



Regional Anaesthesia in Harris Platelet Syndrome for Transurethral Resection of Prostate-A Clinical Conundrum or Certitude?

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Abstract

Routine use of the autoanalyzer has helped uncover the increasing incidence of thrombocytopenia. Disorders associated with macrothrombocytes with thrombocytopenia necessitate a preoperative evaluation to assess the bleeding tendencies and the need for transfusion of blood products. Harris platelet syndrome is one such disorder where macrothrombocytes with thrombocytopenia are associated with no congenital abnormalities and low risk of bleeding intraoperatively. There are cases where Harris platelet syndrome has been treated with steroids or splenectomy, which is unwarranted. We report successful management of a patient with Harris platelet syndrome who underwent transurethral resection of the prostate under spinal anaesthesia with no complications.

Keywords: Bleeding, megakaryocytes, spinal anaesthesia, thrombocytopenia

Introduction

Neuraxial anaesthesia is increasingly being utilized in patients with thrombocytopenia. The routine use of autoanalyzers has uncovered the increasing number of cases of thrombocytopenia (1, 2) and calls for a further evaluation preoperatively. Disorders of associated macrothrombocytes with thrombocytopenia have unique clinical profile. Common to these disorders with low to near normal platelet counts and macrothrombocytes is the varying degree of bleeding. Bengal thrombocytopenia, also called asymptomatic constitutional macrothrombocytopenia or Harris platelet syndrome is the most common inherited giant platelet disorder. It is a commonly observed condition in the population in eastern and north-eastern India, Bangladesh, Bhutan, and Nepal. We present a case report of a 63-year-old male with Harris platelet syndrome who underwent transurethral resection of the prostate. Meager literature is available regarding the bleeding tendencies of such patients and their anaesthetic management.

Case Presentation

A 63-year-old gentleman from north eastern India, was presented to the preanaesthetic clinic for evaluation for transurethral resection of the prostate. He had no known medical comorbid illnesses and had an unremarkable history of past surgical events. Routine blood investigations revealed thrombocytopenia (58×10^9 cells L^{-1}). Peripheral smear for manual platelet counts revealed a platelet count of 150×10^9 cells L^{-1} , and an associated normal appearance of the platelets, macrothrombocytes without inclusion bodies. There were no inclusion bodies in the leukocytes. A focused history of epistaxis, bleeding from minor trauma, and extensive bruising was considered to rule out that the congenital platelet disorder was negative. Acquired platelet deficiencies because of the intake of nonsteroidal anti-inflammatory drugs (NSAID), liver disease, megaloblastic anemia, sepsis, and autoimmune disorders were ruled out. Bleeding time, clotting time, prothrombin time, activated partial thromboplastin time, and international normalized ratios were all normal. A literature search and discussion with the hematologist revealed such a presentation to be Harris platelet syndrome, though there was a paucity of case reports or studies on anaesthetic management of such patients.

The patient was prepared for the procedure under spinal anaesthesia after acquiring written informed consent. A 27 G Pencan needle was used to administer subarachnoid block in L3-L4 interspace in the first attempt, using 2.4 mL of 0.5% heavy bupivacaine. A sensory level of T10 and a modified Bromage scale of 3 was achieved in few minutes. The surgery was successfully completed in 45 minutes, throughout which the patient was stable hemodynamically. Blood loss was 100 mL. The patient was monitored in the post-anaesthesia care unit (PACU) for 60 min, and the motor and sensory blockades had recovered completely by then. The patient was followed up for 3 days postoperatively during which he had no neurologic deficit. The patient was discharged on postoperative day 3. He had no neurologic deficit two weeks after the procedure when he visited the urology outpatient clinic for follow up.

Discussion

The small non-nucleated platelets, derived from the bone marrow megakaryocyte precursors, are discoid shaped cells measuring $2.0 \times 4.0 \times 0.5 \mu\text{m}$, with a mean volume of 7-11 fL. The normal count is $150\text{-}450 \times 10^9 \text{ cells L}^{-1}$. The bone marrow megakaryocytes are the larger parent cells, reaching up to 50 μm in diameter. Congenital deficiency of platelets, along with the presence of megakaryocytes in the blood is rare and occur, for instance, in MYH9 gene-related disorders such as May-Hegglin anomaly and Sebastian syndrome. Many of these disorders have an association with features such as sensorineural hearing loss, glomerulonephritis, and cataract (3). These disorders have been vastly studied and are seen to manifest defective platelet function, in addition to the low counts and large size, and hence, more than usual perioperative bleeding (4).

Harris et al reported patients with mild to severe thrombocytopenia, macrothrombocytes, anisocytosis, absent bleeding symptoms, and normal platelet aggregation studies with absent MYH9 mutation from the Northeast Indian population (5-7). These patients with asymptomatic constitutional macrothrombocytopenia, also called as Harris platelet syndrome, are asymptomatic and do not report exaggerated bleeding manifestations. In fact, they remain undiagnosed until rou-

tine blood work is done for an unrelated cause. The macrothrombocytopenia is attributed to altered levels of secondary cytoskeletal proteins necessary to maintain the structural aspects of the platelets. Hence, the functional integrity of the platelets is maintained but structurally they are macrothrombocytes (8).

This very common inherited giant platelet disorder is prevalent in about one-third of the population in the Indian subcontinent (9). Such patients are unnecessarily subjected to extensive evaluation such as invasive bone marrow examination. Lack of awareness about this syndrome has led to treatment of such patients with corticosteroids, platelet transfusions, immunoglobulin infusion, and even splenectomy (10).

It is of relevance, therefore, that Harris platelet syndrome is recognized preoperatively and unwarranted interventions such as bone marrow examination, steroid therapy, and platelet transfusions are avoided perioperatively. There is very limited information on this issue, partly because of the seemingly localized geographic distribution of the affected individuals, and reported lack of clinical symptoms. We have reported the conduct of regional anaesthesia in a patient with Harris platelet syndrome for transurethral resection of the prostate, a surgery where bleeding is a major concern. Our patient underwent transurethral resection of the prostate (TURP) without major blood loss under spinal anaesthesia with no neurological deficit postoperatively. In this era of enhanced recovery after surgery, we highlight our clinical experience in this case report, in a patient with Harris platelet syndrome, to increase the awareness about this silent entity, and thus help prevent morbidity of unwarranted evaluation, therapy, and unnecessary health care expenditure on these otherwise healthy individuals.

Conclusion

In patients with Harris platelet syndrome, where there is macrothrombocytopenia with no clinical manifestations of bleeding and with normal coagulation parameters, regional anaesthesia can be safely administered without risk of neuraxial hematoma.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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Main Points:

- Macrothrombocytes with thrombocytopenia with no congenital abnormalities have low risk of bleeding peri-operatively.
- Pre-operative manual platelet count and peripheral smear is warranted in such patients which usually reveal normal platelet count with macrothrombocytes without inclusion bodies.
- It is safer to proceed with spinal or neuraxial anesthesia in such patients with Harris platelet syndrome.

Conflict of Interest: The authors have no conflicts of interest to declare.

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