Brief Communication
http://mjiri.iums.ac.ir
Medical Journal of the Islamic Republic of Iran (MJIRI)
Iran University of Medical Sciences

Public Health Problems related to factor V deficiency in southeast of Iran

Majid Naderi[1], Shadi Tabibian[2], Akbar Dorgalaleh[3]
Zahra Kashani Kahtib[4], Shaban Alizadeh[5]

Received: 29 May 2013 Accepted: 2 Oct 2013 Published: 7 May 2014

Cite this article as: Naderi M, Tabibian Sh, Dorgalaleh A, Kashani Kahtib Z, Alizadeh Sh. Public Health Problems related to factor V deficiency in southeast of Iran. *Med J Islam Repub Iran* 2014 (7 May). Vol. 28:27.

Introduction

Factor V (FV) deficiency is a rare autosomal recessive hemorrhagic disorder with incidence of around one per million in general populations (1,3). The disorder results from low or unmeasurable level of FV antigen or activity. This disorder characterized by various bleeding features including mucosal tract bleeding, epistaxis, oral cavity hemorrhage, menorrhagia, bruising, hematomas and hemarthrose. This disorder mostly seen in some regions including Sistan and Baluchistan province in south east of Iran with high rate of consanguineous marriage. Factor V deficiency after Factor XIII deficiency has the highest prevalence in this province. It seems that the high frequency of the disease in the province is associated with different characteristics including ethnic, race and religion. Thus, the aim of this study is to evaluate the role of ethical, racial and other related characteristics of the area and its relation to the high prevalence of disease in this province. Nineteen patients with FV deficiency referred to Iranian Blood Transfusion Organization (IBTO) during 2011 To 2012 were included. The deficiency in each patient established by considering clinical manifestations, family history and prolonged coagulation tests including PT and PTT and the deficiency confirmed by PT-based FV assay. Factor VIII coagulant activity measured in order to distinguished isolated FV deficiency from combined FV and FVIII deficiency. Then each patient was interviewed by an expert staff to complete a questionnaire regarding demographic data and previous medical history, type of treatment, duration, therapeutic response, clinical manifestations and complications of the treatment .The mean age of patients was 23.65 years. Eleven (57. 9%) patients were men and 8 (42.11%) patients were women. Data is shown in Table 1.

The distribution of studied patients showed that the majority of patients (47.3%) were resident in Zahedan. Data is shown in Table 2.

According to the race of the patients, the

1. MD of Hematology and Oncology, Genetic Researcher Center in Non-Communicable Disease, Zahedan University of Medical Sciences, Zaheedan ,Iran, naderifactorxiii@yahoo.com
2. MSc of Hematology, Hematology Department, Allied Medical School, Tehran University of Medical Sciences, Tehran, Iran. Sh.tabibian@gmail.com
3. MSc of Hematology, Hematology Department, Allied Medical School, Tehran University of Medical Sciences, Tehran, Iran. dorgalaleh.1390@yahoo.com
4. MSc of Hematology, Hematology Department, Medicine School, Tarbiat Modares University, Tehran, Iran. zkashanikhatib@gmail.com
5. (**Corresponding author**), Assistant Professor of Hematology, PhD of Hematology, Hematology Department, Allied Medical School, Tehran University of Medical Sciences, Tehran, Iran. alizadehs@sina.tums.ac.ir

Table 1. Distribution of factor V deficiency according to age and sex

Age	Men	Women	Total
<10years	3(75%)	1(25%)	4(100%)
10-20	3(60%)	2(40%)	5(100%)
20-30	2(66.6%)	1(33.4%)	3(100%)
>30	3(42.8%)	4(57.1%)	7(100%)
Total	11(57.9%)	8(42.1%)	19(100%)

Table 2. Distribution of factor v deficiency according different cities of province

City	Number of patient	Relative percent
Zahedan	9	47.3%
Khash	3	15.7%
Saravan	3	15.7%
Zabol	1	5.7%
Dalgan	1	5.7%
Iranshahr	1	5.7%
Chahbahar	1	5.7%
Total	19	100%

majority of them were Fars (63.3%) and a minority was Baluch (36.8%).

A considerable number of patients' parents were close relative including unclegirl–cousin, cousin–cousin and cousin-girl of aunt (Fi.1).

This study is the first report in Sistan and Baluchistan province about FV deficiency and it was focused on social factors causing high rate of FV deficiency in this part of Iran. As we expected, in this study we found that consanguineous marriage in this province has a great impact on high prevalence of FV deficiency and more than 50 percent of patients are result of a close relative marriage. Relatively high percent (21% of patients were lower than 10years) of new cases of FV deficiency is of concern too. In fact this finding suggests that the tradition of consanguinity still continues in this area. Another outcome of this high rate of consanguineous marriages is prevalence of Rare Bleeding disorders in this area such as FXIII deficiency. First report of FXIII deficiency in 2004 revealed that FXIII deficiency has a considerable incidence in south east of Iran. In this study 46 patients with severe FXIII deficiency was diagnosed. But now in 2013 this province has more than 350 patients with severe FXIII deficiency; according to recently published reports, it has the highest prevalence of FXIII deficiency worldwide. This extremely high incidence of FXIII deficiency is the result of continuous traditionally consanguineous marriages and also absent of a prenatal diagnosis protocol and screening method for this factor deficiency. Since a considerable number of patients with factor V deficiency do not have any significant bleeding diathesis, it may not be clinically diagnosed. This in turn will cause an increase in patients with factor V deficiency. Moreover, carriers of factor V deficiency have no clinical presentation that will increase the number of heterozygote and therefore homozygote individuals.

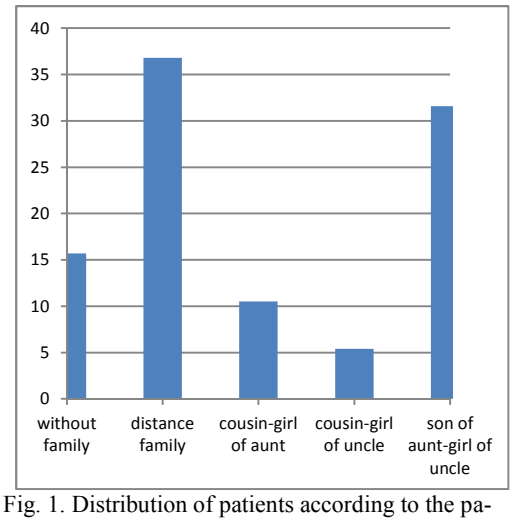

Fig. 1. Distribution of patients according to the patient's relativity

First step in management of RBDs is identification of main genetic disorder that can result in establishment of PND and screening tests for diagnosis of FV deficiency. With the establishment of an effective management strategy, further extension of disorder can be prevented. Such strategy has been used for factor XIII deficiency in this area (7).

Further extension of such diseases will increase the costs of health care services, prophylaxis treatment and also will induce psychological problems in the society. Therefore, introduction of an educational program to prevent the increase of consanguinity, carrier detection and prenatal diagnosis is necessary.

Acknowledgments

This research has been supported by Tehran University of Medical Sciences & health Services grant 25341.

Reference

1. Asselta R, Peyvandi F, editors. Factor V deficiency. Seminars in thrombosis and hemostasis; 2009: Thieme.

2. Lippi G, Favaloro EJ, Montagnana M, Manzato F, Guidi GC, Franchini M. Inherited and acquired factor V deficiency. Blood coagulation & fibrinolysis : an international journal in haemostasis and thrombosis. 2011;22(3):160-6. Epub 2011/01/20.

3. Huang JN, Koerper MA. Factor V deficiency: a concise review. Haemophilia. 2008;14(6):1164-9. Epub 2009/01/15.

4. Lak M, Sharifian R ,Peyvandi F, Mannucci F. Symptoms of inherited factor V deficiency in 35 Iranian patients. British journal of haematology. 1998;103:1067-9.

5. Naderi M, Eshghi P, Saneei Moghaddam E, Alizadeh S, Dorgalaleh A, Younesi M, et al. Safety of human blood products in rare bleeding disorders in southeast of Iran. Haemophilia. 2012.

6. Peyvandi F, Cattaneo M, Inbal A, De Moerloose P, Spreafico M. Rare bleeding disorders. Haemophilia. 2008;14:202-10.

7. Naderi M, Dorgalaleh A, Alizadeh S, Kazemi A, Tabibian S, Younesi MR. Assessment of relationship between CNS bleeding in factor XIII deficiency and Thrombin-Activatable Fibrinolysis Inhibitor polymorphism. Arak University of Medical Sciences Journal. 2013;16(7):0-0.