A RARE CASE OF HEMOPHILIA AND INTRACRANIAL HAEMORRHAGE
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Abstract:
INTRACRANIAL HAEMORRHAGE IS A MORBID COMPLICATION IN PATIENTS WITH HEMOPHILIA. IT IS RARE TO PRESENT WITH SUBTLE SIGNS LIKE HEADACHE IN THE ABSENCE OF LOCALISING SIGNS OR LOSS OF CONSCIOUSNESS. HERE WE REPORT A CASE OF YOUNG ADULT PRESENTING WITH HEADACHE AND SPONTANEOUS INTRACEREBRAL HAEMORRHAGE. THIS CASE IS PRESENTED BECAUSE OF ITS RARITY, TO PRESENT AS HEADACHE ALONE AND TO EMPHASIZE THE GOOD PROGNOSIS OF SUBARACNOID HAEMORRHAGE AND SUBDURAL HAEMORRHAGE IN PATIENTS WITH HEMOPHILIA.

Keyword: HEMOPHILIA, SPONTANEOUS INTRACEREBRAL HAEMORRHAGE, HEADACHE

INTRODUCTION:
Haemophilia blood disorder known as the royal disease because it was common among Queen Victoria’s family. The disease can be hereditary or acquired. The hereditary disorder is a sex linked recessive disorder affecting males, females are silent carriers. In one third of the patients spontaneous mutations (deletion -5%, inversion -40%) were noted. Haemophilia A (factor-viii deficiency) is most common with the incidence of one in 10000 males births. Haemophilia B (deficiency of factor ix) occurs in 1 in 20000 to 34000 male births. The acquired disorder is due to the development of inhibitors to factor vili.

CASE HISTORY:
A 16 yr old male a known case of haemophilia A from 2 yrs of age born to a 3rd degree consanguineous parents with complaints of head ache and vomiting for 5 days. There was no history of trauma, loss of consciousness. There was no history of cranial nerve involvement, sensory or motor disturbance. There was also no history of bleeding elsewhere. His father is also a case of haemophilia A.

On examination- the patient was conscious and vitals were stable. The cardiovascular and respiratory system examination were normal. The examination the central nervous system was normal except for a mild neck stiffness. Fundus normal.

BEFORE TREATMENT (Interhemispherical bleed, tentorial bleed, left frontoparietal SDH, SAH)

CXR and ECG were normal. CT brain showed interhemispheric bleed, left frontoparietal subdural haemorrhage and subarachnoid haemorrhage. Diagnosis of haemophilia A with SDH AND SAH was made. Patients was treated conservatively with Recombinant factor vili 2000 units/day for 10 days along with anti convulsants and supportive measures. The patient improved symptomatically and there was no headache at the end of 15 days. Coagulation profile was normal. He was continued on prophylaxis (800 units three times a week). CT BRAIN repeated after 1 month was normal. The patient was followed for a period of 6 months. There was no history of seizures, motor deficits. Neurological examination done at 6 months were normal.
DISCUSSION

Haemophilia is a bleeding disorder that can cause a wide range of complications like persistent bleeding after trauma to spontaneous haemorrhage. ICH is the most dreaded complication with the incidence of 10% in haemophiliacs and the mortality of 30%. The risk of developing ICH is 2% per year [4,11]. Data from the UNIVERSAL DATA COLLECTION PROJECT of the US centers for disease control and prevention indicate that the predisposing factor for intracranial bleed are HIV infection, presence of inhibitors and age <5 yrs or > 51 yrs.

BRADLEYS NEUROLOGY IN CLINICAL PRACTICE (5TH EDITION) quotes that Haemophilia is a rare cause of ICH with a incidence of 2.5 to 6% of haemophilia developing it with half as intra cerebral and other as subdural, subarachnoid least common [12]. It can present as loss of consciousness brain damage and death. It can occur in the neonatal period (as a part of birth injury) in young adults(young stroke; secondary ICH);[10], and in adults. In most cases it occurs secondary to trivial trauma or sometimes spontaneous. Spontaneous CNS bleeding in individuals with haemophilia is rare except when there has been a recent antecedent CNS haemorrhage (i.e., a recurrent bleed at a previously injured site) or when there is an associated anatomic lesion that predisposes acute haemorrhage (e.g., aneurysm or AV malformation); [14]. It can be intracerebral, subdural, or epidural. Subarachnoid haemorrhage is the least common and carries a good prognosis [9,14]. Bleeding can also occur in the spinal cord (haematomyelia) but it is rare.

TREATMENT OF HAEMOPHILIA (NOV2007. ISSUE NO 43) by the WORLD FEDERATION FOR HAEMOPHILIA states intracranial haemorrhage is a acutely harmful to the tissue of the nervous system causing "space occupying effect". It is therefore essential to raise the factor levels even before bleeding is documented by imaging. The site of bleed can be subdural, epidural or intraparenchymal. All are capable of causing rapidly worsening CNS dysfunction and in extreme cases herniation and death. Because the degree of damage is dependent on volume and duration of bleed, bleed in surgically accessible sites should be removed fast (eg-SDH). When inhibitors are suspected bypass agents like factor via can be used. A retrospective review of the Haemophilia and Thrombosis Research Society 2004–2008 database was conducted to know the safety and efficacy of recombinant factor via (bypassing agents) in congenital haemophilia with inhibitors and concluded that rFVIIa was found to be safe and effective in treating CH with an efficacy rate of 100% for ECH and 82% of ICH [17].

A larger review of neonatal intracranial haemorrhage indicates an incidence of 3.58% [6]. Given that nearly one-third of new cases of severe haemophilia A represent new mutations (with no family history), all male newborns presenting with intracranial haemorrhage should have an activated partial thromboplastin time (aPTT) performed immediately and FVII and FIX assays performed as soon as possible [11]. Suspicion of intracranial haemorrhage is call for an immediate ultrasound of the brain through the fontanelle.[1,17]

Although ICH in haemophilia is not benign it was associated with significant cognitive deficit. Early neuropsychological assessment is indicated when there is a history of ICH [2]. But seizures, motor deficits, mental retardation, aphasia can occur (more with intraparenchymal bleed); [14]

CONCLUSION:
Intracranial haemorrhage is a morbid complication of Haemophilia. Early suspicion and prompt treatment with factor concentrates and surgery when appropriate will reduce the mortality and long term sequelae. Before the use of factor concentrates the mortality was more than 70% and is now reduced to 34% [9]. With the recent advances and the availability of extremely potent concentrates of factor viii, the outlook for long survival is good.

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Head injury in patients with haemophilia or suspected cases should be treated with recombinant factors to maintain a plasma level of 100IU/dl. Radiological procedures can be done while the factors are administered and once the diagnosis is confirmed the factors are administered to maintain a nadir level of more than 30IU/dl. Treatment should for 10 to 14 days and prophylaxis (15-20 IU/kg body weight 2-3 times per week to maintain a factor level of >1%) for at least 6 months [16] and the haemorrhage dealt accordingly.
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