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Cast 36 – Immunological Emergency

A **55-year-old man** with a history of **Multiple Myeloma** presents to the Emergency Department with **fever, shortness of breath and rash**. He underwent an **autologous stem cell transplant 15 days ago**. On examination he has a **maculopapular rash** involving the arms, axilla, torso and abdomen.

Vital signs:

- HR 101 bpm
- BP 144/76 mmHg
- RR 26 /min
- Temp 39°C
- SpO_2 89% on room air (96% on oxygen)

Blood results:

Test	**Result**
Hb	101 g/L
WCC	4.0
Neutrophils	2.9
Lymphocytes	0.4
Monocytes	0.6
Platelets	97
CRP	15

(a) Apart from infection, list four (4) possible diagnoses that could explain this presentation. (4 marks)

- **Transplant-related complications**
  - Engraftment syndrome
  - Peri-engraftment respiratory distress syndrome (PERDS)
  - Acute graft-versus-host disease (rare with autologous transplant)
- **Drug hypersensitivity reaction**
- **Drug Rash with eosinophilia (DRESS)**
- **Diffuse Alveolar Haemorrhage**
- **Cytokine release syndrome**

(b) Patient has been moved to Resus, connected to physiological monitoring and commenced on HFNP oxygen therapy. In addition to haematology and biochemistry, he has had blood cultures sent. State six (6) key points of assessment management for this patient (6 marks).

- **Temperature control:** IV paracetamol 1gm and active cooling (tepid sponging, fan, cooled IV fluids).
- **Treat as neutropenic/immunocompromised sepsis:** Urgent broad-spectrum IV antibiotics (e.g. piperacillin–tazobactam / cefepime / meropenem with doses).
- **Add targeted anti-infectives post Respiratory PCR:** Empiric antivirals (e.g. Tamiflu for influenza or acyclovir if other viral aetiology if suspected) (anti-fungals accepted if justified)
- **Seek and treat other causes of hypoxia:** such as pulmonary embolism (CTPA) or haemorrhage / PERDS (CXR/CT).
- **Commence systemic corticosteroids for suspected engraftment:** Methylprednisolone 1-2mg/kg/day
- **Escalation and disposition:** Early haematology involvement, consider bronchoscopy/BAL, ICU review ± transfer to tertiary centre.

Other Possible SAQ questions that can be asked

Feature	Engraftment Syndrome	Graft-versus-Host Disease (GVHD)
Typical Timing	Around **neutrophil recovery (Day 7–21)**	**Acute GVHD:** usually **<100 days** post transplant **Chronic GVHD:** >100 days
Type of Transplant	Common after **autologous transplant**	Occurs almost exclusively after **allogeneic transplant**
Pathophysiology	Cytokine release during **rapid neutrophil recovery**	**Donor T-cells attack host tissues**
Skin Findings	Diffuse **maculopapular rash**, often transient	**Maculopapular rash** that may progress to erythroderma or desquamation
Pulmonary Features	**Pulmonary infiltrates**, hypoxia, capillary leak common	Lung involvement **uncommon in acute GVHD** (more chronic bronchiolitis obliterans)
GI Involvement	Usually mild or absent	**Prominent diarrhoea**, abdominal pain
Liver Involvement	Usually mild or absent	**Elevated bilirubin / cholestatic LFTs** common
Fever	Very common	May occur but less dominant feature
Course	**Acute inflammatory syndrome**, short course	Can be **progressive and persistent**
Response to Steroids	**Rapid improvement**	Variable response, may require prolonged immunosuppression

Simple Exam Rule (Very High Yield)

Engraftment Syndrome

- Occurs **around neutrophil recovery**
- **Fever + rash + pulmonary infiltrates**

Acute GVHD

- Occurs **within 100 days**
- **Rash + diarrhoea + liver dysfunction**

Practical ED Clues

Think **Engraftment Syndrome** if:

- Occurs **10–20 days post transplant**
- **Autologous transplant**
- Fever + rash + hypoxia

Think **GVHD** if:

- **Allogeneic transplant**
- **Profuse diarrhoea**
- **Abnormal LFTs**
- Progressive rash

Condition	Typical Timing After Transplant	Key Clinical Features	Investigations	Initial ED Management
Neutropenic Sepsis	Usually **Day 0–30** (pre-engraftment)	Fever, rigors, hypotension, minimal inflammatory response	Blood cultures, FBC, lactate, CXR, urine culture	**Immediate broad-spectrum IV antibiotics** (e.g. piperacillin-tazobactam / cefepime), sepsis resuscitation
Bacterial / Viral Pneumonia	**Early post-transplant (first month)**	Fever, cough, SOB, hypoxia	CXR, CT chest, respiratory PCR (COVID, influenza, RSV), sputum culture	Oxygen, **empiric antibiotics**, antivirals if suspected
Fungal Infection (Aspergillus, PJP)	**2–6 weeks**	Persistent fever, cough, dyspnoea, hypoxia	CT chest, BAL, fungal markers	Early **antifungal therapy**, specialist input
Engraftment Syndrome	**Around neutrophil recovery (Day 10–20)**	Fever, **maculopapular rash**, pulmonary infiltrates, hypoxia, capillary leak	CXR, bloods to exclude infection	**Systemic corticosteroids**, supportive care
PERDS (Peri-Engraftment Respiratory Distress Syndrome)	**Engraftment period (Day 10–20)**	Acute hypoxia, pulmonary infiltrates, respiratory distress	CXR, CT chest	Oxygen, **steroids**, ICU if severe
Acute GVHD	Usually **within 100 days** (allogeneic transplant)	**Skin rash**, diarrhoea, abdominal pain, liver dysfunction	LFTs, stool tests, biopsy if required	**Systemic corticosteroids**, haematology involvement
Chronic GVHD	**>100 days**	Autoimmune-like disease: skin sclerosis, dry eyes, oral ulcers, lung disease	Organ-specific tests	Immunosuppression, specialist management
Drug Reactions / Toxicity	Any time	Rash, fever, organ dysfunction	Medication review	Stop offending agent, supportive care

Organ System	Common Features
Skin	Maculopapular rash, erythroderma, sclerosis
GI tract	Diarrhoea, abdominal pain, nausea, GI bleeding
Liver	Elevated bilirubin, cholestatic LFT pattern
Eyes	Dry eyes, keratoconjunctivitis sicca
Lungs	Bronchiolitis obliterans (chronic GVHD)