Hemarthrosis an Unusual Presentation of Glanzmann's thrombasthenia

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INTRODUCTION

Glanzmann thrombasthenia (GT) is an extremely rare autosomal recessive coagulopathy disease .It characterized by significant prolonged bleeding time and normal platelet count. The cause of disease is the quantitative or qualitative defect of the platelet glycoprotein IIb–IIIa. As a result, the aggregation processes via fibrinogen bridging of platelets to other platelets cannot occur properly. The disease characteristically presents as mucosal bleeding such as epistaxis, gum, dental extraction bleeding menorrhagia and increased bleeding post-operatively. Hemarthrosis usually associated with abnormalities of the plasma coagulation factors such as classic hemophilia.

AIM

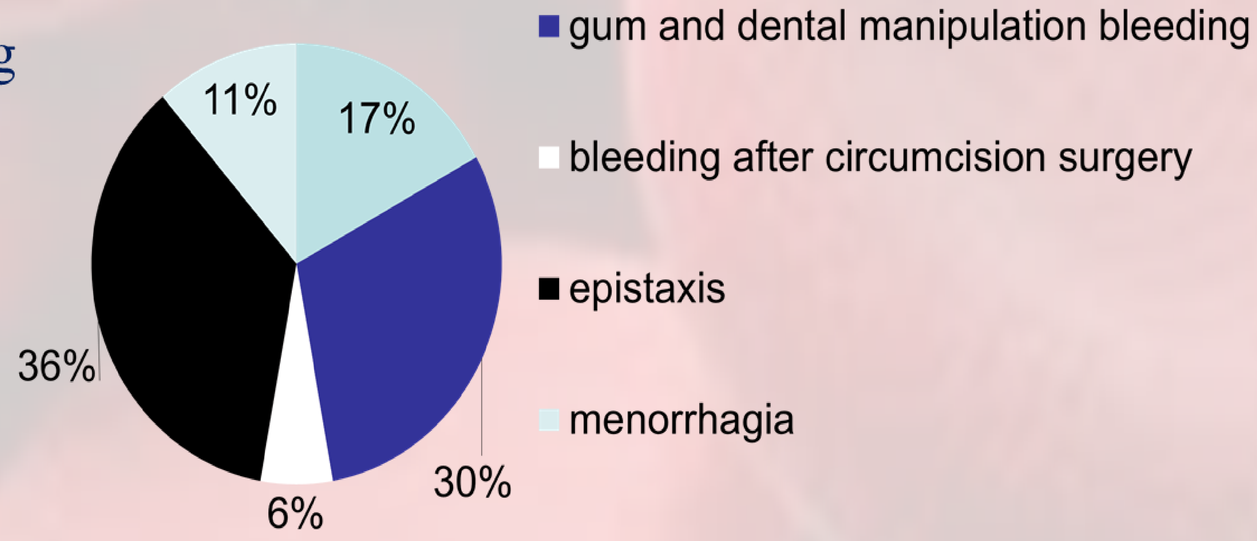
This study aims to present the hemarthrosis and other clinical manifestations of 53 patients with Glanzmann Thrombasthenia (GT) from south west Iran.

MATERIALS AND METHODS

The diagnosis was made in 53 (27 male, 26 female) patients in the range of one to 50 years old in the presence of a normal platelet count, prolonged bleeding time (BT), decreased or absent clot retraction, and absent platelet aggregation to ADP, epinephrine, collagen, and thrombin. A comprehensive evaluation such as clinical manifestations, and demographic data were recorded. This is a descriptive cross sectional study that was conducted at Ahvaz Jundishapur University of Medical Sciences in Khuzestan province. Data were collected by a questionnaire form. Statistical analysis was done by using Statistical Package for the Social Sciences version 17.

RESULTS

Hemarthrosis was the first presenting symptom among 9 cases (17%). 3 patients had recurrent hemarthrosis, hemophilic-like arthropathy. Target joint in 2 patients were the knee (1 patient with both knee involvement) and the elbow was the occurrence of another chronic arthropathy. The former patients were women and the latter was man. The other first usual clinical manifestations of this disorder among the 53 patients were shown in Figure.



CONCLUSIONS

Hemarthrosis is distinctly unusual presentation in GT. The review of literature is noted only few GT patients with hemarthrosis. Some occurred after major trauma but our patients had hemarthrosis after minor trauma and three of patients had repeated hemarthrosis. The optimal treatment of patients were done by single donor platelet transfusion and/or recombinant factor VIIa (Novo Seven).This brief report may have some clinical points for the clinicians and health sector providers.

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