CLINICAL GUIDE

Approach to a Patient of Bleeding Disorder

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Bleeding disorders form a sizeable number of patients in hematology practice. These disorders may be obvious or so subtle that they can be overlooked (1). The evaluation of patient with bleeding disorders must be made in a logical sequence so as to uncover the nature of bleeding disorders, thus facilitating the choice of laboratory studies. The disorders results from abnormality of vessel wall, platelets or coagulation factors (2). This paper reviews the approach to a patient suffering from or having any evidence of bleeding disorder. The evaluation of a patient with bleeding disorders can be divided into two parts.

1. Clinical Evaluation
   (a) History
   (b) Physical examination
   (c) Family History
2. Laboratory Evaluation
   (a) Screening tests
   (b) Specific tests

History

Bleeding history:- A carefully collected history provides the single most effective tool for determining the presence and significance of a bleeding disorder. A meticulous history is essentially necessary in two circumstances.

(a) When clinical finding or past medical history points to a disorder of homeostasis.

(b) In patients who do not show obvious homeostatic disorder but who are scheduled for major surgery.

Milder bleeding are common than severe ones and history provides most assistance when questions are probing and sufficiently detailed. It is essential to ascertain whether the bleeding disorders is congenital or acquired.

Clues to congenital disorders are :

(a) Excessive bleeding initiated by common childhood trauma eg. tooth extraction.

(b) History of bleeding disorder in family and when mode of inheritance can be determined, it may suggest specific diagnosis as well.eg. X-linked in hemophilia A & B. Autosomal inheritance with low factor VIII C level suggests Von Willebrand's disease (3).

Clues to acquired disorders are :

(a) Patients giving history of tolerating stress to homeostatic system. e g., tooth extraction but presenting with a bleeding tendency in recent times.

(b) Exposure to potentially causative factors and onset of bleeding episodes.

Nature of bleeding

(a) Sites of bleeding may suggest where in the coagulation ,the defect may be seen e.g. :
   (i) Mucous membrane bleeding and petechiae are seen in platelet disorders.
   (ii) Haemarthrosis is common in Hemophila.
   (iii) Soft tissue hematoma without petechie, a mucous membrane bleeding suggest a defect in Primary homeostasis.

(b) Severity:

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(i) Spontaneous bleeding usually, is seen in severe bleeding disorders.

(ii) History of bleeding only after major trauma or surgery suggests a mild bleeding disorder (4).

(c) Timing of episodes:

(i) Bleeding uncontrolled from the onset suggests a defect in primary homeostasis.

(ii) Bleeding after apparent initial homeostasis are consistent with factor deficiency. A marked delay in bleeding after the initiating event is sometimes seen in factor XIII deficiency.

Medical History

Review of all medical conditions and their treatment can provide insight into possible nature of bleeding disorders e.g. :

(a) Vitamin K deficiency seen in malnourished, hospitalized patients on multiple antibiotics.

(b) Immune mediated thrombocytopenia seen in association with Systemic lupus erythematosus (SLE) and HIV infection.

(c) Disseminated Intravascular coagulation (DIC) is most commonly seen in septicemia.

Medications

Careful review of all the medications is critical in evaluating a coagulation abnormality.

(1) Aspirin leading to platelet abnormality.

(2) Incidental Heparin used for flushing Intravenous access lines.

(3) Extensive use of drugs known to induce thrombocytopenia.

(4) Acquired factor inhibitors associated with medications like penicillin.

General Questions

Should seek information about :

(a) Previous episodes of abnormal bleeding disorder or bruising.

(b) History of diseases associated with abnormal hemostasis eg, Liver diseases, uremia, SLE, and Malignancy.

(c) All medicines used.

(d) Family history of abnormal bleeding.

(e) Excessive bleeding from multiple sites suggestive of systemic homeostasis than bleeding from a site.

Specific Questions

(a) Bleeding from umbilical stump- typical of factor XIII deficiency.

(b) Bleeding from circumcision common in severe hemophilia.

(c) Nose bleeding - Infrequent but prolonged suggest systemic abnormality.

(d) Mennorrhagia.

(e) Extensive prolonged bleeding from minor cuts.

(f) Bruising.

Ask : (1) How often?

(2) Whether bruises without obvious trauma?

(3) Or appear at inoculation or intramuscular injection sites.

(4) Bleeding after dental extraction.

(5) Bleeding after major surgery or delivery.

(6) Need for blood transfusion.

Physical Examination

A detailed physical examination may provide valuable clue as to where in the coagulation system, the abnormality is likely to be present. e.g. Peteche - found with abnormality of platelet number and function. Mucous membrane bleeding points to coagulation defect but when mucous membranes are the only bleeding sites, thrombocytopenia, platelet function disorder or Von Willebrand's Disease is likely. Hemarthrosis always seen with severe form of hemophila and can occur in severe acquired deficiency as well. Diffuse bleeding from multiple sites seen in severe combined coagulation defect or severe liver disease. Abnormal elasticity of skin or hyperextensibility of joints should be sought as evidence of an hereditary connective tissue disorder associated with vascular bleeding (5). Telengiectasia should be noted.

Laboratory Studies : are guided by the history and physical finding in a patient .However, when no clues are available, a battery of appropriate tests often is the most expedient approach.

Platelet tests

(a) Platelet count- if low, confirm on peripheral smear.

(b) Platelet function.

(c) Bleeding time.

(d) Platelet aggregation with ADP, epinephrine or collagen and with ristocetin will unravel thromboasthenia, defects of arachidonic acid metabolism or storage pool disease or Von Willebrand's disease and Bernard-Soulier disease respectively.

Coagulation tests

Screening tests of coagulation like:

Prothrombin time (PT)

Partial thromboplastin time (PTT).

Thrombin time (TT).

Specific tests

Factor assays would uncover abnormalities of coagulation factor deficiency.

Management

Correct management is unlikely if exact diagnosis is unknown.

Local measures

Local hemostatic agents application like :

Thrombin gel.

Fibrin glue.

Haemolok.

Local pressure and stitching of the wound can also be used.

Specific measures include : (6,7)

Vitamin K.

Platelet transfution.

Fresh frozen plasma (FFP)

Cryoprecipitate / DDAVP (Desmopressin)

Factor concentrate.

Conclusion

As in other disciplines of medicine, prevention is key to success and any patient who is suffering from any bleeding disorder should be educated accordingly. A detailed history coupled with thorough clinical examination would help to order a panel of Screening tests and the results would guide further investigation and the nature and amount of therapy needed.

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