QUESTION 70
A 67-year-old man with a history of heavy smoking presents with new onset angina. On examination he is found to be plethoric and hypertensive with no other abnormalities noted.
His full blood examination shows:
- **Haemoglobin**: 205 g/L [128-175]
- **Haematocrit**: 57% [36-50]
- **Mean cell volume (MCV)**: 78 fL [80-97]
- **White cell count**: 11.6 x 10^9/L [3.9-12.7]
- **Neutrophils**: 9.50 x 10^9/L [1.50-6.00]
- **Lymphocytes**: 0.60 x 10^9/L [0.70-3.15]
- **Monocytes**: 0.30 x 10^9/L [0.15-0.60]
- **Eosinophils**: 0.80 x 10^9/L [0.00-0.40]
- **Basophils**: 0.40 x 10^9/L [0.00-0.15]
- **Platelets**: 499 x 10^9/L [150-396]
- **Red cell mass**: 39 mL/kg [28-35]
- **Plasma volume (derived)**: 49 mL/kg [40-50]
- **Serum erythropoietin**: 4.9 IU/L [4.8-21.9]

The most likely explanation for these findings is:
A. carcinoma of the lung.
B. high affinity haemoglobin.
C. dehydration.
D. chronic obstructive pulmonary disease.
E. polycythaemia vera.

**Polycythaemia**

<table>
<thead>
<tr>
<th>Type</th>
<th>Condition</th>
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<tbody>
<tr>
<td>True</td>
<td>Polycythaemia rubra vera</td>
</tr>
<tr>
<td>Primary</td>
<td>EPO appropriately increased</td>
</tr>
<tr>
<td>Secondary</td>
<td>EPO inappropriately increased</td>
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<tr>
<td></td>
<td>1. Renal disease: hypernephroma, renal cyst,</td>
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<tr>
<td></td>
<td>hydrenephrosis</td>
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<td></td>
<td>2. Uterine myoma</td>
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<td>3. Other tumours eg. Hepatocellular carcinoma,</td>
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<td></td>
<td>bronchial carcinoma</td>
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<tr>
<td>Relative</td>
<td>Plasma volume depletion</td>
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<tr>
<td></td>
<td>Stress (‘pseudopolycythaemia’)</td>
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<tr>
<td></td>
<td>Dehydration</td>
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<td>Diuretic therapy</td>
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True polycythaemia exists when the **total red cell mass (RCM)**, measure by dilution of isotopically labelled red cell, is increased above normal.

Relative polycythaemia exists when an elevated haemoglobin concentration is caused by **reduction in plasma volume**

**Clinical features**
1. Raised RCM – ruddy complexion
2. Hyperviscosity – headaches and visual disturbances, thrombosis
3. Haemorrhage
4. Pruritus, gout
Year 2005 Paper two: Questions supplied by Ilynn

5. Enlarged spleen (75%) (distinguishes between PCV and others)

**Lab findings**
1. Raised haematocrit, red cell count and RCM
2. Raised WCC (75%)
3. Bone marrow: hypercellular with prominent megakaryocytes
4. Iron stores are depleted
5. Abdo U/S – exclude renal disease, assess spleen size

**Differential dx**
1. Relative polycythaemia arise when plasma volume is reduced
   - Dehydration
   - Vomiting
   - Diuretic therapy
2. Young male adults (especially smokers)
   - Ass with stress, increased vasomotor tone and HT
   - WCC, plt count, RCM and bone marrow normal

Other test:
1. Cxr – exclude lung disease
2. Haemoglobin oxygen dissociation curve – to identify a variant Hb with increased O2 affinity
3. Serum EPO assay

**Treatment**
1. Asprin
2. Regular venesection
3. Chemotherapy (oral hydroxyurea)

**Prognosis**
Median survival is 16 years – thrombosis main cause of morbidity and mortality
30% develop myelofibrosis
5% develop AML