THE TREATMENT OF LATE-ONSET HEPATIC VENO-OCCLUSIVE DISEASE FOLLOWING HEMATOPOIETIC STEM CELL TRANSPLANTATION USING DEFIBROTIDE: A CASE

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Case study

- 27 year old lady was diagnosed with Acute Myeloid Leukaemia (AML) in March 2018 at 24 weeks gestation on routine blood tests.
- Treatment with DA 3 + 7 (Daunarubicin and Cytarabine) was commenced, under the joint management of haematology and obstetrics. A complete remission (CR) was achieved.
- A healthy baby boy was delivered in July 2018.
- Patient declined further treatment and follow up, and subsequently presented with relapsed AML in November 2018.
- Patient agreed to further treatment and received FLAG-IDA . CR2 was achieved.
- A myeoloablative hematopoietic stem cell transplant (HSCT), with Cyclophosphomide and Bulsuphan conditioning was undertaken in March 2019.
- Donor was a female, 10/10 matched sibling, CMV pos/neg.
- Patient engrafted timely and was discharged home on day 24 post transplant.
- She presented on day +32 post transplant with nausea, acute abdominal pain and was admitted with suspected acute cholecystitis; no clinical findings were suggestive of VOD at that time. A liver ultrasound scan was unremarkable.
- Abnormal liver function tests were found on admission and monitored daily.
- Patient did not comply with fluid balance and urine collection/ measurement.
- Distended abdomen and right upper quadrant pain on palpation was identified.
- Weight increased from 50.5kg to 59.5kg over a 10-day period (17.8% weight gain).
- Bilirubin began to rise rapidly to 37 µmol/L over a 5 day period. Liver ultrasound scan was repeated.
- These factors along with fluid retention, oedema and portal vein disruption on repeat liver ultrasound scan supported the diagnosis of severe late onset VOD (Mohty et al, 2016).

Background

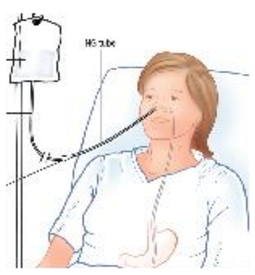
- Veno-Occlusive Disease (VOD) is also known as sinusoidal obstruction syndrome (SOS).
- It is a potentially life-threatening complication of HSCT.
- Prevalence is 10-15% of adult HSCT patients (Richardson et al, 2019).
- VOD usually occurs within the first 21 days after HSCT.
- VOD presenting after this time is called late onset VOD (Mohty et al, 2016).
- VOD occurs when the small blood vessels in and around the liver become blocked.
- There are several factors that increase the risk of VOD such as age, gender, conditioning regime, prior treatment and underlying diagnosis (Duncan et al, 2019).

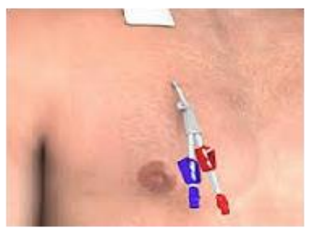
VOD characteristics

Rapid weight gain
Not ahributed to excessive fluid intake, together with oedema and/or ascites

Hepatomegaly and/or RUQ pain
Hepatomegaly may only be evident on ultrasound

Hyperbilirubinemia
Rapid in both onset and progression
Anicteric VOD has been reported in 13% of adult pa8ents with VOD post-HCT (Corbacioglu et al, 2018)





Blood products keep Platelets over 30

Physio therapy and exercise

Daily weights & strict fluid balance

Psychological support

Diuresis & albumin

Management

Optimise nutrition

Daily bloods tests

Analgesia & supportive medications

Consider IV access i.e., central line

Treatment

- Defibrotide is approved for the treatment of severe hepatic VOD post HSCT in patients >1 month of age (Richardson et al, 2019).
- Treatment with defibrotide 25mg/kg in four divided doses was administered intravenously.
- Patient was reviewed by gastroenterology and other causes of acute hepatitis were excluded.
- Platelets were transfused to maintain a level greater than 30 to reduce the risk of bleeding.
- The patient was also treated for CMV reactivation with IV foscarnet.
- IV electrolytes were given to manage low potassium , magnesium and phosphate.
- A urinary catheter was inserted so diuresis and fluid balance could be managed effectively.
- Defibrotide was administered for 21 days, liver function normalised and weight returned to baseline.
- Patient was discharged home.

Conclusions

- This case study highlights the successful treatment of late onset hepatic veno-occlusive disease following HSCT, with intravenous defibrotide and supportive medications.
- Interventions and management such as strict fluid balance, daily bloods, daily weights and nutritional interventions are integral to the successful outcome of this potentially life threatening complication.
- Psychological support should be offered by the nursing team, clinical nurse specialist and/ or psychologist.
- In this case the patient was discharged home and was subsequently monitored in the transplant clinic

References

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