AN ELDERLY PATIENT WITH MYELODYSPLASTIC SYNDROME

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DISCLOSURES

• None
CASE PRESENTATION

• 70 year old Caucasian female patient Mrs. CC is married, has 2 children and works as a secretary

• She was referred to CHBAH from a lower level hospital with persistent pancytopenia and increasing transfusion requirements (October 2019)

• When she came to our hospital, she presented with:
  • Fever
  • Fatigue, dizziness and occasional headaches
  • Easy bruisability

• Comorbidities: Hypothyroidism on levothyroxine

• No known significant drug or toxin exposures

• No prior malignancies or exposure to radiation or chemotherapy

• No family history of malignancies
Alert, palor, pyrexial but haemodynamically stable

Bilateral tonsillitis with no exudate

No lymphadenopathy, hepatomegaly or splenomegaly

Skin: bruising on venipuncture site and some petechiae on both legs

No clinical evidence of failure of the heart, liver or kidneys
# LABORATORY WORK UP

<table>
<thead>
<tr>
<th>Full blood count</th>
<th>Urea &amp; Electrolytes</th>
<th>Haematinics</th>
<th>Liver function tests</th>
<th>Culture results</th>
</tr>
</thead>
<tbody>
<tr>
<td>WCC: 1,24 x 10^9/L</td>
<td>Normal</td>
<td>Ferritin: Normal</td>
<td>Unremarkable</td>
<td>Blood: Neg</td>
</tr>
<tr>
<td>HB: 7,2 g/dl</td>
<td>Folate: Normal</td>
<td>B12: Normal</td>
<td></td>
<td>Urine: Neg</td>
</tr>
<tr>
<td>MCV: 93,7 fl</td>
<td></td>
<td>EPO: 647 mU/dl</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PLT: 27 x 10^9/L</td>
<td></td>
<td>RPI: 0,3</td>
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</tbody>
</table>

CXR: Normal
Trilineage haemopoiesis with dysplasia in all three haemopoietic cell line. Additionally there are 12% myeloid blasts. Features in keeping with **Myelodysplastic syndrome with excess blasts (MDS-EB-2)**
FURTHER RESULTS

- Flow cytometry: myeloid blasts positive for myeloperoxidase
- Cytogenetics: 46, XX and deletion 7q
- FISH: Negative for t(8;21), inversion 16 and del 17p
  - Deletion 7q
- NGS myeloid mutational panel: Not done
PRE-TREATMENT ASSESSMENT

• Revised international prognostic scoring system (IPSS-R score):
  • 8 (very high-risk)

• Functional status: ECOG PS 1

• Echocardiogram: 69% ejection fraction & structurally normal heart

• MDS Comorbidity index score: 0 (low risk)

• Independent in all activities of daily living (ADLs) and IADLs, though with fatigue

• Frailty score: Intermediate fitness

• **Summary:** Mrs CC is a 70 year old patient who presented with features of bone marrow failure. We diagnosed her with very high-risk MDS. Pre-treatment assessment revealed good functional status, low risk comorbidity score and intermediate frailty score
MANAGEMENT

• Supportive care
  • Blood products support
    • Leuco-depleted packed red cells
    • Single-donor platelets
  • Treatment of infection
  • Patient education and psychosocial support
  • Involvement of palliative care team
SPECIFIC TREATMENT

- Allogeneic HSCT
- Hypomethylating agents: Azacitidine or decitabine
- Intensive chemotherapy
- Enrolment into clinical trial
• Our patient received Cytarabine 100mg IVI daily for 5 days as an inpatient

• Monitored and supported for 37 days in hospital

• She had delayed and incomplete recovery of cytopenias
  • Hb and neutrophil count improved initially for ~2 months but subsequently dropped to slightly above baseline
  • Blood transfusion requirements 2-3 weekly
  • Platelets improved to >50 x 10^9/L

• Repeat BMAT: ~10% myeloblasts with 7q deletion on FISH analysis

• **Way forward??**
• We discussed with Mrs. CC and her family regarding the limited treatment modalities in our (public health) setting

• A decision was made for best available supportive care

• Patient is being seen 2 weekly in haematology clinic
  • On thioguanine, deferasirox, prophylactic antibiotics & antifungals

• Patient is willing to be treated with HMAs whenever they are available in the hospital

• She would like to participate in a clinical trial for MDS treatment whenever it becomes available in our clinic
CONCLUSION

• Outcomes in MDS depends on:
  • Disease factors: Risk category
  • Patient factors: Medical fitness
  • Available treatment modalities & experience of the clinical team in managing the condition

• There is need to improve treatment modalities in low- to middle income countries to improve outcomes in MDS
THANK YOU FOR LISTENING