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The May–Hegglin anomaly in a kidney transplant recipient

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We report a 37-year-old renal transplanted patient with thrombocytopaenia. He had May–Hegglin anomaly (MHA) inherited from his father, who died due to myocardial infarction subsequent to two renal transplantations. At the age of 15 years, proteinuria was found, but he was not biopsied. In 1982, he underwent splenectomy because of trauma. In 1989, when he was 17, renal function was found to be decreased. Five years later, he started haemodialysis and at 24 years he underwent live related kidney transplantation. Serum creatinine was always found stable around 1.9 mg/dl on cyclosporine, azathioprine and steroids. Automated blood count reported a number of platelets around 8000/μl with a mean volume of 20.3 fl (normal range 7.2–11.1 fl). However, blood cell counters have a defined volume window for platelet counting and large macrothrombocytes are classified as red blood cells, meaning that platelet number is underestimated (Figure 1). The true value of platelets after count with the microscope was around 30 000/μl. He was completely asymptomatic, without any bleeding tendency.

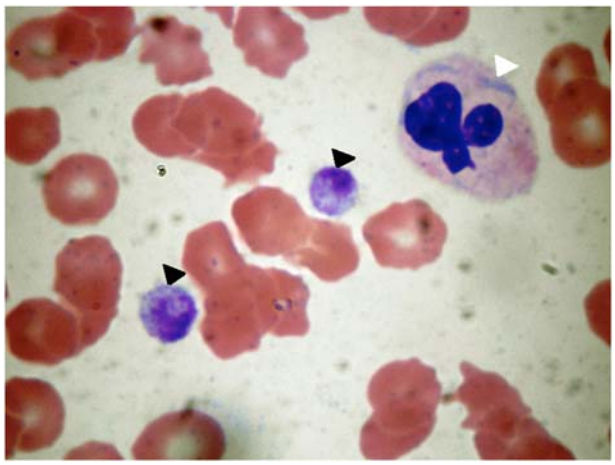

Fig. 1. Large platelets (black arrow head) and granulocyte with cytoplasmic inclusions (white arrow head); blood smear, May–Grunwald Giemsa stain.

MHA is a rare autosomal dominant platelet disorder, characterized by macrothrombocytopaenia and granulocyte inclusion bodies. The mutation involves the MYH9 gene, localized in chromosome 22q, which encodes a heavy chain of non-muscle myosin IIA. MYH9-related hereditary macrothrombocytopaenia disorders include: MHA, Ebstein, Fechtner and Sebastian syndromes, which are characterized by nephritis, hearing loss and cataract [1]. In 1992, Nel *et al.* [2] reported two cases of MHA incidentally discovered in a patient and his brother during investigation for end-stage renal failure and workup for renal transplantation. Alhindawi and Al-Jbour [3] described an association between Epstein syndrome and chronic nephropathy. Recently, Singh *et al.* [4] suggested that MYH9 gene alterations are associated with a greater risk of focal segmental glomerulosclerosis and hypertensive nephrosclerosis in African Americans.

Conflict of interest statement. None declared.

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