International Journal of Research in MEDICAL SCIENCE

ISSN Print: 2664-8733
ISSN Online: 2664-8741
Impact Factor: RJIF 8
IJRMS 2023; 5(2): 05-08
www.medicalpaper.net
Received: 05-05-2023
Accepted: 11-06-2023

Dr. Devanshu Kakani
MBBS, Dr. Vasantrao Pawar Medical, College, Hospital and Research Center, Nashik, Maharashtra, India

Dr. Sumit Biniwale
Junior Resident, Department of Paediatrics, Vilasrao, Deshmukh Government Medical College, Latur, Maharashtra, India

Dr. Sohan Sandeep Raut
MBBS Student, Dr. Vasantrao Pawar Medical, College, Nashik, Maharashtra, India

Corresponding Author:
Dr. Devanshu Kakani
MBBS, Dr. Vasantrao Pawar Medical, College, Hospital and Research Center, Nashik, Maharashtra, India

A rare case of dress syndrome with pancytopenia

Dr. Devanshu Kakani, Dr. Sumit Biniwale and Dr. Sohan Sandeep Raut

DOI: https://doi.org/10.33545/26648733.2023.v5.i2a.46

Abstract
Background: Drug Rash with Eosinophilia and Systemic Symptoms (DRESS) syndrome is a rare, uncommon and unexpected drug reaction that is usually associated with anticonvulsants, allopurinol, and antibiotics. The primary clinical signs are fever along with cutaneous abnormalities, eosinophilia and very rarely organ involvement. Pancytopenia is another rare but possible symptoms.
Case Presentation: We report a 47-year-old male patient with maculopapular rash on forehead and trunk, progressive breathlessness and decreased urine output after antibiotic treatment, which was initially diagnosed as erythema multiforme, acute renal failure and pancytopenia. Patient was made hemodynamically stable, renal replacement therapy and skin biopsy was done which evidently proved the diagnosis of DRESS with aplastic anemia and hence was appropriately treated with discontinuation of offending antibiotic and colony stimulating factor resulting in improvement of patient.
Conclusion: DRESS syndrome is a rare, possibly lethal illness that can present as pancytopenia, fever, eosinophilia, and atypical lymphocytes along with delayed onset of rash. The most recent drug use reportedly associated with DRESS should cause a preliminary suspicion to be raised.

Keywords: Dress syndrome, maculopapular rash, pustules

Introduction
A wide variety of pharmaceutical drugs such as sulfonamides, allopurinol, phenytoin or other anticonvulsants can cause a rare but potentially fatal adverse reaction known as the Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) syndrome. The syndrome is caused by wide variety of clinical symptoms including fever, skin rash, lymphadenopathy, hematologic abnormalities (most frequently eosinophilia and atypical lymphocytes) and internal organ involvement, as well as long latency period between 2 and 6 weeks after the offending agent is initiated [3]. Additionally, it takes time for symptoms even after the drug have been withdrawn. Autoimmune disorders including Graves's disease, Hashimoto disease, or autoimmune hemolytic anemia might develop over time as long-term consequences of the condition [1].

Case presentation
A 47-year-old male, known diabetic and hypertensive presented to casualty with severe rash, breathlessness and decreased urine output.
Two weeks ago, before his admission, he went to a general surgeon with a complaint of pustules over both feet for which he was given Amoxicillin and an analgesic. After that he developed a maculopapular rash which covered his entire body and face over a period of 2 days and breathlessness which progressed from initially at exertion to even at rest for which he was referred to our hospital for further management. Initially, though the patient did not have the classic rash, the diagnosis of erythema multiforme was suspected in view of the combination of the rash and recent drug exposure, with acute renal failure. On physical examination, the patient had fever 39 °C, bilateral pedal edema, raised JVP, bilateral basal crept on lung auscultation, BP 160/90, Pulse 112 bpm, SpO2 99% on 10 L of oxygen and RR 24/min. Patient had no clubbing, cyanosis and lymphadenopathy. A complete blood count revealed leukocytosis of 1200 cells/mm, comprising of 60% neutrophils and 8% of eosinophils (relative eosinophilia), platelets count of 82,000 cells/mm and Hemoglobin count of 6.8 mg/dl. His liver function tests with levels of total bilirubin 1.1 mg/dL, and SGOT/PT 10/25.

Urine microscopy, renal function tests with low urine output and serum creatinine of 5.4 mg/dL, serum calcium 8.2 mg/dL, serum uric acid 3.2 mg/dL, serum electrolytes [sodium 126 mg/dL, potassium 5.4 mg/dL and chlorides 103mg/dl] and blood urea of 167 mg/dL.

For initial hemodynamic stabilization and uremic symptoms, hemodialysis was done, and he was transfused with packed red cells and platelets. The patient was started on broad spectrum antibiotic Meropenem, as the symptoms and renal failure were attributed to the sepsis from the pustules in feet. After the administration of meropenem, he developed severe itching and rashes, which was suspected to be due to similarity of beta lactam ring between Amoxicillin and Meropenem, which points towards diagnosis of DRESS.

Bone marrow aspirate showed markedly hypocellular marrow which suspects the condition of aplastic anemia. The skin biopsy was done which showed eosinophilic infiltration in dermal layer.

Hence the diagnosis of DRESS syndrome was made (criteria given below); the offending drug recognized was amoxicillin. The component of renal failure was attributed to Acute Interstitial Nephritis (criteria) progressing into Acute Tubulo-interstitial Nephritis. The patient was kept on alternate day hemodialysis and eventually the urine output and creatinine improved but it did not normalize which was attributed to a background of CKD secondary to Diabetes and Hypertension. Initial therapy of amoxicillin and meropenem was discontinued as total counts normalized and thrombocytopenia resolved.

Table 1: Patients history

Clinical examination	P 112/min regular, BP 160/90, Spo2 99% on 10 L Oxygen, RR 24/min Pedal Oedema +, JVP+ Cyanosis, Clubbing and Lymphadenopathy absent
Auscultation	RS Fine Basal Crept Heard
Investigations	Hemoglobin: 6.8 mg/dL Leucocytes: 1200 cells/mm, 60% Neutrophils, 8% Eosinophils (Relative Eosinophilia) Platelets: 82000 cells/mm Blood Smear: Microcytic hypochromic Ferritin: 1106 CRP Quantitative:83.7 mg/dL Procalcitonin:6.6mg/ml
Renal function test	Blood Urea:167mg/dL Serum Creatinine: 5.4 mg/dL Serum Calcium: 8.0mg/dL Serum Uric Acid: 3.2mg/dL Serum Electrolytes: (Sodium/Potassium/Chlorides: 126/5.4/103 mg/dL)
Liver function tests	SGOT/PT:10/25 Total Bilirubin:1.1 mg/dL
Bone marrow biopsy	Markedly hypocellular marrow
Skin biopsy	Dermis showing eosinophilic infiltrate
Antibiotics given	Amoxicillin and Meropenem

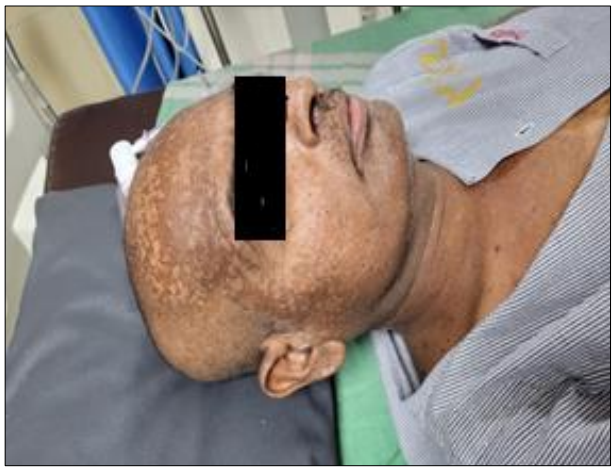

Fig 1: Widespread maculopapular rash over forehead and trunk (recovering)

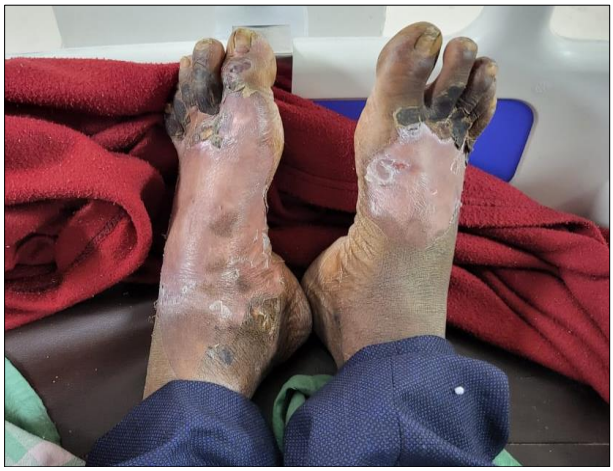

Fig 2: Pustules over both feet (recovering)

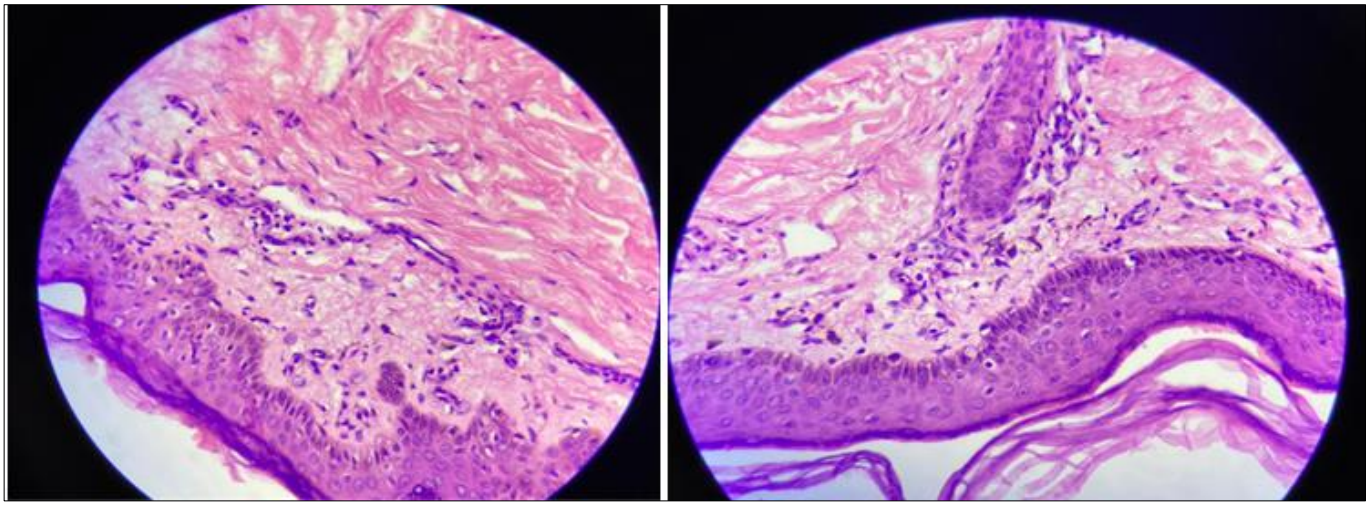

Fig 3(a), 3(b): Skin Punch Biopsy (Dermis showing eosinophilic infiltrate.)

Discussions

Adverse drug reactions leading to severe systemic involvement can be difficult to diagnose, especially when patients are critically ill. The already tricky diagnosis of DRESS becomes even more challenging with the superadded complication of bone marrow suppression. Various diagnostic criteria which are useful for diagnosing DRESS syndrome have been suggested which are listed below (Table 2) [4].

Table 2: Comparison of three diagnostic criteria in DRESS syndrome [4]

COMPARISON OF THREE DIAGNOSTIC SCORING SYSTEMS

RegiSCAR study group	Japanese consensus group	Bocquet *et al.*
More than 3 of the criteria are required for the diagnosis of DRESS	Typical DRESS (presence of all 7 criteria); atypical DIHS (all criteria present except lymphadenopathy and HHV-6 reactivation)	DRESS is confirmed by presence of 1 and 2 and 3
1. Hospitalization	1. HHV-6 reactivation	1. Cutaneous drug eruption
2. Reaction suspected to be drug related	2. Prolonged clinical symptoms 2 weeks after discontinuation of causative drug	2. Adenopathies >2 cm in diameter or hepatitis (liver transaminases >2 times upper limit of normal) (or) interstitial nephritis (or) interstitial pneumonitis (or) carditis
3. Acute rash	3. Maculopapular rash developing >3 weeks after starting drug	3. Hematologic abnormalities eosinophilia >1.5×10⁹/L (or) atypical lymphocytes
4. Fever above 38°C	4. Fever above 38°C	
5. Enlarged lymph nodes involving at least two sites	5. Lymphadenopathy	
6. Involvement of at least one internal organ	6. ALT >100 U/L or other organ involvement	
7. Blood count abnormalities	7. Leukocyte abnormalities (at least one)	
Lymphocytes above or below laboratory limits	Leukocytosis (>11×10⁹/L)	
Eosinophils above laboratory limits (in percentage or absolute count)	Atypical lymphocytosis (>5%)	
Platelets below laboratory limits	Eosinophilia (1.5×10⁹/L)	

DRESS=Drug reaction with eosinophilia and systemic symptom, ALT=Alanine aminotransferase, HHV-6=Human herpes virus-6, DIHS=Drug-induced hypersensitivity syndrome

Regardless of classification, all criteria were being met except for Eosinophilia/ Raised Counts which was obviously due to Pancytopenia. But, if bone marrow suppression is considered as an organ involvement, we may conclude that relative eosinophilia without the actual elevation of absolute eosinophil counts will fulfill the criteria for DRESS syndrome. Hence, the diagnosis of DRESS syndrome was made with the offending drug being amoxicillin.

Conclusion

Patients who have taken a new medication within the last two to six weeks may be suspected of having DRESS syndrome since early detection of this illness and immediate withdrawal of the responsible drug have the potential to save life. Unfortunately, there is no specific diagnostic approach that can clearly identify DRESS Syndrome; however, we have still mentioned some criteria that can identify DRESS syndrome (Table 1). (4) Re-exposure of drug should be avoided in patients with a history of DRESS syndrome. Due to the severity and the possibility of life-threatening consequences associated with this kind of drugs reaction, it is essential that every doctor is aware of DRESS.

Conflict of Interest

Not available

Financial Support
Not available

References

1. Kounatidis D, Vourlakou C, Panagopoulos F. Fever with pancytopenia in a patient with lamotrigin induced DRESS syndrome: A case-based review. EJMCR. 2022;6(4):64-67.
2. Gudi S, Biswas TK, Kane S. DRESS syndrome: A case report. MGM J Med Sci. 2021;8:312-6.
3. Lorenz Z A, Kish SM, Kalisky I, et al. DRESS Syndrome-An Unusual Presentation with Pancytopenia after Salazopyrin Treatment. Am J Biomed Sci& Res. 2020, 7(3).
4. https://www.jrpp.net/viewimage.asp?img=JResPharmPract_2017_6_2_130_207360_t3.jpg

How to Cite This Article
Kakani D, Biniwale S, Raut SS. A rare case of dress syndrome with pancytopenia. International Journal of Research in Medical Science 2023; 5(2): 05-08.