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(CASE REPORT)

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A case report on drug rash with Eosinophiilia and systemic symptoms (DRESS) syndrome

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Abstract

Background: DRESS syndrome is a distinctive, severe, and atypical drug reaction includes drug rash with eosinophilia and organ dysfunction symptoms, as well as hematological abnormalities. This disease is diagnosed using the Regis CAR (European Registry of Severe Cutaneous Adverse Reaction) scoring system.

Case presentation: A 25-year-old lady presented with complaints of skin lesion over whole body since 3 days along with multiple clear fluid filled blisters over face and chin and lesions in oral cavity, multiple ill-defined areas of erythema with multiple bullae over face and chin. She also had complained about was also having cough, breathlessness and chest pain.

Conclusion: DRESS syndrome is a severe hypersensitivity reaction caused by a reaction from drug, which in this case is found to be allopurinol. The treatment for the same is firstly the withdrawal of causative drug and using corticosteroids,in this patient dexamethasone was used.

Keywords: DRESS; Eosinophilia; (HHV)-6 and -7; RegiSCAR; Allopurinol-induced

1. Introduction

DRESS syndrome is a distinctive, severe, and atypical drug reaction [1] includes drug rash with eosinophilia and organ dysfunction symptoms [2], as well as haematological abnormalities [3]. This disease is diagnosed using the RegiSCAR (European Registry of Severe Cutaneous Adverse Reaction) scoring system [3] which was created to assign DRESS cases a "no," "possible," "probable," or "definite" status [4]. DRESS syndrome symptoms appear 2-8 weeks after taking the reaction-inducing drug [2], Fever and rash are usually the first symptoms [5] and Dysphagia, agranulocytosis, and chylous ascites are some of the unusual symptoms [1]. There is a limited number of laboratory data that can help distinguish DRESS syndrome from other severe drug reactions and identify asymptomatic internal organ involvement. These data include a complete blood cell count, which typically reveals eosinophilia and mononucleosis-like atypical lymphocytosis, liver function parameters, serum creatinine levels, and urinalysis. Thyroid stimulating hormone levels should also be measured and repeated after 2-3 months, as hypothyroidism can develop as a late complication [5]. Involvement of skin which occurs in 99-100% of patients, including both adults and children is found to be most common finding [6]. DRESS syndrome has been linked to antiepileptic drugs such as phenytoin, lamotrigine and carbamazepine. Antibiotics that contain sulfa, minocycline, and vancomycin have also been linked to this syndrome [7]. Kawasaki disease is the second most common cause of vasculitis in children after Henoch Schonlein purpura like DRESS

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Syndrome [8]. The foremost goal for managing the patients is to control the signs and symptoms promptly and safely, ultimately improving the quality of life of the patients and decreasing further number of hospitalizations [9].

The pathogenesis is still unknown [5]. Detoxification defects leading to reactive metabolite formation and subsequent immunological reactions, slow acetylation, and reactivation of human herpes, including Epstein-Barr virus and human herpesvirus (HHV)-6 and -7, have all been linked to its development [4]. In DRESS syndrome, the liver is the most commonly affected organ [5]. Studies recommend that L-ornithine-L-aspartate will lower blood concentration of ammonia and doubtless improve internal organ neurological disorder [10]. Adult mortality is estimated to be 10% and 5.4% in children [2].

2. Case study

A 25 year old female was admitted to skin department with complaints of skin lesion over whole body since 3days. Patient was relatively asymptomatic before 4 days after which she developed multiple areas of redness over face which increased in size and number, Since 2 days there was development of multiple clear fluid filled blisters over face and chin and lesions in oral cavity , multiple ill-defined areas of erythema with multiple bullae over face And chin. Multiple ill-defined areas of erythema, erythromatous papules over bilateral upper limbs coalescing over both arms, chest and abdomen. Multiple erythromatous papules patches over back and buttocks, palms and soles. This was followed by development of similar lesions over chest, abdomen, back, buttocks. This gradually increased in size and number to involve whole body. Patient complaints were associated with occasional itching (mild in intensity), fever (high grade) with chills and rigor, body ache, backache which were relieved by medications. Patient was also having cough, breathlessness and chest pain.

2.1. General examination

On general examination patient was conscious, cooperative and well oriented to time, place, person. Patient was examined in natural light, and consent was taken. CNS- conscious oriented; CVS- S1S2 (positive) no murmur ; RS- BLAE (+); GIT- soft, non-distended.

2.2. Medical history

Known case of gout and on medication since 20 days.

2.3. Treatment history

Patient was taking T.allopurinol for gout since 15 days. History of taking diclofenac + paracetamol combination 3days ago. T.Azithromycin, T.Levocet and T.methylprednisolone(10mg) was taken 2 days ago from local practitioner.

2.4. Family history

Not any similar history found.

2.5. Menstural history

G3P1L1A2. Last menstrual period was on 14/10/2021. Periods are irregular, painless and moderate flow.

2.6. Vitals

Table 1 Scorten Criteria

Prognostic Factor	0	1	Weight
Age	>40		0
Tachycardia	>120bpm	150bpm	1
Neoplasia			0
Detached or compromised body surface	>10%		1
Serum urea (mg/dl)	>28	27	0
HCO3 (mEq/L)	>20	<20	0

WBC	16500		0
Total			2

Temperature - 102.6°F, Pulse Rate- 150 beats/minute, Respiratory Rate - 18 breaths/minute, SpO2 - 95%on RA, BP-116/72 mm of Hg.

Table 2 Coagulation profile

Parameters	Result	Biological Ref. Range
PT Value	11.00 sec	11-13sec
PT Control	14.50 sec	(11-14sec)
ISI	1.20	-
INR	0.717	0.8-1.20
APTT Test Value	28.7	26-36
APTT control	34.90	-

Table 3 Laboratory findings

Parameters	Obtained value	Normal range	Interference
Hemoglobin	11.9	12-17.5 gm/dl	Decreased
WBCs	12030	4500-11000 /cmm	Normal
Neutrophils	74	40-80%	Normal
Lymphocytes	18	20-40%	Lymphocytopenia
Monocytes	1	2-10%	Monocytopenia
Eosinophils	8	1-6%	Eosinophilia
ESR	41	0-21mm/hr	Increased
MCV	77	80-96 fL	Normal
PCV	35.90	40-56%	Decreased
Platelet	340000	150000-450000 /cmm	Normal
RDWs	15	11.6-14%	Increased
ABSOLUTE EOSINOPHIL COUNT	1284	20-450 /cmm	Increased
CRP	96	0-6	Increased
SGOT(AST)	22	5-40 IU/L	Normal
SGPT(ALT)	34	7-56 IU/L	Normal
Serum ALB	3.5	3.5-5.5 g/dl	Normal
Serum urea	26	7-20 mg/dl	Increased

2.7. Urine analysis

- PH- 5.5 (4.6-8.0)
- PROTEIN- Trace SUGER – Nil
- PUS CELL- 8-12 /hpf BLOOD- Nil LEUCOCYTES- (+) Present

Table 4 Treatment plan

SR.NO.	DRUG	DOSE	FREQ.	
1.	INJ. CEFOTAXIME	1 gm	12Hrly	For 13 days
2.	ZOXAN DROPS (CIPROFLOXACIN)	0.3%w/v	BID	For 13 days
3.	INJ. INFUPAR (PARACETAMOL)	500 mg	12Hrly	For 7 days
4.	SYP. CODISTAR	4 mg/5 ml+ 10 mg/5ml	TDS	For 13 days
5.	INJ. PANTOPRAZOLE	40 mg	12Hrly	For 7 days
6.	TAB. LEVOCET	5 mg	BD	For 13 days
7.	BETAMETHASON CREAM	0.1%	LABD	For 13 days
8.	WHITE SOFT PRAFFIN	-	LABD	For 13 days
9.	TAB. AZITHROMYCIN	500 mg	OD	For 7 days
10.	FUSIDIC ACID CREAM	2% w/w	BD	Day 1 & then continued from day 4 to 13
11.	BETADINE GARGLES	2% w/v	TDS	For 13 days
12.	INJ. FLUCONAZOLE	200 mg	24Hrly	For 7 days
13.	FUCIBET CREAM	0.1%w/w	BD	For 13 days
14.	CLOTRIN MOUTH PAINT	1%w/v	BD	For 13 days
15.	CMC EYE DROP	0.5% w/v	BD	For 7 days
16.	TESS BUCCAL PASTE	0.1%w/v	BD	From 4th day
17.	INJ.DEXAMETHASONE	2cc	In the morning	For 13 days
18.	INJ.IV Ig	2 g/kg - 20cc/hr		For 7 days

3. Discussion

A rare condition known as DRESS Syndrome is considered to affect 0.9% out of every 1000,000 people. It is a systemic, severe drug hypersensitivity syndrome, typically encountered in adults but is uncommon in children. [2,7]. According to the RegiSCAR scoring system, which categorizes DRESS Syndrome as “excluded,” “possible,” “probable,” or “definite” and on this basis the diagnosis of the patient is made 3. The prolonged asymptomatic period that occurs between taking a medication and the onset of symptoms makes diagnosis convoluted, however, the difficultly of diagnosis results in a lower incidence of recognition and reporting [2],[1]. Ultrasound, if required, remains the preferred imaging modality. When further detailed images are needed, MRI without contrast is safe in pregnancy [11]. In our case report, A 25 year old female presented with the complaints of skin lesion over whole body since 3 days. The patient was relatively asymptomatic before 4 days after which she developed multiple areas of redness over face which gradually increased in size and number. This lesion coalesced to form large areas of reddish lesion over face in a span of 2 days. It was followed by development of similar lesion over chest, abdomen, back and buttocks. The lesion gradually increased in size and numbers to involve the whole body. Patient also had mild itching occasionally, High Grade Fever with chills and rigors, body ache, backache that worsen in the evening which was relieved upon medication. She was taking Tab. Allopurinol for gout since 15 days, Tab. Diclofenac + Paracetamol in combination. The patient was diagnosed with DRESS syndrome based on symptoms and laboratory reports while, the prominent cause was allopurinol. The most common mediations that cause DRESS syndrome are anticonvulsant (Carbamazepine, phenytoin, lamotrigine and phenobarbital), anti-tuberculous treatments, captopril, calcium Channel blockers, thalidomide, Sulphasalazine antibiotics (Sulphonamides, vancomycin), Non-steroidal anti-inflammatory drugs and antiretroviral drugs (zalcitabine, neviparine). It has been suggested that the development of the syndrome is caused by an accumulation of the allopurinol metabolite oxy-purional, especially when there is a deterioration in renal clearance and the use of thiazide diuretics [5].

For the treatment of dress syndrome, immediate withdrawal of causative drug and use of corticosteroids for treatment the hypersentitivity reaction. In this case report patient was given Inj. dexamethasone which is a potent glucocorticoid, decreases inflammation by suppressing migration of polymorphonuclear leukocytes and reducing capillary permeability. For topical treatment, betamethasone cream which is also a potent glucocorticoid was given the patient to relieve inflammation over the skin. Fusidic acid cream is an antibiotic, works by preventing synthesis of essential proteins required by bacteria to carry out vital functions, thus preventing spread of infection, was also given to the patient to treat multiple erythematous papules. Inj. fluconazole an antifungal medication was used to prevent fungal infection in the patient. For fever Inj. infupar (paracetamol) was given, syrup codistar for cough, zoxan drops for eye infection, CMC drops as a lubricant for dry eyes, pantoprazole for acidity, Tab. azithromycin for skin infection. Lastly Inj. Intra Venus IgG was given as supportive therapy.

4. Conclusion

DRESS syndrome is a severe hypersensitivity reaction caused by a reaction from drug. Particularly, in this case report, the culprit drug is allopurinol due to which hypersensitivity reaction was found. It is essential to diagnose this syndrome at an early stage so that the identified medication can be discontinued and re-exposure can be avoided. Thereafter an appropriate treatment regimen can be initiated. The high rate of reactivation of HHV-6 and other herpes viruses associated with DRESS suggests that HHV-6 and other herpes viruses should be detected in routine clinical practice. SCORTEN criteria is also used to to identify severity-of-illness in in-hospital mortality**.** RegiSCAR is a simplistic and trustable tool for confirming a clinical suspicion of DRESS. More research is needed for early detection of DRESS cases, as well as to develop a better treatment protocol to reduce morbidity and mortality associated with it.

Compliance with ethical standards

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Disclosure of conflict of interest

All authors declare that they have no conflict of interest.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors.

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