ACQUIRED HAEMOPHILIA AT CHRIS HANI BARAGWANATH ACADEMIC HOSPITAL

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INTRODUCTION

Acquired haemophilia is a rare bleeding diathesis caused by inhibitors/auto-antibodies to factor VIII, and is associated with significant morbidity and mortality. It typically presents with spontaneous soft tissue/muscle bleeds, associated with a prolonged partial thromboplastin time. More than half the patients have no underlying cause (idiopathic). Therapy is aimed at arrestting the bleeding (usually with the use of bypassing agents), treating the underlying cause (where detectable) and eradicating the auto-antibody with immunosuppressive therapy.[1-3]

PATIENTS AND METHODS

This study is a retrospective review of all adult patients with acquired haemophilia seen at Chris Hani Baragwanth Academic Hospital during the period 01/01/1996 to 31/12/2011.

NO.	F8 LEVEL %	GENDER	AGE	MAJOR SITES OF BLEEDING	UNDERLYING CONDITIONS	OUTCOME
1.	2	F	73	Soft tissue/muscle, haemarthrosis, mucosal	Idiopathic Developed Ca Breast 10 years after initial presentation	Died. Survival 128 months
2.	0	M	32	Haematuria, post-surgical abdominal wound site, haemarthrosis	HIV	Died. Survival <1 month
3.	0	F	57	Haemetemesis, soft tissue/muscle, haemarthrosis	SLE	Died. Survival 2 months
4.	1	M	49	Soft tissue/muscle	Rheumatoid arthritis	Alive, 19 months after diagnosis
5	2	F	82	Sof tissue/muscle, haemarthrosis, haematuria	Idiopathic	Alive, 5 months after diagnosis

RESULTS

There were 3 females and 2 males with a F;M ratio of 1.5:1. The mean age was 58 years, with a range of 32 to 82 years. A secondary cause was evident in 60% of the patients , while one of the two patients with no underklying cause (idiopathic), developed a malignancy 10 years after her initial presentation, bringing into question the possible association of malignancy, manifesting years after the initial diagnosis. The uncommon association of HIV with acquired haemophilia is noteworthy in the context of an area of high endemicity for the virus. The patiients were supported with blood products, given appropriate factor replacement (and more particularly FEIBA or Novo 7) and immunosuppressive therapy. 3 of the 5 patients have died(one with a survival of 128 months) and 2 are alive and well at present.

CONCLUSION

Acquired haemophilia is rarely encountered at our institution. It typically manifests in older patients with soft tissue/muscle bleeds as well as haemarthrosis and mucosal bleeding in our patients. Auto-immune disease is present in 40% of the individuals. HIV, despite being highly prevalent , is an uncommon cause. Therapy includes measures to arrest the bleeding (supportive measures and bypassing agents), immunosuppressive therapy and treatment of the underlying cause.

1. Collins PW and Percy CL. Advances in the understanding of acquired haemophilia A.: implications for clinical practice. British Journal of Haematology 148:183-194, 2009.
2. Franchini M and Lippi G. How I treat Acquired Factor VIII Inhibitors. Blood 112@):250-255, 2008.

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