Transplant Eligible Patients with Amyloidosis

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Objectives

- Brief overview of Amyloidosis
- Eligibility criteria for auto BMT
- Considerations for stem cell mobilisation/collection and transplant.
- Case Study and discussion
**Amyloidosis**

- General term used to refer to the deposition of fibrils in extracellular tissues.

- Fibrils arise from variety of proteins that have undergone structural change.

- These deposits result in wide range of clinical manifestations depending on their type, location and the amount of deposition.
Type / Classification

- **AL** (formerly known as Primary Systemic Amyloidosis)
  - Immunoglobulin light chain Amyloid deposition
  - Associated with plasma cell dyscrasias – multiple myeloma, some B cell lymphomas

- **AA** (previously known secondary Amyloidosis)
  - Amyloid A Protein
  - Associated with chronic inflammation- rheumatoid arthritis, Tuberculosis.
Clinical Presentation

- Nephrotic syndrome
- Restrictive Cardiomyopathy
- Peripheral Neuropathy
- Hepatomegally
- Macroglossia
- Purpura and other skin manifestations - racoon eyes.
Diagnosis

- AL amyloidosis is suspected in a patient presenting with any one of the following
  - Non diabetic proteinurea
  - Restrictive cardiomyopathy in otherwise unexplained CCF
  - Unexplained Oedema, hepatosplenomegaly or carpel tunnel syndrome
  - macroglossia
Eligibility Criteria

<table>
<thead>
<tr>
<th>Clinical Factors</th>
<th>Organ Function</th>
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<tbody>
<tr>
<td>Age 65yrs</td>
<td>Cardiac Biomarkers</td>
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<tr>
<td>NYHA class I-II</td>
<td>GFR &gt;50</td>
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<tr>
<td>ECOG 2</td>
<td>Bilirubin &lt;1.5 ULN</td>
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<td>SBP &gt;90</td>
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<td>Negative features, no syncope, cardiac failure, arrhythmias</td>
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<td>Positive features- isolated renal amyloid, isolated peripheral neuropathy</td>
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</tbody>
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Case Study

- Mr DM  61 yr  Music Teacher
- Diagnosed April 2017, significant proteinurea, Nephrotic syndrome
- Renal Biopsy confirmed AL
- Induction therapy 4 Cycles CVD, put forward for AutoSCT
Stem Cell Mobilisation

- Higher rates of complications during mobilisation and collection
- GCSF only
- Consideration for cardiac monitoring / telemetry during mobilisation / apheresis and infusion.
Stem Cell Transplant (HPC reinfusion)

- 4 bags HPC-A infused in CCU, nil complications.
- Discharged to ward 4 hours post infusion
- Remained inpt until neutrophil recovery.
Peri Transplant

- Addition of arrythmia prophylaxis
- Attention to Fluid balance
- Avoidance of GCSF
- Prevention GI Haemorrhage
- Avoidance nephrotoxins
- MDT care
DM

- Admission 6/12/2017
- ANC >0.5 day 15
- Plts >20 day 16 (transfused day 9)
- Afebrile throughout, blood product support entailed single unit of pooled platelets, nil PRBC
- Discharged day 16
Outcomes
Discussion Point

- Induction therapy before AutoSCT
- Cardiac monitoring
- Collection experiences.
Questions?
References

- eViQ id 3045v.2  Considerations for patients with Primary AL amyloidosis prior to autologous BMT.
- EBMT guidelines 2012
- ABMTRR Annual Data Summary 2014
- ABMTRR Annual data Summary 2016.