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# The Holster Sign: An Overlooked Cutaneous Finding in the Diagnosis of Dermatomyositis

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## Abstract

**Objective:** Characteristic skin findings in dermatomyositis (DM) include Gottron's papules (Gp), heliotrope rash (He) and shawl or V (S/V) sign. Noted in dermatology publications is a violaceous rash over the lateral hip, called the "Holster sign." Our objective was to determine the incidence of the Holster sign (HS) in comparison to other DM cutaneous signs and to raise rheumatologists' awareness of the HS.

**Methods:** Patients with ICD-9 codes for DM, PM, MCTD and SS were identified. Patients received a survey that included pictures of He, Gp, S/V and HS and were asked to check if they "had any of the following skin findings?" Charts were reviewed to confirm the diagnosis and providers' description of skin findings. We calculated the incidence rate, sensitivity and specificity of each skin sign for DM.

**Results:** From 117 surveys, positive HS self-reporting was 22/28 in DM, 2/13 in PM, 0/21 in MCTD, and 6/55 in SS. Hs positive reporting of 78.5% in DM, was comparable to S/V (75%), Gp (85%), and He (71%). When compared to other signs, HS reporting was lowest in PM, MCTD and SS patients. Of all the skin signs, the HS had the highest specificity for DM (91%, range 75-91% for other signs) and comparable sensitivity (79%) to other signs (71-85%).

**Conclusion:** This survey indicates that the HS has a comparable incidence to the more established and recognized cutaneous findings in DM. Based on these findings, we recommend looking for the HS sign in all patients with suspected DM.

**Keywords:** Dermatomyositis; Holster sign; Diagnosis

## Introduction

The idiopathic inflammatory myopathies include polymyositis (PM), dermatomyositis (DM) amyopathic dermatomyositis (ADM), and inclusion body myositis [1] In those patients who have DM and ADM, the cutaneous findings are characteristic and are considered to be very important in the early recognition and diagnosis of the disorders [2,3]. Patients with cutaneous manifestations can have a violaceous macular or popular rash over many different body areas. However, specific skin areas are characteristically involved, resulting in the description of "hallmark" cutaneous signs. These include: heliotrope rash (violaceous erythema of the eyelids or periorbital skin), Gottron's papules (violaceous papules overlying the dorsal and lateral aspects of the MCP and PIP joints), Shawl and V sign (macular violaceous erythema over the nape of the neck and shoulders and the V of the neck). Cutaneous signs have been used for both the diagnosis of the inflammatory myopathies, and also to measure disease activity [4-6].

The Holster sign (HS) was noted in a 2002 review of dermatomyositis by Richard Sontheimer [7]. He described it as a "macular, violaceous erythema, often displaying reticulated or livedo array over the lateral aspects of the hips and upper thighs." The erythema is usually centrally located and below the greater trochanter. It is frequently bilateral and symmetrical. Since its shape and location is similar to where a leather holster would lie, it has been called the Holster sign.

Reference to the HS has been included in several publications and anthologies [2,3,8], however it is rarely mentioned in American texts. Our academic rheumatology group was introduced to the HS by a dermatology colleague (KS). In discussions with rheumatologists at other US centers we found that many were not aware of the HS.

We performed a retrospective self-reporting survey in our patients with DM, polymyositis (PM), mixed connective tissue disease (MCTD), and scleroderma (SS), to determine the sensitivity and specificity of the HS in comparison to the other classic cutaneous signs in patients with DM.

## Patients and Methods

### Patient selection

Patients with ICD-9 codes for dermatomyositis, polymyositis, mixed connective tissue disease or scleroderma who had been seen between January 2003 and December 2008 by rheumatology or dermatology providers at Fletcher Allen Health Care were mailed a survey packet. The packet included pictures of a heliotrope rash, shawl sign, holster sign and Gottron's sign (Figure 1).

Patients were asked to check yes or no above each picture in response to the question "Have you had any of the following skin

findings?" Subjects who completed the survey and a signed informed consent for chart review were entered into the study. The study was approved by the Institutional Review Board of the University Of Vermont College Of Medicine.

### Chart review

The medical records of the patients who completed the questionnaire and the informed consent were reviewed. Providers notes, diagnostic and laboratory studies were used to confirm the ICD-9 diagnosis. Peters/Bohan criteria [9,10] were used to confirm the diagnosis for the DM/PM patients. Presentation, clinical findings and positive anti-RNP antibody were used to confirm the diagnosis of the MCTD patients. Clinical features, including sclerodactyly and Raynaud's were used to confirm the diagnosis of the scleroderma patients [11].

We reviewed all providers' records (rheumatology and dermatology) of the DM patients to document the patient's cutaneous exam findings, with attention to the presence or absence of the hallmark cutaneous signs.

### Statistics

The incidence rate for each skin sign was calculated for each diagnosis, and the sensitivity and specificity of each sign for dermatomyositis was calculated.

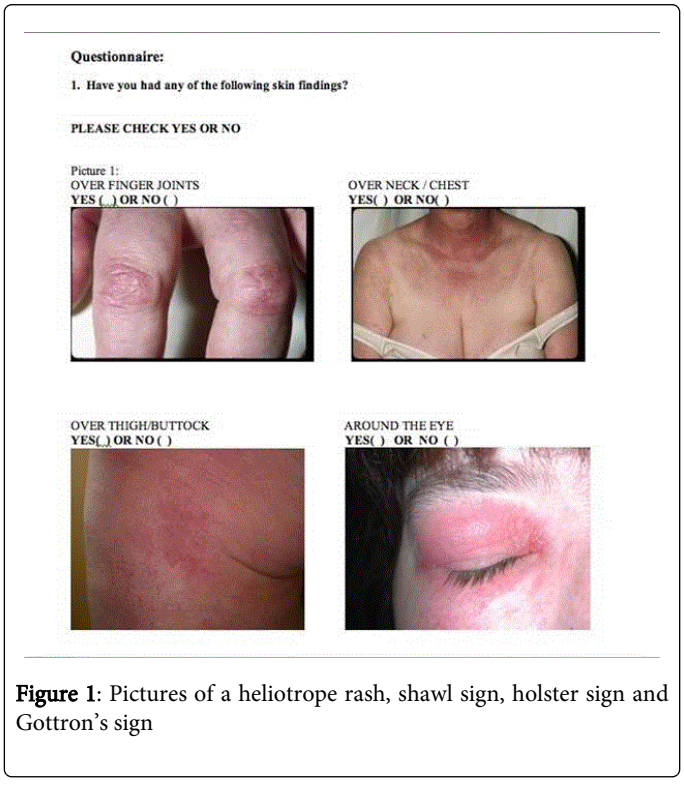


**Figure 1**: Pictures of a heliotrope rash, shawl sign, holster sign and Gottron's sign

## Results

A total of 117 surveys and informed consents were returned from the 315 surveys that were sent to the patients in the 4 diagnostic groups. Chart reviews of the 117 patients who returned the survey confirmed the original diagnoses. The return rates were: DM 28/46 (61%), PM 12/38 (34%), SS 55/139 (39%), and MCTD 26/93 (24%). The higher return rate from the DM patients is likely due to the fact that more DM patients recognized the skin findings shown on the survey sheet, and were more motivated to return the survey.

Table 1 shows the rates of positive self-reporting for the 4 dermatologic signs in the 4 patient groups. Within the DM group, self-reporting of the selected dermatologic findings revealed that 22/28 (78.5%) patients had the holster sign.

The incidence of the other cutaneous signs ranged from 67-86%. By comparison, the incidence of the HS in the three control diagnostic groups ranged from 0-15%. We performed chart reviews on all PM, SS and MCTD patients who reported positive responses to the survey questions and did not find any description of the cutaneous findings in the providers' notes.

The sensitivity and specificity of each skin finding for the diagnosis of DM within the cohort was calculated (Table 1). The HS had the highest specificity of the skin findings at 91%, with a range of specificity from 75%-91% for the other signs. Sensitivity of the Holster sign was 79%, with a range of 71%-85% for the other signs.

We determined the rate at which rheumatologists and dermatologists recognized and noted the presence of the 4 cutaneous signs in their records. Identification of the HS by dermatologists was 58% as compared to 35% by rheumatologists. In comparison, rheumatologists' recordings of the shawl/V, Gottron's and heliotrope signs were 56%, 74% and 60%, while dermatologists' recordings were 47%, 63%, and 42% respectively.

## Discussion

The results support the validity of the holster sign as another specific cutaneous finding in patients with dermatomyositis. However, in clinical practice, Gottron's sign, heliotrope, and shawl/V sign remain the pathognomonic "hallmark" signs. One of the limitations of the study is the relatively small number of patients included. However, both the patient survey and chart review demonstrate that there is comparable incidence of the holster sign to the more well established cutaneous findings associated with dermatomyositis. In addition, the holster sign appears to be highly specific for dermatomyositis.

A limitation of a questionnaire-based study, is the evident bias toward collaboration with the asked patient. The significant difference in the survey return rate between DM patients (61%) and control groups (24%) can also have an effect upon the comparisons of sensitivity and specificity.

The patient survey and chart review demonstrate that there is a comparable incidence of the holster sign to the more well established cutaneous findings associated with dermatomyositis. In addition, the holster sign appears to be highly specific for dermatomyositis. The rate at which dermatologists recorded the holster sign was higher than rheumatologists. The patient survey and chart review demonstrate that there is a comparable incidence of the holster sign to the more well established cutaneous findings associated with dermatomyositis. In addition, the holster sign appears to be highly specific for dermatomyositis. The rate at which dermatologists recorded the holster sign was higher than rheumatologists.

| Signs: | Self-Reporting of signs for each diagnosis | | | | Sensitivity for DM | Specificity for DM |
|---|---|---|---|---|---|---|
| | Dematomyositis | Polymyositis | Scleroderma | MCTD | | |
| **Holster** | 22/28<br>79% | 2/13<br>15% | 6/55<br>11% | 0/21<br>0% | 79% | 91% |
| **Shawl/V** | 21/28<br>75% | 2/13<br>15% | 12/55<br>22% | 7/21<br>33% | 75% | 76% |
| **Gottron's** | 24/28<br>86% | 3/13<br>23% | 13/55<br>23% | 7/21<br>33% | 85% | 75% |
| **Heliotrope** | 20/28<br>60% | 3/13<br>23% | 7/55<br>13% | 1/21<br>4.7% | 71% | 87% |

**Table 1**: Shows the rates of positive self-reporting for the 4 dermatologic signs in the 4 patient groups

However there was comparable recognition by dermatologists and rheumatologists of the three other hallmark signs. The traditional cutaneous signs are easily visible on the face, neck and hands. Since dermatologists are more likely to perform a full skin exam, they are more likely to observe the holster sign. It is important that all patients with suspected dermatomyositis be examined in either a gown or shorts and T shirt to allow for a full skin exam.

A comparison of dermatomyositis patients seen by dermatology and rheumatology practices at a tertiary medical center showed, as expected, more patients with amyopathic dermatomyositis (ADM) in the dermatology practice [12]. Five patients in our study presented with ADM, and all were dermatology patients. All of these patients had hallmark cutaneous signs, including 4 with the HS.

While the cutaneous hallmark signs are well-recognized and appreciated, the frequency of these signs in classic dermatomyositis or amyopathic dermatomyositis has not been formally evaluated. We do not know if DM patients typically present with 1, 2 or more hallmark cutaneous signs. In our population, it is interesting that 46% of DM patients reported the presence of all 4 signs on the questionnaire. In addition the HS was always self-reported with 2 other signs,

In summary, the HS appears to be highly specific for dermatomyositis, and its presence is comparable to the other hallmark cutaneous signs. Based on these findings, we recommend looking for the holster sign when evaluating all patients suspected of having a myopathy.

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