Indian J Hematol Blood Transfus (July-Sept 2019) 35(3):603–604
https://doi.org/10.1007/s12288-019-01129-2

IMAGES

Peripheral Blood Smear: A Clue to MYH9-Related Disorder

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Received: 6 January 2019 / Accepted: 25 April 2019 / Published online: 4 May 2019

An 11-year-old boy presented with severe thrombocytopenia. There was no history of bleeding or fever. Complete blood count showed hemoglobin-10.8 g/dl; TLC 5.54×10^3/cu.mm; platelet count of 20×10^3/cu.mm; MPV of 9.5 fl (reference interval: 6.5–10 f) and ESR of 15 mm/1st h. Immunologic tests showed normal Anti DS DNA: 7.2 U/ml, Anti Cardiolipin Ab: 1.5 U/ml, and ANA: 0.2 (Negative result less than 1.0) and CRP (< 0.3 mg/dl). Review of Peripheral Blood Smear (PBS) revealed giant platelets with large blue intracytoplasmic inclusions in periphery of the cytoplasm of WBC series (Fig. 1). No toxic change or left shift was seen in the peripheral smear. The findings were suggestive of May–Hegglin anomaly. Similar finding was seen in the PBS of the patient's father. Molecular testing for confirmation could not be done. The morphology was persistent during 2 years by checking every 6 months interval. Döhle body like inclusions within neutrophils are highly suggestive of MYH-9-related disorders. MYH-9-related disorder is a rare autosomal dominant macrothrombocytopenia syndrome which was previously classified as May–Hegglin anomaly with subtypes of Sebastian, Fechtner and Epstein syndromes [1]. Misdiagnosis with immune thrombocytopenia often leads to inappropriate administration of immunosuppressive and steroid therapy [2]. Evaluation of the PBS for giant platelets and Döhle body-like inclusions in the granulocytic series is important for correct diagnosis of this condition.

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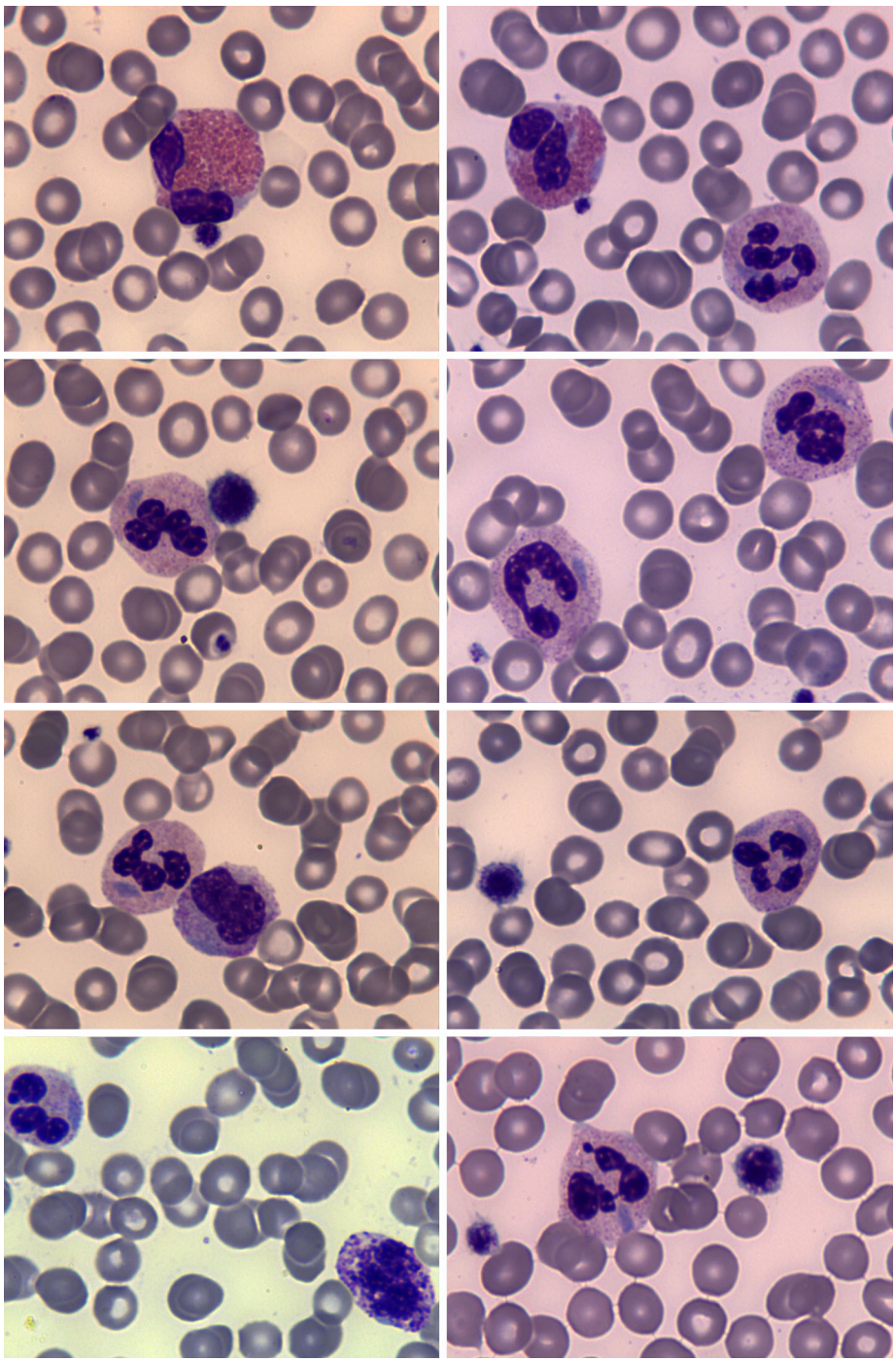

Fig. 1 Giant platelets with large blue intracytoplasmic inclusions in periphery of the cytoplasm of WBC series

References

1. Bolton-Maggs PH et al (2006) A review of inherited platelet disorders with guidelines for their management on behalf of the UKHCDO. Br J Haematol 135(5):603–633

2. Pecci A et al (2018) MYH9: structure, functions and role of non-muscle myosin IIA in human disease. Gene 664:152–167