Validity of dermatomyositis case identification using diagnostic codes

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Background

Dermatomyositis (DM) is an uncommon idiopathic inflammatory myopathy (IIM) resulting in characteristic rashes and muscle weakness. Typical cutaneous manifestations include heliotrope rashes (periorbital erythema), Gottron's papules, and Gottron's sign (erythema of knuckles, elbows, and knees). DM is strongly associated with malignancy and various autoimmune conditions.

Objective

The purpose of this study was to assess the validity of specialist-specific diagnostic coding to identify patients with DM in an outpatient clinical database.

Methods

Data source: Outpatient electronic medical record database at Northwell Health

Key inclusion criteria: DM diagnosis applied at a rheumatology or dermatology encounter (ICD-9 710.3; ICD-10 M33.10, M33.11, M33.12, M33.13, M33.19, M33.90, M33.91, M33.92, M33.93, M33.99)

Data collection: Review of physicians' clinical notes, laboratory, and imaging results for signs and symptoms

Case confirmation: *Primary* = clinical documentation of DM diagnosis by treating physician; *Secondary* = satisfaction of 2017 European Alliance of Associations for Rheumatology/American College of Rheumatology (EULAR/ACR) criteria for "probable" or "definite" IIM and relevant cutaneous findings.

Results

Table 1. Positive predictive values (PPV) of ICD-code case definitions for dermatomyositis from electronic medical records

Case Definition (# of codes and provider specialty)	Reference Definition: Diagnosed by treating physician		Reference Definition: 2017 EULAR/ACR Criteria	
	PPV (n/N)	95% CI	PPV (n/N)	Positive Predictive Value (95% CI)
≥ 1 dermatology or 1 rheumatology	82.1% (128/156)	77.3%-86.8%	44.9% (70/156)	38.8%-51.0%
≥ 1 dermatology	93.2% (41/44)	82.0%-98.3%	52.3% (23/44)	38.3%-66.2%
≥ 1 rheumatology	82.0% (123/150)	77.1%-86.9%	46.7% (70/150)	40.4%-53.0%
≥ 2 dermatology or 2 rheumatology	85.7% (108/126)	80.7%-90.8%	49.2% (62/126)	42.0%-56.4%
≥ 2 dermatology	96.4% (27/28)	82.2%-99.8%	57.1% (16/28)	39.4%-74.8%
≥ 2 rheumatology	85.8%(103/120)	80.6%-91.1%	50.8% (61/120)	43.3%-58.3%

Discussion

- We observed PPVs ranging from 82%-96% for ICD-9/-10 coding of DM by rheumatologists or dermatologists when compared against the treating physician's diagnosis.
- Definitions requiring at least 2 codes marginally improved the PPV, and coding by dermatology resulted in PPVs approximately 10 percentage points higher compared to rheumatology.[1,2]
- EULAR/ACR criteria represent classification criteria designed to identify a homogeneous cohort of patients with common shared features for clinical research. They may be less suitable than treating physician's diagnosis for the validation of administrative coding to identify real-world DM cohorts.[3,4]

Conclusions

- Application of at least 1 or 2 ambulatory diagnosis codes by dermatology or rheumatology, or by each separately, may be used to identify an accurate case cohort of patients with DM for analyses within clinical databases.
- A validated algorithm to identify a DM cohort within a large clinical database will support analyses that may better characterize the relationship between dermatomyositis and malignancy, interstitial lung disease as well as other comorbid conditions.

References

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Disclosures

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