The magic of immersion oil: gray platelet syndrome

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A 28-year-old male presented with copious spontaneous epistaxis. He had a history of mild epistaxis, easy bruising and long-lasting hemorrhages after accidental cuts. Blood count revealed: leukocytes: 3.7×10^9/L with a normal differential, hemoglobin: 147 g/L and platelets: 47×10^9/L with a mean platelet volume of 10.5 fL.

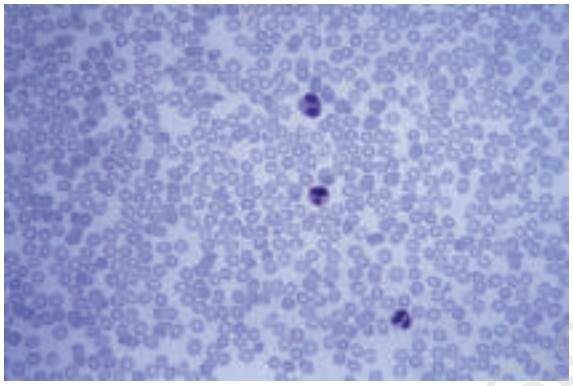

Figure 1. Peripheral blood film showing erythrocytes and 3 neutrophils; it seems to be an absence of platelets (May-Grünwald-Giemsa, 400×).

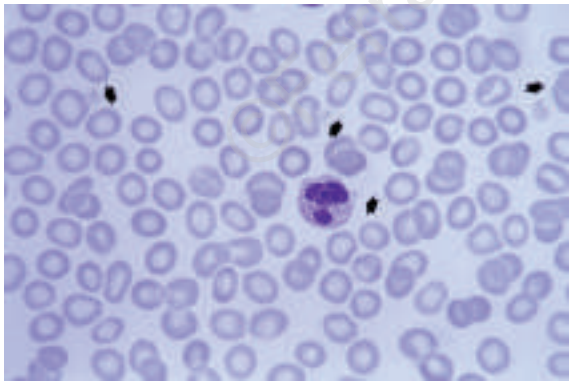

Figure 2. Magnification of the central area of Figure 1. Agranulated platelets with a grayish hue (arrows) can be seen and differentiated from erythrocytes (May-Grünwald-Giemsa, 1000×).

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Well-prepared peripheral blood smears, without anticoagulant in order to avoid platelet swelling, were stained with May-Grünwald-Giemsa. Examining these smears with a 40× objective on light microscope, erythrocytes and leukocytes showed no morphological abnormalities but, surprisingly, platelets seemed to be absent (Figure 1). Platelets were identified, however, using the 100× oil-immersion objective (Figure 2) and had no granules, a grayish or bluish tonality and well-defined edges. They were round and moderately large, many with vacuoles. A diagnosis of gray platelet syndrome was made.

Transmission electron microscope confirmed the diagnosis, revealing a total absence of α-granules in more than 80% of platelet sections (Figure 3). In the remaining sections α-granules were very few and small. In contrast, dense bodies were absolutely normal in morphology and number.

Other outstanding ultrastructural features were a marked development of surface-connected canalicular and dense tubular systems, and an increased number of mitochondria. A specific α-granule proteins release study was performed on platelets stimulated with collagen 3 µg/mL. Protein levels, determined by ELISA technique, were clearly reduced: platelet-factor 4: 38 $U\times10^3$ platelets (normal value

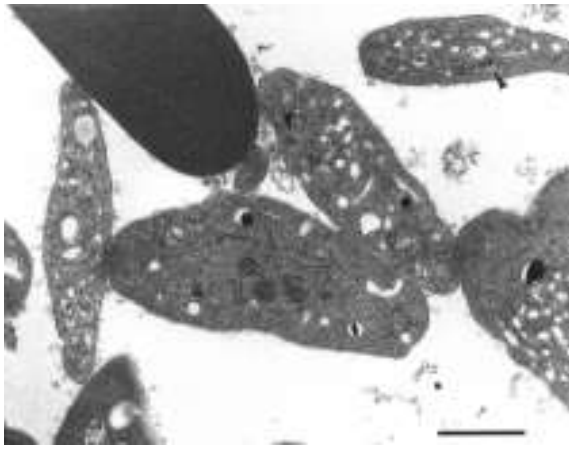

Figure 3. Ultrastructural platelet morphology showing α-granules deficiency. Few and small α-granules can be identified (arrowheads). Dense bodies appear in normal number and morphology, bar. 1 µm.

> 450), β-thromboglobulin: 92 U×10^3 platelets (NV > 500) and thrombospondin: 154 μg×10^3 platelets (NV > 200).

Gray platelet syndrome is a rare disease characterized by thrombocytopenia, an almost complete absence of platelet α-granules and a lifelong hemorrhagic diathesis.[1-4]

Since the first description by Raccuglia in 1971,[1] very few patients have been reported;[5] however, this diagnosis may be more frequent if careful examination of peripheral blood smears is performed. Figures 1 and 2 emphasize the importance of microscopic immersion oil in the routine studies of platelet morphology and illustrate how the complete gray platelet syndrome may otherwise be overlooked.

References

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