Transplant Eligible Patients with Amyloidosis

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July 2018

NSW GOVERNMENT
Health
Central Coast
Local Health District

Objectives

- Brief over 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

Clinical Factors	Organ Function
Age 65yrs	Cardiac Biomarkers
NYHA class I-II	GFR >50
ECOG 2	Bilirubin <1.5 ULN
SBP >90	
Negative features, no syncope, cardiac failure, arrhythmias	
Positive features- isolated renal amyloid, isolated peripheral neuropathy	

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

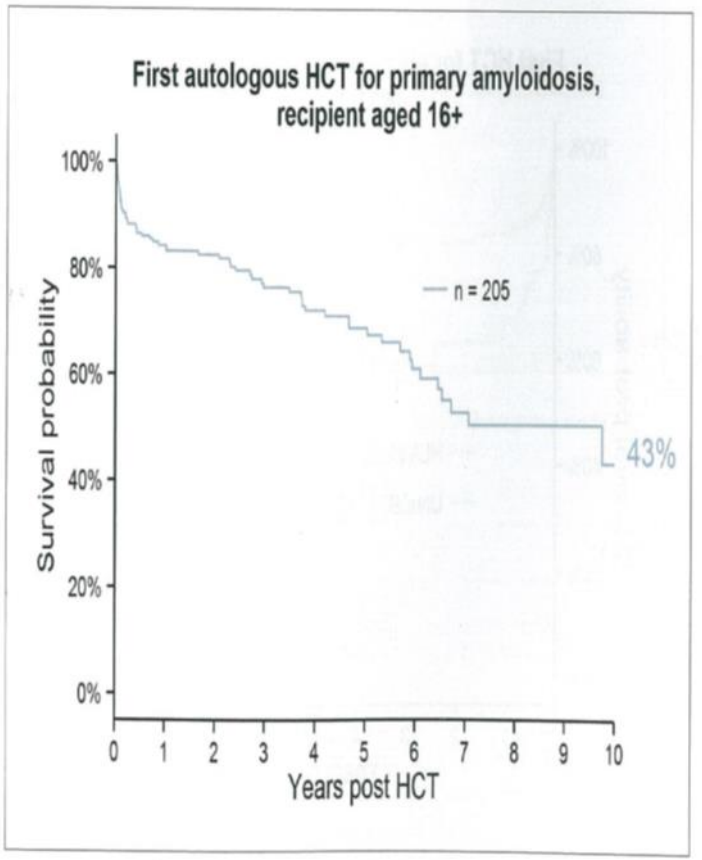
DM

- 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

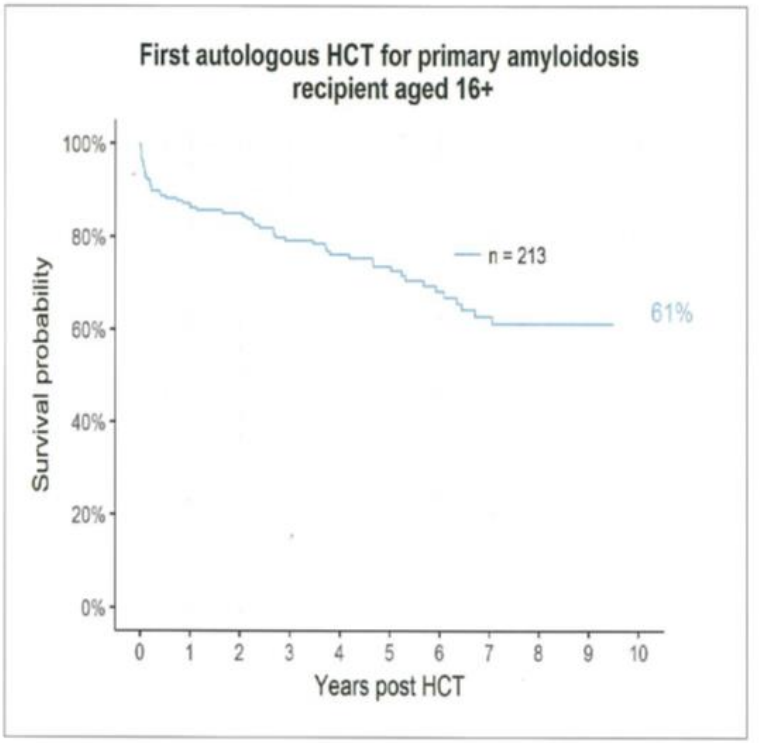
Primary amyloidosis



ABMTRR Annual Data Summary 2016

Ten-year survival - first transplants 2001-2015

Primary amyloidosis



Discussion Point

- Induction therapy before AutoSCT
- Cardiac monitoring
- Collection experiences.

Questions?

References

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