Mass General Brigham

Identification of Drug Rash with Eosinophilia and Systemic Symptoms (DRESS) Syndrome in Electronic Health Records

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Background

Drug reaction with eosinophilia and systemic symptoms (DRESS) syndrome is a rare and severe drug hypersensitivity reaction associated with substantial morbidity and mortality[1]

DRESS syndrome typically develops from 2 to 8 weeks after the exposure to the culprit medications including antibiotics, anticonvulsants, allopurinol, and antiretrovirals[2]

Typical clinical manifestation of DRESS syndrome include rash, facial edema, lymphadenopathy, multiple visceral organ involvement of eosinophilia and lymphadenopathy with systemic symptoms like fever, rigors, and hypertension[3]

Due to the low incidence rate, ranging from 1 in 1000 to 1 in 10,000 drug exposures, a small study sample size was the most common limitation encountered by studies of DRESS syndrome in the US, limiting our understanding of this rare and complex disease, its risk factors, and long-term sequelae[4]

Due to its rarity and inconsistent coding, large number of cases of DRESS syndrome are not coded in the diagnosis for the patients. No single electronic health records (EHR) component can be enough to find all the cases

Study Aims

Studying how DRESS syndrome has been documented in the EHR will enable more efficient case identification that supports patient enrollment for epidemiological and pharmacogenetic studies and drug safety

Methods

We applied rule-based approaches to search for DRESS syndrome cases from Mass General Brigham (MGB)'s enterprise data warehouse (EDW) between January 1, 1980 and June 19, 2020 via multiple Epic EHR components, including problem list, medical history, allergy list and billing/encounters

For billing or encounter data, we used ICD-9 codes 693.0, E947.9, 288.3 and ICD-10 codes D72.12, T50.905A to search for DRESS cases, while for problem list and medical history both ICD codes and keyword-based search. For the allergy list, we used keyword-search approach as well as reaction codes

Each case was screened by a researcher to rule out negated cases and cases with insufficient records

The remaining cases were chart reviewed by trained researchers by performing a detailed REDCap data entry form to confirm whether the cases indeed have DRESS syndrome. Patients with RegiScore ≥4 were classified as DRESS syndrome cases

We assessed the prevalence, positive predictive value (PPV), and sensitivity of each EHR component for DRESS case identification

We evaluated how many true cases were found from the problem list, billing/encounters, and medical history but not documented in the allergy list

Results

Sources	Patients identified	Reviewed positive	PPV	Sensitivity
Allergy list	265	206	0.777	0.858
Encounter/Billing diagnosis	104	94	0.904	0.392
Medical history	33	30	0.909	0.125
Problem list	102	93	0.912	0.388
Total	310	240	0.774	1

Venn Diagram to Show the Distribution of DRESS Cases from Four EHR Sources
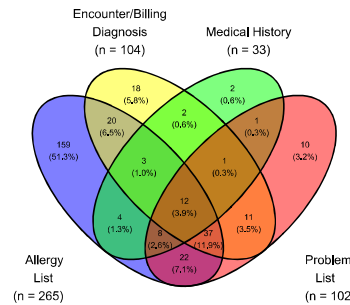
The Distribution of Positive DRESS Cases from Four EHR Sources
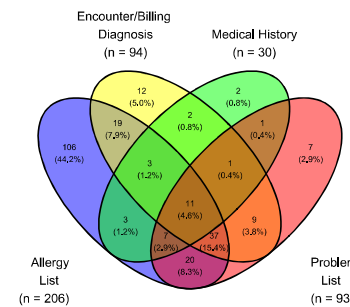
Quantitative findings

A total of 446 patients were identified from the EHR, and 136 were excluded during the first round of review due to insufficient records in the EHR

Among the remaining 310 cases, 245 from allergy list, 104 from problem list, 102 from billing diagnosis, and 33 from medical history.

DRESS syndrome was presented in 240 (77.4%) patients

For the four source EHR components, the PPVs were: 77.6% for the allergy list, 90.4% for the problem list, 91.2% for encounter billing/diagnosis, and 90.9% for medical history

Sensitivities were 85.8% for the allergy list, 39.2% for problem list, 38.6% for encounter billing/diagnosis, and 12.5% for medical history

189 patients were identified by a single EHR sources, of which 127 (67.2%) were positive DRESS cases

121 patients were identified by two or more sources, and 113 (93.4%) were positive cases

Conclusion

An EHR component alone is not sufficient to identify all DRESS cases and documentation quality varies

Although most of cases were only mentioned in allergy list, allergy documentation still requires improvement to support case finding and safe medication prescription

References

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