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### Acute erythremic myelosis (true erythroleukaemia): a variant of AML FAB-M6

Our group has been actively researching the acute erythroleukaemias since the late 1980s, during which time, we have developed and extensively published our classification<sup>1-3</sup>:

- acute erythroleukaemia, M6a (traditional FAB-M6, DiGuglielmo's syndrome, 1917)
- acute erythroleukaemia, M6b (pure erythroleukaemia, DiGuglielmo's disease, 1926)
- acute erythroleukaemia, M6c (mixed erythroleukaemia).

We have also published abstracts and presented this classification at meetings of the International Academy of Pathology, International Society of Haematology, and the American Society of Haematology, and our recommendations have also been cited in the recent literature. Therefore, we are surprised to find the M6b subtype now "discovered" and re-named in the article by Hasserjian *et al.*<sup>6</sup> and the M6c subtype completely overlooked. Although we suspect this incongruity to be an oversight, we believe it is important to set the record straight.

This established classification of the acute erythroleukaemias is based partly on the old FAB criteria and also upon morphological, cytochemical, and immunophenotypical criteria.<sup>2</sup> All bone marrow aspirates demonstrate  $\geq 50\%$  erythrocytic precursors, with erythroid dysplasia. Dysplasia of the granulocytic and megakaryocytic cell lines may or may not be present. The M6a subtype is defined as  $\geq 30\%$  blasts of the non-erythrocytic component (FAB exclusion criteria); the M6b subtype is defined as  $\geq 30\%$  pronormoblasts of the erythrocytic elements; and the M6c subtype has  $\geq 30\%$  blasts and  $\geq 30\%$  pronormoblasts by the aforementioned exclusion criteria. Because the dysplastic changes may, at times, make definitive characterisation of the blasts as erythrocytic versus non-erythrocytic difficult, the morphological features must always be confirmed by cytochemical stains, immunohistochemical stains, and/or flow cytometric analysis.

These three separate subtypes must be distinguished from one another to provide useful prognostic information for the clinician and the patient. When treated with the standard myeloid protocol, the M6a and M6c subtypes demonstrate a very high remission rate, whereas most patients with the M6b subtype remain refractory to treatment. Notably, patients with the M6c subtype remain in remission for a significantly shorter time than the M6a group. Mean survival for these subtypes is: M6a, 31.4 (SD, 32) months; M6b, 3.15 (SD, 4.2) months; M6c, 10.5 (SD, 12.7) months.

The malignant clonal cell of origin manifesting as acute erythroleukaemia of any subtype appears to be a multipotential stem cell,<sup>3</sup> which shows varying degrees of erythrocytic and granulocytic lineage maturation. Therefore, the three distinct subtypes of acute erythroleukaemia are not three separate diseases, but rather represent a spectrum of the same disease. The poor remission rate and short survival characteristic of this disorder<sup>3</sup> are dependent upon: (1) a high pronormoblast to myeloblast ratio within diagnostic bone marrow aspirates, (2) a high proliferative index, (3) "unfavourable" cytogenetic aberrations, and (4) a high

incidence of P-glycoprotein expression (the multidrug resistance phenotype).

**F M Mazzella**

Medical College of Georgia, Augusta,  
30912-3605 Georgia, USA;  
[fmazzella@mail.mcg.edu](mailto:fmazzella@mail.mcg.edu)

**H R Schumacher**

University of Cincinnati Medical Centre, Cincinnati,  
45367 Ohio, USA

### References

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- 4 Mazzella FM, Kowal-Vern A, Shrit MA, *et al.* Effects of multidrug resistance gene expression in acute erythroleukaemia. *Mod Pathol* 2000;**13**:407-13.
- 5 Mazzella FM, Alvares C, Kowal-Vern A, *et al.* The acute erythroleukaemias. *Clin Lab Med* 2000;**20**:119-37.
- 6 Hasserjian RP, Howard J, Wood A, *et al.* Acute erythremic myelosis (true erythroleukaemia): a variant of AML FAB-M6. *J Clin Pathol* 2001;**54**:205-9.

## CORRECTIONS

**A comparison of 13 guinea pig and human anti-tissue transglutaminase antibody ELISA kits.** Wong RCW, Wilson RJ, Steele RH, *et al.* *J Clin Pathol* 2002;**55**:488-95. The affiliation of Dr RH Steele should be South Western Area Pathology Service.

**Detection of the CD56+/CD45- immunophenotype by flow cytometry in neuroendocrine malignancies.** Bryson GJ, Lear D, Williamson R, *et al.* *J Clin Pathol* 2002;**55**:535-7. The quotation on page 535 was inadvertently cut out of the first paragraph (it should have remained there as the second sentence) and should begin with CD56 not CD59.



## Detection of the CD56+/CD45– immunophenotype by flow cytometry in neuroendocrine malignancies.

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