

Dermatological manifestations of hemophilia

Opinion

Hemophilia is a coagulation disorder due to deficiency or deficient activity of clotting factor VIII or IX. Congenital Hemophilia is an X-linked Recessive disorder due mutation in F8 gene in case of classic. Hemophilia or Hemophilia A and F9 gene in case of Hemophilia B. Acquired Hemophilia develops due to antibodies to factor VIII or IX. Both the variants have no clinical difference. The disease can be classified according to residual factor activity as severe (<1%), moderate (1-5%), or mild (6-30%) in cases with activity >25% of normal the disease is usually asymptomatic.

The patient of hemophilia can present to the dermatologist with recurrent bruises all over the body especially on trauma prone areas. Bruises or ecchymoses are large extravasations of blood usually associated with trauma or coagulation disorders. A hemophiliac can present with hemorrhagic subcutaneous nodules which may be the first presenting sign in many patients.¹ The colour of bruise can help in approximately assessing the duration of bruise. It can also be affected by patient factors like age, sex, skin colour and site of body involved. For instance, in older patients due to low elasticity, on cheeks and groin due to more vascularity and in fairer patients the bruise will be prominent. It can also be affected by the type of trauma, amount of bleeding and the time elapsed since the occurrence of bruise. The usual temporal sequence of colour change in bruise is; reddish to bluish-purple in first 2-3days, turns green within a week and yellow between 1-2weeks due to hemosiderin deposit.

The various differential diagnosis to be ruled out in the patient will be child abuse (age and history may help), drug, Munchenchausen syndrome, anticoagulant over dosage or usage in few cases. Other causes of non - palpable purpura must always be ruled out. Acquiring of hemophilia like features in adult or old age should alert the dermatologist to the possibility of acquired hemophilia. The patient can

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be diagnosed on the basis of associated clinical features like recurrent hemarthroses, hematomas and gingival leading. Factor VIII or Factor IX assay should be done. The residual activity of the factors should also be checked. Platelet counts and function should also be checked to rule out purpura. The patient can be symptomatically managed with cold compresses in initial stages and warm compresses in later stages of the bruise. The patient should be counseled to wear padded clothing and avoid trauma. Cryoprecipitate and factor infusions can be given in severe disease or in case of stress.

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Conflict of interest

The author declares no conflict of interest.

References

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