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Case Report: An Infant with Severe Hemophilia

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Abstract

Hemophilia is a hereditary bleeding disorder in which a person lacks or has low levels of certain protein in the blood called "factors" and the blood does not clot. Hemophilia A–Deficiency of factor VIII, Hemophilia B –Deficiency of factor-IX, Hemophilia C-Deficiency of Factor XI. Males are affected more. Severity of the disease is classified as per the amount of clotting factors. We should link them with the hemophilia society or local support groups. (1,2 &3)

Key words: Clotting factor, Bleeding, & coagulation profile.

Introduction

A 7-month-old baby who is developmentally normal child brought with multiple firm swellings at different sites of body for one month,which appears spontaneously over multiple nonspecific sites,not localized to joints,associated with purplish skin discoloration over the swellings, resolves spontaneously in a week.

Clinical history

Present History

Associated with mild pain to touch present. No involvement of joints No history of epistaxis, melena, hematemesis. No history of mucosal bleeding, history of suggestive of petechiae. No history of Trauma, drug administration. No history of fever seizures, altered sensorium, weakness.

Past History-No similar history in the past

Family history- he is the first born out of non consanguineous marriage. No similar history in other family members in 3 generations

Development history Developmentally normal upto age

Immunization history Immunized upto age

Antenatal history-uneventful.

Natal/postnatal history: term/AGA/SVD/birth weight-2.9kg/no NICU stay/no bleeding manifestations at birth

Vitals

Temp afebrile; pulse-128/min;Resp.rate-28/min;spo2-98%on RA;CFT<3sec;BP-89/52mmHg(50-90p)

Anthropometry

Weight-8.5kg, Height-72cm Head circumference-43cm which indicates normal for age

Physical examination

On general examination child was alert, febrile, no pallor icterus/cyanosis/clubbing/pedal edema

4 hematomas, largest measuring 3x3 cm with ecchymosis over it on the right shoulder, right arm abdomen and chin.

P/A soft, hepatomegaly, spleen-2cm under RCM

Respiratory system; BAE, NVBS, No added sounds

CVS:s1 s2 No murmur

CNS-No FND

Investigations

Hb-10.3

TLC-15830

DLC-N19l75

PLT-3.04lakh

Ps-MCHC RBcs anisopoikilocytosis

Urea/creat-11/0.25

Na/K-136/4.12

STP/Alb-6.2/4.2

AST/ALT-39/23

T.Bil/d-Bil-0.52/0.09

ALP/GGT-446/8

PT/INR-13/1.06

aPTT-110.7

USD Abdomen and cranium-NAD, No c/o hematomas

PT/INR -13.3"/1.08

APTT-193"

TT-18.8"

Factor IX assay-<01% activity of normal

Imp-severe hemophilia

Course and treatment

7 month old child was admitted with the above complaints. On examination multiple hematoma were found, largest on their right shoulder, measuring 3x3 cm with ecchymosis over the skin.3 small hematomas were found on right arm, leftarm, abdomen and chin. Hemophilia was considered as a possible diagnosis. Hence coagulation profile was sent, which showed elevated aPTT with normal PPT. Fa assay showed severe factorIX deficiency (<1%).hemophilia B was diagnosed didn't have any new onset hematomas or echymosis during the hospital stay,thermodynamically stable.Condition of the child and precautions to be taken is explained to the mother (4).

Condition at Discharge

Child was alert, active, and afebrile

PR-128/min

BP-82/54mmhg

RR-26/min

P/A :soft,nohepatomegaly,spleen-2cm under LCM

CVS-S1 S2 heard,no murmur

RS-BAE present.No added sounds

CNS-No FND

Advice at Discharge

1. orals as accepted

2.Vit D 3 drops (1ml/400IU)1ml OD

3 To prevent trauma

4. Review in Paediatric OPD after 2weeks

5.Follow-up under paediatric haematology clinic

Care givers Stress

Baby's father working as agraphic designer and being a young mother found it difficult to accept the disease. Haemophilic is a chronic condition and there will be continuous stress in taking care of these children. Parents may find support groups such as the haemophilic society useful. It enables the parents to find support and find methods to cope with these children as the society organises talks and seminars to educate about haemophilia and organises meeting with similar haemophilia children. Hence these parents would be able to encourage one another and share tips on caring for haemophilic children.

Conclusion

It's often-neglected diagnosis missed out many a times. It is important to diagnose the disease early and treat appropriately.India is one among the 5 countries harbors the highest number of hemophilia cases. The other countries are USA, Brazil, China &UK. In India a reported number of patients with hemophilia A is 11,586 while estimated prevalence could be around 50,000 patients. The reasons of this less count could be, under diagnosis and incomplete mortality recording. Epidemiologists mentioned that actual prevalence could be 70,000 in India. Among union territories Puducherry is leading. (5)

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