

## Slide 2 - September 2006 / Cycle 29

### ESSENTIAL THROMBOCYTHAEMIA

#### Please read this bit first

The HPCSA and the Med Tech Society have confirmed that this clinical case study, plus your routine review of your EQA reports from Thistle QA, should be documented as a "Journal Club" activity. This means that you must record those attending for CEU purposes. Thistle will **not** issue a certificate to cover these activities, nor send out "correct" answers to the CEU questions at the end of this case study.

The Thistle QA CEU No is: MT00025

Each attendee should claim **THREE** CEU points for completing this Quality Control Journal Club exercise, and retain a copy of the relevant Thistle QA Participation Certificate as proof of registration on a Thistle QA EQA.

#### FORWARD

*This clinical page may not exactly match the slide due to the need to vary the clinical descriptions for CPD purposes.*

Scanned and edited from Essential Haematology by Hoffman, Pettit and Moss. Blackwell. (2001).

In this condition there is a sustained increase in platelet count because of megakaryocyte proliferation and over production of platelets. A persisting platelet count of  $>600 \times 10^9 / l$  is the central diagnostic feature but other causes of a raised platelet count need to be excluded before the diagnosis can be made.

#### Clinical and laboratory findings.

The dominant clinical features are thrombosis and haemorrhage. Many cases are symptomless and diagnosed on routine blood counts. Thrombosis may occur in the venous or arterial systems (Fig. 17.4b) whereas haemorrhage, as a result of abnormal platelet function, may cause either chronic or acute bleeding. A characteristic symptom is erythromelalgia, a burning sensation felt in the hands or feet and promptly relieved by aspirin. Up to 40% of patients will have palpable splenomegaly whereas in others there may be splenic atrophy because of infarction. Abnormal large platelets and megakaryocyte fragments may be seen in the blood film (Fig. 17.7). The bone marrow is similar to that in PRV but an excess of abnormal megakaryocyte is typical. Cytogenetics and molecular analysis for the *BCR-ABL* fusion gene are analysed to exclude chronic myeloid leukaemia. The condition must be distinguished from other causes of raised platelet count (Table 17.4). Platelet function tests are consistently abnormal, failure of aggregation with adrenaline being particularly characteristic.

#### Treatment.

The principle is to control the platelet count so as to reduce the risk of thrombosis which is the major clinical problem. The patients may be placed in risk groups according to age, size of platelet count and previous episodes of thrombosis or haemorrhage. The thrombotic risk depends on other risk factors such as smoking history and hypertension, and the treatment should take account of these risks. In those with a high risk, the aim is to keep the platelet count below  $600 \times 10^9 / l$ . Hydroxyurea is probably the most widely used treatment although  $\alpha$ -interferon is also valuable in younger patients.

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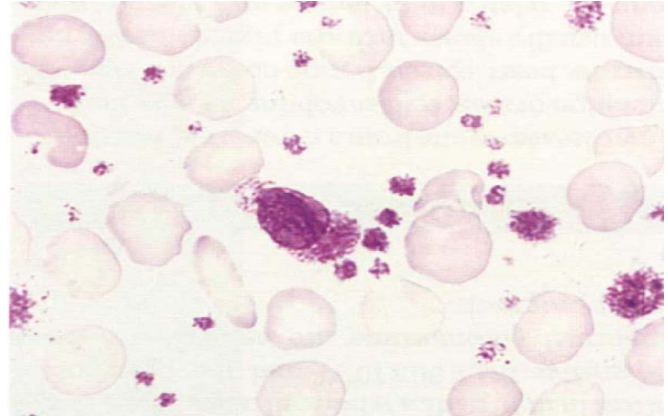


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Busulphan and  $^{32}\text{P}$  were used but are not now favoured because of possible long-term complications. Platelet pheresis may be helpful in short-term management. Aspirin is commonly used to reduce thrombotic risk and in patients younger than 60 years with no previous thrombosis or haemorrhage and platelets  $< 1000 \times 10^9 /\text{l}$  it may be the treatment of choice.



**Fig. 17.4 (b)** Gangrene of the left fourth toe in essential thrombocythaemia.



**Fig. 17.7** Peripheral blood film in essential thrombocythaemia showing increased numbers of platelets and a nucleated megakaryocytic fragment.

### **Course.**

Often the disease is stationary for 10-20 years or more. Patients may transform after a number of years to myelofibrosis; the risk of transformation to acute leukaemia is relatively low (<5%).

### **Table 17.4** **reactive**

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Haemorrhage, trauma, postoperative  
Chronic iron deficiency  
Malignancy  
Chronic infections  
Connective tissue diseases, e.g. rheumatoid arthritis  
Postsplenectomy

### **Endogenous**

Essential thrombocythaemia  
In some cases of polycythaemia vera, myelofibrosis and chronic myeloid leukaemia

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### **CEU Questions:**

1. Discuss the links between an increased platelet count and the risk of thromboses and haemorrhage.
2. What morphological feature would help confirm the diagnosis of essential thrombocythaemia ?

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