A Patient with May-Hegglin Anomaly who underwent cervical laminectomy and laminoplasty

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Background

May-Hegglin anomaly (MHA) is an autosomal dominant disorder with MYH9 mutations, characterized by a triad of thrombocytopenia, giant platelets, and granulocyte inclusion bodies. We report the case of MHA patient who underwent cervical laminectomy and laminoplasty without blood transfusion.¹⁾

Case

Case: 58-year-old woman, 144cm, 42kg
Diagnosis: Cervical Spondylotic Myelopathy
Past History: Hemodialysis for 43 years (due to nephrotic syndrome), Splenectomy
Preoperative Blood Test: Platelet Count 6,000/ μ l
Surgical Procedure: Laminoplasty and foraminotomy on three cervical vertebrae
Estimated Blood Loss: 300ml

Discussion

- Counters do not count giant platelets correctly.^{2) 3)}
Since the counters do not count giant platelets correctly, automated counters underestimate 30-50% compared to the true platelet counts.
- Platelet aggregation study tends to be with-in normal limits.^{2) 3)}
Although some case studies of MHA patients performed preoperative platelet transfusions, gathering these facts and considering the risk of platelet transfusion, we decided that preoperative platelet transfusion is not required in our case.

Learning points

- ✓ Most of the MHA patients do not require preoperative routine platelet transfusion.
- ✓ Understanding the character of MHA and distinguishing with other thrombocytopenia diseases is important to prevent unnecessary treatments.

Based on Alport syndrome-like symptoms, presence of giant platelets on peripheral blood smear testing(Fig. 1), and thrombocytopenia with intact platelet function(Fig.2), genetic test was performed, and diagnosed as MHA. Her postoperative course was uneventful.

Fig.1. Giemsa-stained peripheral smear confirmed the presence of giant platelets.

Fig.2. Platelet aggregation study with ristocetin was within normal limit.

Our case	Platelet count
Automated count	6,000/ μ l
Direct visual count	80,000/ μ l

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3) Sehbai AS, Abraham J, Brown VK. Perioperative management of a patient with May-Heggling anomaly requiring craniotomy. American Journal of Hematology 2005;79:303-308