



PHPWS
Rawalpindi/Islamabad

Intracranial bleeds in bleeding disorders: Northern Pakistan experience



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Introduction

Intracranial bleeds are a serious problem in bleeding disorders in the world resulting in increased morbidity and mortality. They are the most common cause of death in hemophilia. In Pakistan where consanguineous marriages are common, intracranial bleeds are also seen in rare bleeding disorders.

The Rawalpindi/Islamabad chapter of PHPWS has 658 registered patients of which 547 are males and 112 are females. 366 males (66.9%) have hemophilia A and 56 (10.2%) have hemophilia B. von Willebrand's Disease (vWD) is seen in 53 (9.7%) males and 54 (48.2%) females. Rare bleeding disorders are present in 49 (8.9%) males and 45 (40.2%) females. These patients are managed in collaboration with hemophilia treatment centre (HTC) at PIMS.

From Jan 2007 to Dec 2011 thirteen males presented with fifteen episodes of intracranial bleeds. This study was conducted to document risk factors and evaluate management in these cases.

Objectives

To study all patients presenting with intracranial bleed in relation to the bleeding disorder & its severity and to document risk factors, presenting features, diagnostic and management modalities and complications.



Methods

Data of all patients was recorded on a Proforma with details of age, sex, type & severity of bleeding disorder, presence of inhibitor & presenting symptoms & signs. Diagnostic investigations & treatment details were also documented. The patients were followed up to 2 years.

Tables

Table 1: Intracranial bleeds Age Distribution

Age (yrs)	No. (%)
< 5	2 (15.384)
5 – 10	8 (61.53)
11 – 20	2 (15.384)
More than 20	1 (7.692)

Table 2: Intracranial bleeds Type & Severity of Bleeding Disorder

Factor Deficiency	No.	Severity
Factor VIII	09	Severe
Factor IX	01	severe
vWD	01	severe
Factor XIII	02	severe

Table 3: Intracranial bleeds Presenting Symptoms

Presenting Symptoms	No. (%)
Headache	10 (76.92)
Vomiting	10 (76.92)
Fits	02 (15.384)
Irritability	01 (7.69)
Coma	02 (15.384)

Results

13 male patients presented with 15 episodes of intracranial bleeds. 9 patients were of Haemophilia A, 1 of haemophilia B, 1 patient had vWD & 2 with FXIII deficiency. All cases had intracranial bleed documented by CT scan except 1 patient who arrived in coma & died before any investigations & treatment.

Age & severity

The mean age was 7.3 years with a range of 4 months to 11 years. All patients had severe disease. Inhibitors were not present in any case.

Causes

History of head trauma was present in 12 of the 15 episodes (80%). This was secondary to fall from stairs in 10 patients (83.3%). 3 cases had no evidence of any form of trauma.

Symptoms

The most frequent presenting symptom was headache & vomiting present in 10 patients. 2 presented with fits, 1 patient was irritable and 2 were in coma on arrival.

Site

The bleeds were sub Dural in 5 & intra cerebral in 9 patients. The probable diagnosis of intracranial bleed was made by the history of trauma and symptoms of headache & vomiting and was confirmed by computerized tomography scanning in all patients except 1 who arrived in coma & died before any investigations could be carried out.

Recurrent bleeding

2 patients had repeat bleeds in the 1st year post the initial bleed.

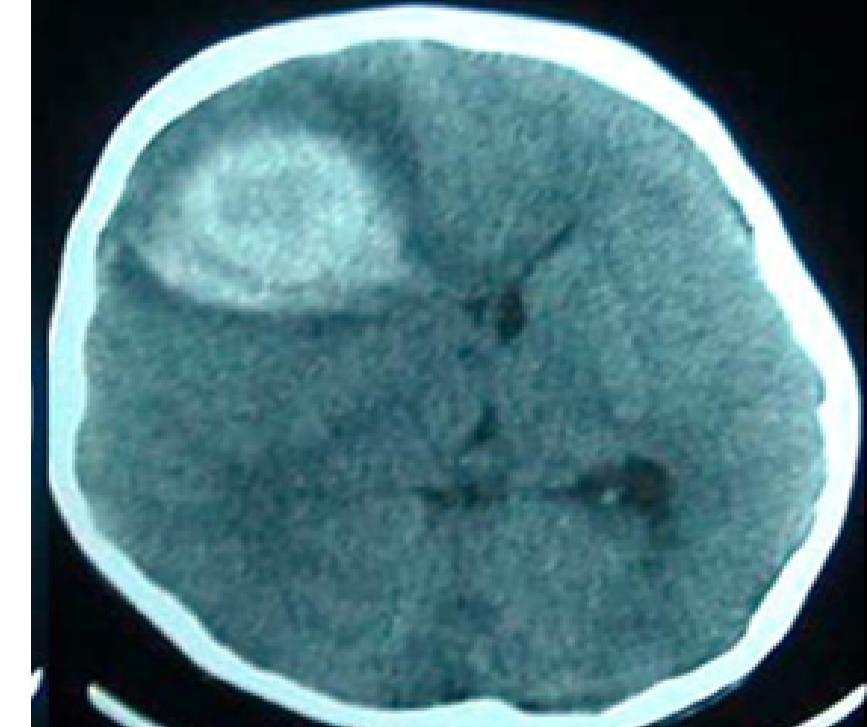
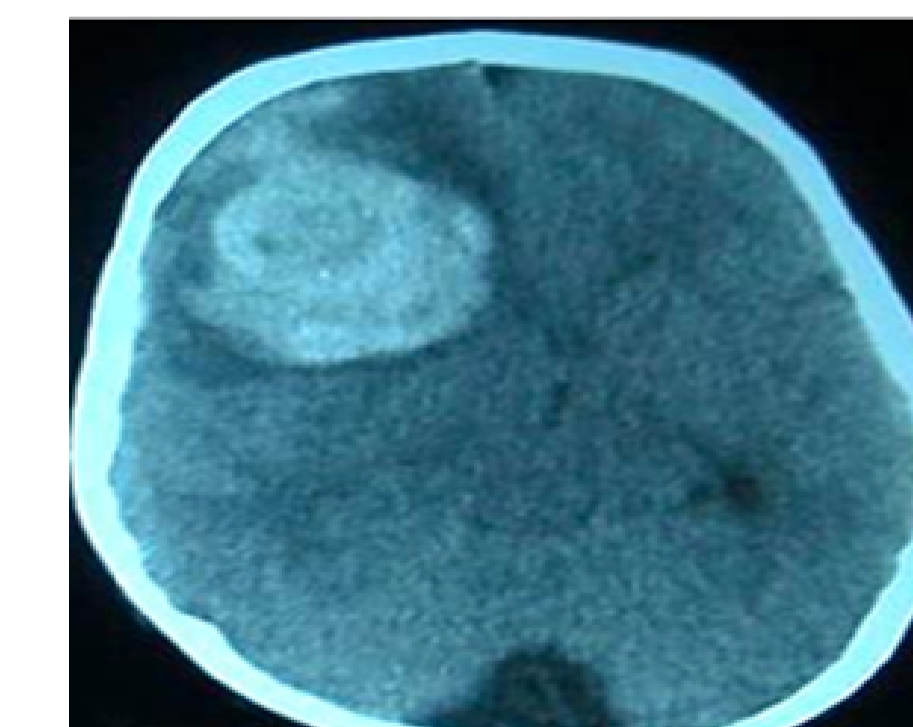
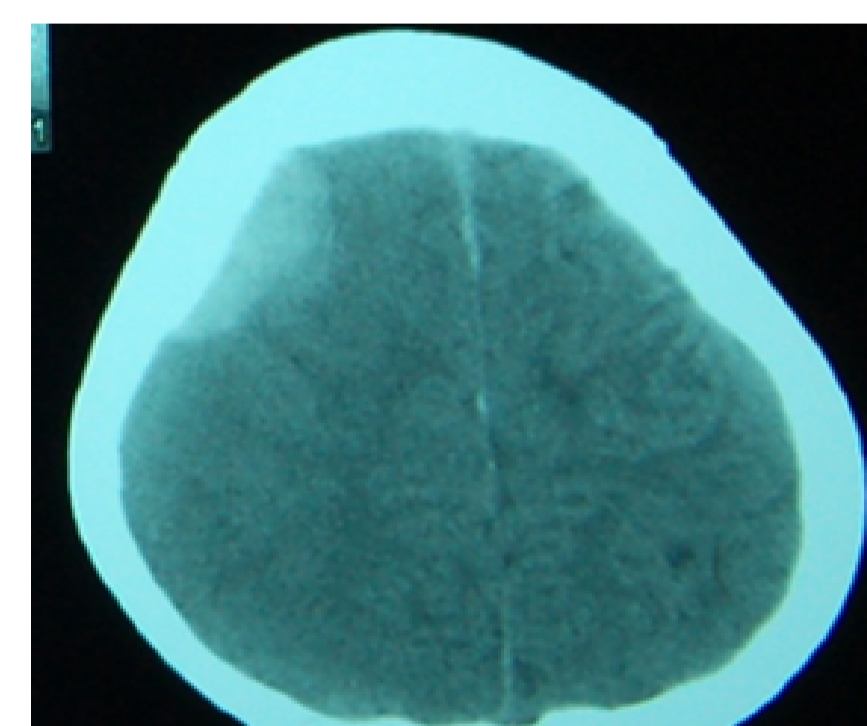
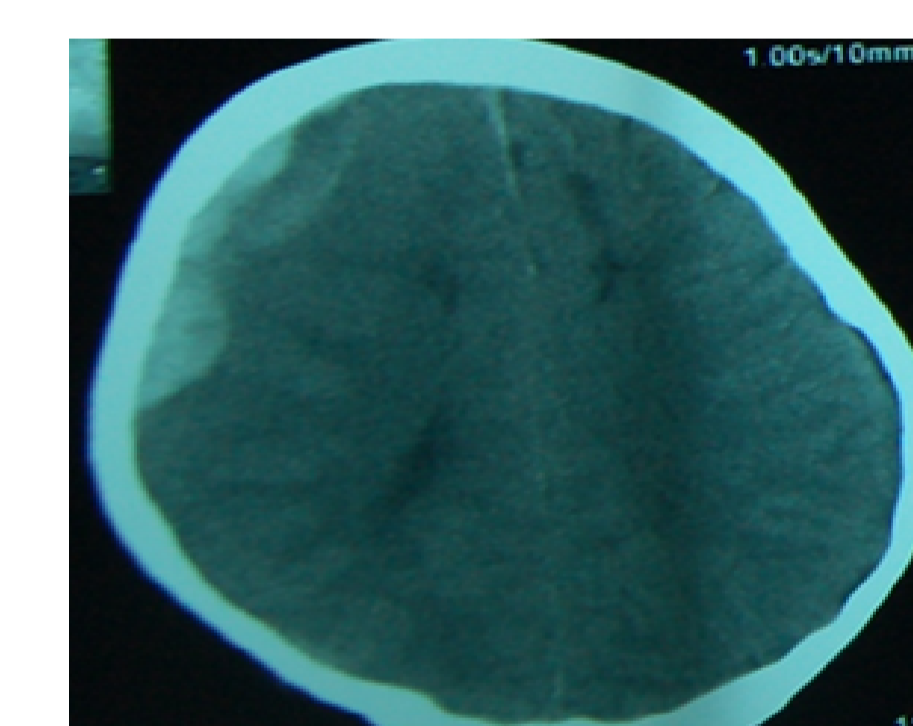
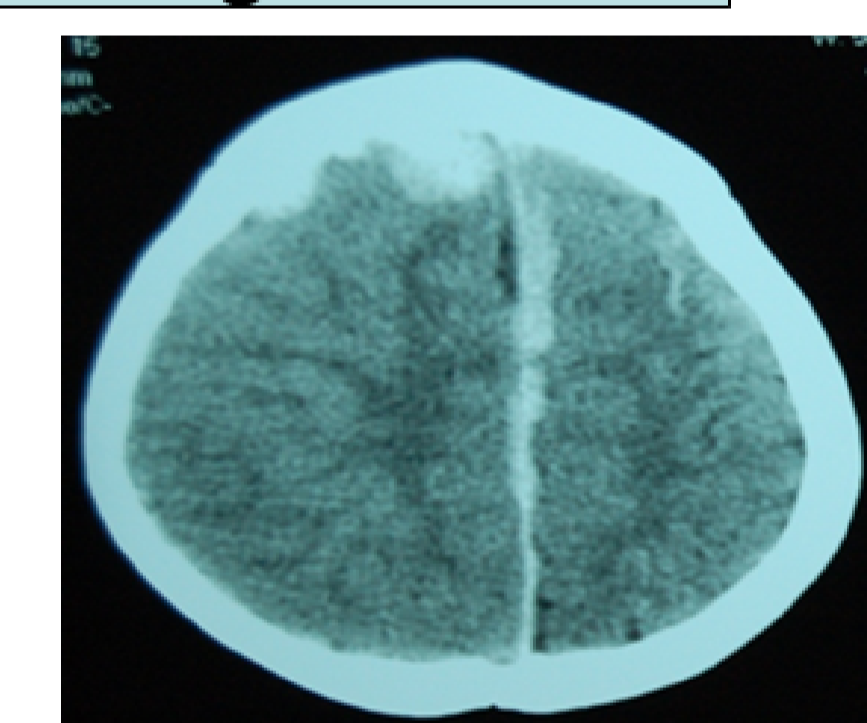
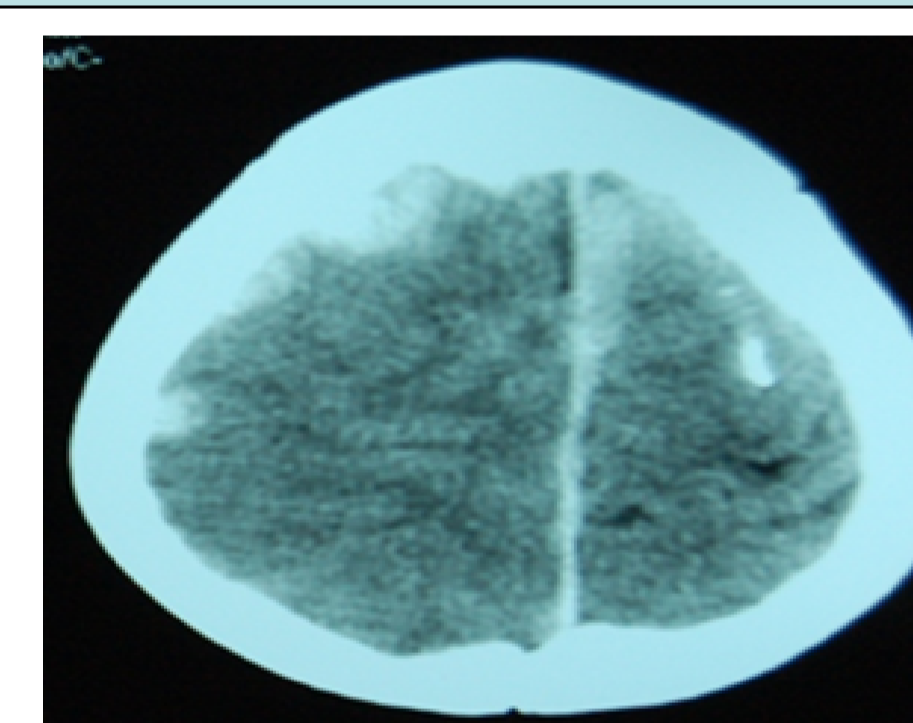
Mortality

There was complete resolution in 10 patients. The patient with vWD, 1 with FXIII deficiency and 1 with FVIII deficiency, who arrived in coma died.

Complications

1 patient developed epilepsy and required treatment with anticonvulsants for a year.

Intracranial bleeds CT Images



Conclusions

Intracranial bleeds are not uncommon in developing countries. Appropriate and adequate treatment is required for full recovery. There is need to create awareness about this serious complication of bleeding disorders and efforts should be made for provision of factor concentrates in all cases.

References:

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- Elaine Eyster M, Gill FM, Blatt PM et al. Central nervous system bleeding in hemophiliacs. Blood 1978 June;51(6):1179-1188.

N.B tables age & severity, presenting symptoms,

