Management of patients with Hemophilia A in the south of Tunisia

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Introduction

Hemophilia A is a rare inherited bleeding disorder resulting from factor VIII deficiency. Clinical features include repeated intra-articular and intramuscular bleeding. It is the most common subtype of hemophilia. The aim of our study was to evaluate the clinical characteristics and the treatment outcomes of patients with hemophilia (PWH) A in southern Tunisia.

Methods:

Our study concerned all PWH A followed in the hemophilia treatment centers of south Tunisia, during a period of 24 years (from January 1982 to September 2024). The epidemiological and clinical characteristics, prophylaxis state and patients complications were evaluated.

Results :

Among 155 PWH diagnosed during the study period, 126 (82%) had hemophilia A. The severe form represented the majority in 66% followed by moderate form in 24 % and minor form in 10%. The hemorrhagic symptoms during the evolution of our PWH were represented mainly by hemarthrosis (82%), hematomas (79%). Intramuscular hematomas on the psoas muscle occurred in 21% of cases. Visceral hemorrhages were noted in 27% of PWH mostly represented by hematuria. Intracranial hemorrhage was noted in 6 % of cases. Among severe hemophilia A patients, prophylaxis was administered in 60 patients (48%). Chronic hemophilic arthropathy concerned 38% of our patients. The knee and elbow were the most affected joints. Transfusion transmitted infection with HIV and hepatitis C was 1,5% and 21% respectively. Inhibitor positivity was found in 21% of cases and 52% are high responders. High responder patients were treated with recombinant factor VII in case of bleeding and 4 patients are under prophylaxis with emicizumab, all of them performed immune tolerance induction with failure in all the cases.

Conclusion

Hemophilia A constitutes 80-85% of all hemophilia cases in the literature which is similar to our serie. It is a serious disease causing repeated bleeding which can be severe enough to be life-threatening and require urgent and appropriate treatment. In the last few years, hemophilia care has improved significantly in our center, in fact, clotting factor VIII concentrates are becoming increasingly available for our patients. Currently, the standard-of-care for hemophilia A includes prophylaxis with factor VIII replacement therapy or emicizumab for high responder PWH A.