A Divine Diagnosis: The Christian Church's Discovery of Gardner-Diamond Syndrome

Katherine McDonald, BScH[1]; Asfandyar Mufti, BMSc[1]; Robert Jackson, MD, FRCPC[2]

[1]Department of Medicine, University of Ottawa
[2]Division of Dermatology, University of Ottawa

Abstract

The Roman Catholic community was the first to observe and describe what later became known as Gardner-Diamond syndrome (GDS), otherwise known as psychogenic purpura. They attributed the dermatological manifestations of GDS to the 'Holy Stigmata' or the 'Five Holy Wounds,' physical and sensory signs linked to Jesus Christ's crucifixion. Between 1927-1989, medical curiosity resulted in the discovery of a link between these stigmata and a recognizable hematologic condition, which later became known as GDS. GDS is an autoimmune hematological disease associated with emotional and physical stress that involves painful ecchymosis. This stress is often related to psychiatric conditions such as depression, anxiety, and obsessive-compulsive disorder. GDS, the medical condition, must not be confused with mimicking presentations, such as variants of fictional disorder in which so-called 'fraudulent stigmatists' self-inflict similarly-presenting wounds. As such, clinicians must be able to distinguish patients with true GDS from religious individuals who have self-mutilated to mimic the 'Five Holy Wounds'. Reaching this diagnosis of true GDS might require a multidisciplinary approach between psychiatry, hematology and dermatology.

The History of Gardner-Diamond Syndrome

While history often demonstrates a volatile relationship between religion and medicine, their coexistence has sometimes resulted in positive outcomes for society. For instance, the Roman Catholic community was the first to observe and describe the Holy Stigmata, which the medical community later reinterpreted as the rare but fascinating disease, Gardner-Diamond syndrome (GDS).

In the Christian tradition, the term stigmata (singular stigma) refers to the bodily wounds (physical stigmata) or sensations of pain (invisible stigmata) that correspond with the crucifixion marks on Jesus Christ's body.[1] These are the traditional 'Five Holy Wounds.'

Each stigma has a specific biblical origin. The involvement of the hands and feet are associated with the nails that secured Jesus Christ to the cross. The involvement of the unilateral abdomen is related to the lancet that pierced Him. Finally, the involvement of the scalp is related to His crown of thorns.[1]

Stigmata were first referenced centuries ago, by the Church. In the Holy Bible (Galatians 6:17), Saint Paul refers to stigmata when he states, "…for I bear on my body the marks of Jesus."[2] The Christian faith dictates that when these markings appear on an individual's body, it is a holy tribute to the affected individual, marking their divinity and deeper connection to Jesus Christ.

Holy Stigmata were first documented as appearing on St. Francis of Assisi (1181-1226) but have also appeared on other notable Roman Catholics, including St. Catherine of Sienna (1347-1380) and St. Padre Pio (1887-1968).[1]

Unfortunately, the divine status conferred to affected individuals by the Church has sometimes also encouraged self-infliction of wounds among religious followers. These self-inflicted wounds are not legitimate cases of GDS, but rather, are examples of factitious disorder, wherein individuals feign Holy Stigmata as an attempt to gain recognition from the Church.[3] Fraudulent stigmatists include historical individuals such as Magdalena de la Cruz (1487-1560), as well as devout modern day religious followers who replicate the Holy Stigmata as a symbol of Jesus Christ's courage within themselves.[4] The differentiation between GDS and imitating wounds with a religious connotation is critical, as the former is a diagnosable medical condition and the latter is a cultural action. The latter is not medical unless it is intended as self-harm.

In 1927, the medical community investigated the Holy Stigmata for the first time. German psychiatrist Rudolf Schindler documented sixteen patients with the purpuritic lesions of the holy stigmata and discovered that they could be healed in due time by hypnosis.[5,6] The sixteen patients gradually recovered from the purpura rather than experiencing the typical worsening of skin lesions due to their distress. Since the goal of the hypnosis was to reduce psychological stress, it was hypothesized that the purpuritic lesions were a result of stress, emotional trauma, or psychiatric illness. This idea garnered further support when in the following year two more cases of GDS (known as hysterical bleeding or purpura at the time)

emerged, both associated with what was then described as a 'hysterical personality' and delusions.[5-8] Similar to the previous example, these two cases involved patients in extreme distress with a possible underlying psychiatric disorder.

The tangibility of medical evidence further advanced when in 1955, hematologists Dr. Frank Gardner and Dr. Louis Diamond identified four cases of Caucasian women presenting similarly to those with Holy Stigmata.[9] It was discovered that these women developed sudden erythema and edema followed by painful ecchymosis secondary to auto-sensitization to a component of their own blood.[7,9] This observation prompted the term 'painful bruising syndrome' as well as the official medical term, GDS.

The long list of synonymous medical terminology did not end with the terms GDS, painful bruising syndrome, hysterical bleeding, and auto-erythrocyte sensitization syndrome. In 1989, GDS gained the attention of notable hematologist Oscar Ratnoff, the founder of the coagulation cascade. He coined the term, 'psychogenic purpura' to more accurately describe the psychiatrically related auto-erythrocyte sensitization syndrome.[3]

The skin lesions in GDS are not fixed to the locations of the Stigmata of Crucifixion, but they can sometimes demonstrate a resemblance due to the typical ecchymotic pattern on the face, hands and feet. After a period of pruritus and burning sensation, edematous lesions present in the extremities and advance to ecchymosis within twenty-four hours.[5] A minimum of one week is required for the lesions to heal, but they often persist and new bruises develop. The patients may have associated symptoms, including but not limited to fever, arthralgia, myalgia, headache, dizziness, gastrointestinal symptoms, hematuria, subconjunctval hemorrhage, vaginismus, and menorrhagia.[5]

GDS is also linked to some medically recognizable hematologic conditions, such as thrombocytosis.[5] Further research has demonstrated that the auto-sensitization is specific to phosphatidylserine, a component of the erythrocyte cell membrane, and that the auto-sensitization seems to increase during times of stress on the body, in line with Schindler's original stress-related hypothesis.[3]

The religious explanation of psychogenic purpura was publically accepted for centuries, but it did not subdue the curiosity of the medical community and physicians. Although arguments for 'genuine' Holy Stigmata still exist, the Christian theory behind the stigmata has failed to withstand rigorous analysis within the medical realm.[1,5] Devout Catholics may continue to support the religious theory because there is no adequate medical explanation for the repeated involvement of the five holy body regions.

The history of this rare cutaneous condition highlights the importance of the Church's keen observation of the Holy Stigmata as well as the scientific community's recognition of the medical merit behind the religious claim. Members of the Church recognized a cutaneous ecchymotic pattern and provided a religious explanation. Physicians saw the validity in this pattern recognition and felt it was worth medically investigating. Ultimately, it was the Roman Catholics who first identified the signs of GDS. Despite proposing an incorrect mechanism of pathogenesis, the Church accurately observed and documented a dermatological manifestation of a systemic disease centuries before the medical community.

The Current Clinical Application of Knowledge on Gardner-Diamond Syndrome

GDS is a rare condition with an interesting etiology and history. If encountered, this condition would more likely present to family medicine, emergency medicine, dermatology, hematology, or psychiatry. The syndrome is most often seen in Caucasian women, particularly among individuals with a history of psychiatric diagnoses such as depression, anxiety or obsessive compulsive disorder (OCD).[10] The etiology of GDS remains unclear, but it has been suggested that severe stress or emotional trauma, combined with a hematologic abnormality, is the inciting cause. It is uncertain whether GDS patients require a concurrent psychiatric diagnosis, or if a psychiatric disorder merely predisposes the patients to more profound distress.

Clinicians should be able to differentiate this auto-erythrocyte sensitization syndrome from self-inflicted wounds in order to determine appropriate management. This can be done based on the patient's history. For instance, GDS is usually preceded by a prodrome of pain and warmth at the sites of ecchymosis as well as by headaches, nausea and occasionally, by vomiting.[10] A minor trauma or surgery that results in both bleeding and psychological distress can sometimes trigger GDS in a predisposed individual.[10]

Although commonly coinciding with the location of the 'Five Holy Wounds,' GDS-associated bruising may be located anywhere on the body. Therefore, when considering the differential diagnosis, the inciting factors (e.g. stress/psychiatric disorder, erythema and burning sensation prior to bruising) and the patient profile (e.g. Caucasian female) might be more suggestive of GDS than the exact location of the ecchymosis. However, in the majority of individuals with GDS, bruising typically starts in the periphery (hands and feet).[5] Ecchymosis is often extensive, severely painful and debilitating to the patient, and lasts for an indefinite period of time. Unfortunately, there are no laboratory investigations that can definitively diagnose GDS; it is a diagnosis of exclusion.[10]

The differential for GDS primarily includes other bleeding disorders. If GDS is suspected, laboratory investigations should include prothrombin time (PT), activated partial thromboplastin time (aPTT), and a complete blood count (CBC) with platelet count, all of which should be within the normal ranges.[11] Additional coagulopathy testing may also be ordered depending on the individual case and the clinical suspicion. If necessary, a skin biopsy may be performed. In GDS, the skin biopsy will show extravasated red blood cells and no evidence of vasculitis.[7,9]

The management of GDS highlights the strong connection between the psyche, the immune system and the skin. This is not a new discovery, as the literature already demonstrates that 30% of patients with a skin disease have a concurrent psychiatric diagnosis.[12,13] Although the etiology of the disease remains uncertain, it has been proposed that in an individual who is predisposed to auto-erythrocyte sensitization, emotional or physical stress might trigger and/or worsen GDS, particularly when an underlying psychiatric diagnosis is present.[7,10] Support for this theory has been demonstrated in clinical treatment of GDS. For example, psychotherapy has been shown to improve GDS-related ecchymosis, most likely due to a reduction in patient stress.[5] In addition, selective serotonin reuptake inhibitors (SSRI) and anxiolytics have been shown to improve the GDS patient's mood and to reduce the healing time.[10,14]

Clinicians and trainees should be educated in the emerging fields of psychodermatology and psychoimmunology in order to succeed in the diagnosis and management of rare and challenging conditions such as GDS. Self-mutilation must always be considered in the differential diagnosis. A detailed history, focused attention on the patient, and blood work with additional coagulation tests are paramount for the diagnosis of this perplexing medical condition.

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