EHA EUROPEAN HEMATOLOGY ASSOCIATION

EHA-ISHBT Hematology Tutorial

Self-assessment Case – Session 1

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Hyderabad, India
March 1-3, 2024

CASE STUDY 1

- 25-year-old male
- Exertional dyspnoea for 8 months
- Increased fatiguability for 8 months

EXAMINATION FINDINGS

- MODERATE PALLOR
- NO ICTERUS
- NO CLUBBING
- NO CYANOSIS
- NO LYMPHADENOPATHY
- NO EDEMA

EXAMINATION FINDINGS

- Cardiovascular and central nervous systems – NORMAL
- Respiratory system – bilateral rhonchi and wheeze
- Abdomen – soft, non tender, no organomegaly
- Skin/oral mucosa – within normal limits
- Musculoskeletal system – within normal limits
- Premature graying of hair starting from 11 years of age

MEDICATION HISTORY

- No significant history

FAMILY HISTORY

- Family history: death of his father due to interstitial pulmonary fibrosis at the age of 60 years.

COMPLETE BLOOD COUNT

- Hb – 90 g/l
- WBC – 2.5 × 10^9/l
- Platelet count – 120 x 10^9/l
- Differential count – neutrophils 30%/lymphocytes 60%/eosinophils 5%/monocytes 5%
- MCV – 98 fl
- MCH – 32 pg
- MCHC – 300 g/l

OTHER LABORATORY PARAMETERS – 1

- Reticulocyte Count – 0.5%
- Direct Coombs test – negative
- Lactate dehydrogenase – 325 U/L
- Ferritin – 325 ng/ml
- Iron – 90 µg/dl
- Total iron binding capacity – 300 µg/dl
- Transferrin saturation - 30%
- Renal function tests – within normal limits
- Liver function tests – within normal limits

OTHER LABORATORY PARAMETERS - 2

- Hepatitis B, Hepatitis C and human immunodeficiency virus (HIV)-1 & 2 – negative
- Vitamin B12 – 245 pg/ml
- Testing for paroxysmal nocturnal haemoglobinuria (PNH) by FLAER – negative

RADIOLOGY

- Computed tomography (CT) CHEST – INTERSTITIAL PULMONARY FIBROSIS

- CT ABDOMEN – WITHIN NORMAL LIMITS

PULMONARY FUNCTION TESTING

- Moderate to severe restrictive lung defect

CARDIAC IMAGING

- No significant abnormality detected on 2-D echocardiography

BONE MARROW BIOPSY

- Hypoplastic bone marrow

CONVENTIONAL KARYOTYPING

- NORMAL KARYOTYPE

CHROMOSOMAL BREAKAGE ANALYSIS

- NORMAL

NEXT GENERATION SEQUENCING

- *TERC* GENE MUTATIONS

TO SUMMARISE….

- 25-year-old male
- Non-transfusion dependent aplastic anaemia
- Telomeropathy
- Interstitial pulmonary fibrosis
- Premature graying of hair
- Family history of death of father due to interstitial pulmonary fibrosis

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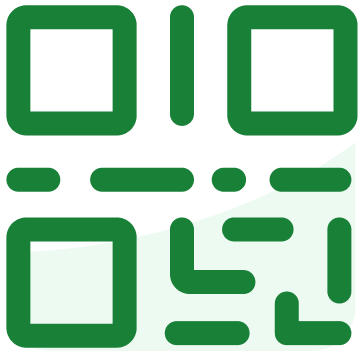

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Question 1

How would you manage this case ?

1. Upfront Allogenic Stem Cell Transplant
2. Danazol + Cyclosporine
3. Danazol only
4. Cyclosporine + Elthrombopag

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1.41 How would you manage this case ?

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Question 2

Type of Stem Cell Transplant Preferable for this patient

1. Matched sibling Allogenic stem Cell Transplant with Myeloablative conditioning.
2. Matched unrelated Allogenic stem cell transplant with Myeloablative conditioning.
3. Matched sibling Allogenic stem cell transplant with Reduced intensity conditioning.
4. Matched unrelated Allogenic stem cell transplant with reduced intensity conditioning.

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1.42 Type of Stem Cell Transplant Preferable for this patient

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QUESTIONS

- Your line of management for this case?
- Should transplant be done upfront in this case?
- Type of conditioning regimen to be used?
- Does transplant help to reverse his interstitial pulmonary fibrosis?
- What type of donor to be selected for transplant?
- Medical line of management for such case?
- Does elthrombopag have any role in this case?
- Does romiplostim has any role in this case?

CASE STUDY 2

- 45-year-old male
- Non-A , non-B , non-C hepatitis induced Acute liver Failure
- Opted for cadaveric liver transplant
- 1 month after liver transplant – develops pancytopenia requiring packed red cell and platelet support.
- Patient was on steroids + ciclosporin (liver transplant protocol)

EXAMINATION FINDINGS

- Marked Pallor
- No icterus
- No cyanosis
- No clubbing
- No lymphadenopathy
- Moderate bilateral pedal edema

EXAMINATION FINDINGS

- Cardiovascular, central nervous and respiratory systems – within normal limits
- Abdomen – soft, non tender, no organomegaly
- Skin/oral mucosa – within normal limits
- Musculoskeletal system – normal

Medication and family history

- No significant medication or family history

COMPLETE BLOOD COUNT

- Hb – 45 g/l
- WBC count – 1.5 × 10^9/l
- Platelet count – Less than 10 × 10^9/l
- Differential count – neutrophils 10%/lymphocytes 80%/eosinophils 5%/monocytes 5%
- MCV – 96 fl
- MCH – 32 pg
- MCHC – 30 g/l
- RDW – 15.2 %

OTHER LABORATORY PARAMETERS -1

- Reticulocyte count – 0.3%
- Direct Coombs test – negative
- Lactate dehydrogenase – 146 U/L
- Ferritin + iron studies – within normal limits
- Creatinine – 1.1 mg/dl (0.7 mg/dl – 1.3 mg/dl)
- Total bilirubin – 3.0 mg/dl (0.1 mg/dl – 1.2 mg/dl)
- Direct bilirubin – 2.5 mg/dl (less than 0.3 mg/dl)
- Indirect bilirubin – 0.5 mg/dl
- SGOT (AST)(Aspartate Aminotransferase)– 175 U/L
- SGPT(ALT)(Alanine Aminotransferase) – 185 U/L

OTHER LABORATORY PARAMETERS - 2

- Hepatitis B, hepatitis C and human immunodeficiency virus (HIV) -1 & 2 - negative
- Vitamin B12 – 350 pg/ml
- Testing for paroxysmal nocturnal haemoglobinuria (PNH) by FLAER - negative

RADIOLOGY

- Computed tomography (CT) – chest and abdomen - No significant abnormality

CARDIAC IMAGING

- No significant abnormality detected on 2 –D echocardiography

Bone Marrow Biopsy

- Hypoplastic marrow

CONVENTIONAL KARYOTYPING

- Normal

CHROMOSOMAL BREAKAGE ANALYSIS

- Normal

NEXT GENERATION SEQUENCING

- NO MUTATION DETECTED

TREATMENT

- Started on horse antithymocyte globulin (ATG) (40 mg/kg) x 4 days
- Ciclosporin dose upgraded to 5 mg/day in 2 divided doses
- Eltrombopag could not be given – rejected by liver transplant team.
- After 6 months of horse ATG and ciclosporin, he remains transfusion dependent with no improvement in counts.

To summarise…..

- 45-year-old male
- Transfusion-dependent severe aplastic anaemia
- Developed 1 month after cadaveric liver transplant for non-A/non-B/non-C Hepatitis induced acute liver failure
- Failed ATG and ciclosporin
- Elthrombopag could not be given

Question 3

How would you manage this case ?

1. Upfront Allogenic stem cell transplant
2. Repeat immunosuppressive therapy with Rabbit ATG + Cyclosporine
3. Cyclosporine + Inj. Romiplostim
4. Cyclosporine + Elthrombopag (at reduced Doses)

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1.43 How would you manage this case ?

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Question 4

How would you manage this case?

1. Should this patient undergo allogeneic stem cell transplant?
2. Conditioning regimen of choice?
3. Any correlation with liver transplant and development of aplastic anaemia?
4. Any role of romiplostim in this case?

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1.44 How would you manage this case?

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CASE CONTINUED …….

- Patient was started on romiplostim (10 µg/kg/week) and subsequently increased to 15 µg/kg/week
- After 6 months of romiplostim his counts stabilised
- Hb – 80 g/l
- WBC – 4.0 × 10^9/l
- Differential count – neutrophils 60%/lymphocytes 35%/eosinophils 2%/monocytes 2%
- Platelet count – 38 × 10^9/l

CASE CONTINUED ……

- After 12 months of romiplostim
- Hb – 80 g/l
- WBC count – 5. 0 × 10^9/l
- Platelet count – 40 × 10^9/l

QUESTIONS

- How long to continue Inj. Romiplostim ?
- Can we stop Inj.Romiplostim now and how to stop Inj. Romiplostim?


thank
you