waste a tremendous amount of each day trying to satisfy the requirements of prior authorizations and other stubborn regulations while the disease proceeds without regulatory boundaries. All this has taught me that many people clearly fear illness and death and will find all kinds of inventive ways to not face their fears. I am now even more sensitive to the patients' fears and find my own inventive ways to provide comfort when I can. I have also developed a greater understanding of how a person's social, ethnic, and financial status can completely undermine even the best efforts at good health. All health care providers can recount these same kinds of stories of struggle. As physicians, we are often required to peel away other possibilities to get to the source of a patient's medical problem and repair what has been disturbed or broken. And with each problem comes a whole underlying story that has preceded the person's appointment, a pentimento of starts and stops that finally leads to being seen.

And the two main players—politics and money—will most likely always play the roles of central characters in our future drama as regards health and happiness. In the United States, we are not even able to get to grips with the moral imperative to establish a single-payer system. Given the huge discrepancies in health care access, much of what I and other practitioners deal with on a day-to-day basis is so incredibly wasteful, including days filled with burdensome prior authorizations, prescription eligibilities, and delayed and denied treatments. Insurance and pharmaceutical companies may be practicing legalized gambling or perhaps “legalized” stealing, but we clearly should prefer high-quality health care instead of insurance companies and bank buildings being the highest peaks in the vista of urban America. Most medical practitioners have taken on the unwilling role of medical advisors instead of decision-makers, given the pedantic and controlling interference of insurance and pharmaceutical mavens in not allowing our patients to have the best possible care.

Although it is impossible to delineate the reason for delaying or avoiding treatment in each case, over time it appears that certain characteristics contribute to the overall outcome and often delay appropriate and timely care. Many consistent reasons for hesitation can be noted—fear, frustration, denial, feeling vulnerable, lack of trust, symptoms that did not interfere enough with daily life, misdiagnosis, a lack of funds, poor compliance, a perceived or real paucity of available and timely care, limited transportation—all can contribute to someone not seeking a remedy.

Events such as skin cancer screenings and the incorporation of skin cancer checks in primary and dermatology care help detection. New techniques such as non-invasive applied test strips to detect various skin disorders may help to decrease the need for skin biopsies, allowing for an easier and more comfortable diagnosis, especially for the estimated 10% of people who suffer from trypanophobia, a fear of needles.

A Priscilla type exists in every medical practice. We need to be aware that, for whatever reason, sometimes *now* is the right time to care for her.

3

FULL OF BUMPS

“I’ve got like bumps all over me,” the 55-year-old Juan said. “I’ve had these for most of my life, but now I’m getting more and they are getting bigger and I think they are going to cover all my skin.”

I looked him over. A few of the growths on his forehead and scalp were somewhat protuberant and could easily be seen. On further examination, I saw many scattered growths on his abdomen, arms, and scalp.

Juan was obviously worried and concerned about these growths, not only because he mentioned them, but because this was the first time he had gone to a dermatologist for help. It was hard to imagine that he had waited so long to seek help. He didn’t seem to have a clue why he had these lesions and why he was getting more. He said that in the last eight years since moving from his home in Colombia to the United States, he kept getting more lesions. Within the past few months, they had shown an accelerated increase in size and frequency of new lesion appearance and had also started appearing more on his scalp.

“I’ve also got a dark spot on me that looks like it’s getting bigger,” he said, pointing to his abdomen.

He stated that he had no complaints as regards the quality of his life caused by the lesions. He described no itching or pain with the lesions, and never any bleeding or drainage.

Juan stated that several of his family members suffered from similar fleshy lesions and colored spots but that no one had been diagnosed as a result of any recognizable medical assessment because, until recently, they had lived in an area without access to in-depth medical technology. He stated that he was otherwise healthy and had not had any significant medical conditions.

“Did anyone ever say anything to you about these before?” I asked.

“One of my grandkids asked me why I was growing a horn on my head.”

On physical examination, the patient appeared to be a moderately healthy male for his stated age. The ovoid lesions were flat and between 1 mm and 3 cm in diameter and up to 8 mm in height, scattered diffusely throughout his trunk, scalp, and upper arms bilaterally, totaling approximately 50 in number. The lesions were fleshy in color but slightly reddish in hue with a glassy appearance. They were freely moveable in the skin and were non-blanching; also, they were semi-compressible.

Juan did not complain of pain or tenderness with palpation of the lesions. His hair growth appeared normal and distribution was only absent where the scalp lesions arose. His skin was tan in color and uniform throughout, except for a large rhomboid-shaped café au lait spot visible on the abdomen. Café au lait spots or café au lait macules are pigmented birthmarks, named after the French for “milky coffee,” a reference to their light-brown color.

Two of the raised lesions on the scalp were biopsied. A few days later, the report was returned as neurofibromas. Although these can occur in other diseases, the widespread nature of Juan’s lesions was characteristic of neurofibromatosis (NF), a neurocutaneous disorder that is separated into two distinct entities. NF type 1, also called von Recklinghausen’s disease