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GLANZMANN'S THROMBOAESTHENIA – A RARE CAUSE OF EPISTAXIS: A CASE REPORT

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ABSTRACT Very few case reports exist of patients presenting with epistaxis as a manifestation ofGlanzmann's thromboaesthenia (GT). Here we report one such case of a young male who presented with severe nasal bleeding and was diagnosed with Glanzmann's, Thromboaesthenia.

KEYWORDS : epistaxis, Glanzmann's thromboaesthenia, platelet aggregration disorder

Introduction

Epistaxis is frequently encountered in ENT emergency. In cases of severe, intractable bleeding where no local cause can be found, an effort should be made to rule out haematalogical disorders. Glanzmann thromboaesthnia is one such rare cause of severe recurrent epistaxis [1]. In this condition the platelet count is normal or subnormal, the bleeding time is prolonged, and platelet aggregation is deficient or absent. It mostly presents as a bleeding disorder characterized by mucocutaneous hemorrhage of varying severity. Nasal bleed can be managed with anterior and/or posterior nasal packing.

Case Report

A 21 year old male presented to the ENT emergency of our hospital with profuse nasal bleed which was sudden in onset, with no history of antecedent trauma. The bleeding could not be stopped by nasal pinching and cold fomentation. The patient was admitted and vitals were recorded which were stable. Bilateral anterior nasal packing was done in an attempt to control the bleeding. However, the patient started bleeding again after only a few hours of anterior nasal packing. There was fresh post nasal bleed, which seemed significant. The anterior pack was removed in order to identify the point of bleeding but the bleeding was so profuse on removing the anterior pack that we immediately had to proceed with posterior nasal packing with a more securely inserted fresh anterior nasal pack. Patient was started on parenteral antibiotics along with hemostatic drug (trenaxamic acid) and other supportive treatment. Routine blood investigations revealed normal haemoglobin (12.7gm/dl), normal platelet count (2,89,000/ mm^3), PT (14 seconds), aPTT (28 seconds) and clotting time (9 seconds). However the bleeding time was prolonged. Detailed history from parents revealed one previous similar episode of nasal bleed in childhood. There was no other significant history suggestive of bleeding disorders. On further probing it was revealed that the patient's elder sister had expired due to excessive bleeding when she was four years of age. She had history of recurrent episodes of bleeding and had been diagnosed with GT at that time. This prompted us to focus our search for secondary systemic causes. As the coagulation studies already showed results within normal limits, the next step was to carry out platelet aggregation tests. The platelets failed to aggregrate in response to natural agonists e.g., adenosine diphosphate (ADP), collagen, thrombin, epinephrine and amino acids but showed aggregation with ristocetin, which was highly suggestive of GT. Patient was kept under observation for 72 hours. Both anterior and posterior nasal packs were removed after 3 days and the he was further kept under observation for a period of two days, during which there was no fresh episode of bleeding. Patient was discharged after a stay of five days in the hospital. He was discharged on topical decongestants and was adviced to follow up in case of an episode of recurrent bleed. The patient and his relatives were further counseled and were advised to visit the hematological clinic for any further course of action.

Discussion

Glanzmann's thrombasthenia is an autosomal recessive platelet abnormality characterised by a defective clot retraction and abnormal appearance on stained film. It has an incidence of about 1/1,000,000 and seen primarily in populations in which consanguineous marriage is common [2]. The gene for GPIIb-IIIa is present on chromosome number 17 in humans and is said to affect men and women equally [3].

GT results from either qualitative or quantitative abnormalities in the glycoprotein IIb/IIIa complex located on the platelet membrane. GPIIb/IIIa once activated will bind to one end of fibrinogen. Another platelet, with its own GPIIb/IIIa can then bind to the other end of the fibrinogen. This allows the formation of a large aggregation of bound platelets, commonly called a blood clot. In GT the GPIIb/IIIa complex is defective, the platelets can no longer bind with one another, and the blood clot fails to form [3].

It commonly presents in infancy or early childhood with multiple bruises and petechial spots following minimal or unrecognisable trauma. Large muscle bleeds and bleeding into joints are uncommon but menorrhagia can be severe in girls. Serious accidental or surgical trauma can be life threatening.

Epistaxis is the most common cause of severe bleeding in thromboaesthenia [2]. It is typically more severe in childhood[4]. Severe epistaxis is unusual in adult patients, and this may be the primary reason for the impression that the risk of bleeding in thrombasthenia decreases with age [2].

The essential diagnostic features are a normal platelet count and morphology and prolonged bleeding time, associated with a complete failure of platelet aggregation with all agents (ADP, adrenaline, thrombin and collagen) except von Willebrand factor. Having demonstrated the aggregation defect, the diagnosis can be confirmed by measuring the amounts of the platelet glycoproteins GpIIb and IIIa on the platelet surface membrane which are either deficient or functionally abnormal [2].

There is no known cure for GT. Platelet transfusion is the standard therapy. However, approximately 15-30% of patients become refractory to platelet transfusion or develop antibodies to GPIIb-IIIa and/or HLA antibodies [3]. Bone marrow transplants have been used successfully in rare cases [2]. Meanwhile, the management of GT consists of the avoidance of trauma and medicines such as aspirin, which may further affect platelet function, and the treatment of major bleeding episodes by platelet transfusion.

The role of the otolaryngologist is to control bleeding during major episodes of nasal hemorrhage that does not respond to medical management. These patients usually have remarkable improvement in the frequency and severity of epistaxis in adolescence and then require much less aggressive therapy [5]. The overall morbidity and mortality have been difficult to estimate due to its rarity, but in most studies the prognosis with proper supportive care has been found to be very good.

Conclusion

GT is a rare inherited bleeding disorder. Epistaxis is a rare manifestation which can present with severe recurrent bleeding. Supportive treatment is the best means for care, and nasal bleed may require nasal packing and platelet transfusions. With proper supportive care GT has a very good prognosis.

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