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TYPE 2M VON WILLEBRAND DISEASE IN A 6-YEAR-OLD BOY: A RARE FORM REVEALED BY RECURRENT EPISTAXIS

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Résumé

We report a rare case of type 2M von Willebrand disease in a 6-year-old boy, initially misdiagnosed as immune thrombocytopenic purpura. Recurrent epistaxis and a positive family history prompted further investigations, which confirmed the diagnosis. This case underscores the need to consider inherited bleeding disorders in children with unexplained mucocutaneous bleeding.

Keywords: von Willebrand disease, type 2M, epistaxis, pediatrics, platelet function

1-INTRODUCTION

Von Willebrand disease (VWD) is the most common inherited bleeding disorder. The type 2M form, a qualitative defect of von Willebrand factor (VWF) affecting its binding to platelets, is rare in children. We report a case of type 2M VWD revealed by recurrent epistaxis in a child initially misdiagnosed as immune thrombocytopenic purpura (ITP).

2-MATERIALS AND METHODS

A 6-year-old boy with recurrent nosebleeds and a family history of epistaxis underwent basic and specialized coagulation tests due to persistent symptoms.

Figure 1: child with epistaxis

3-RESULTS AND DISCUSSION

Initial results showed anemia (Hb: 7.4 g/dL), thrombocytopenia (68,000/mm³), and a peripheral smear with microcytosis and hypogranular small platelets. Coagulation tests revealed a TQ of 15.3 sec, PT at 80%, aPTT at 35.4 sec, and fibrinogen at 1.83 g/L. VWF:RCo was 44%, VWF:Ag was normal, with a RCo/Ag ratio of 0.45, suggesting type 2M VWD. Platelet aggregation was normal except for decreased response to ristocetin, indicating secretory-type platelet dysfunction.

The child was initially managed as ITP. The persistence of bleeding led to further investigations that confirmed type 2M VWD. Management included iron supplementation, an emergency care card, and therapeutic strategies such as platelet transfusion, nasal packing, desmopressin, and consideration of VWF concentrate prophylaxis in case of severe bleeding or surgery.

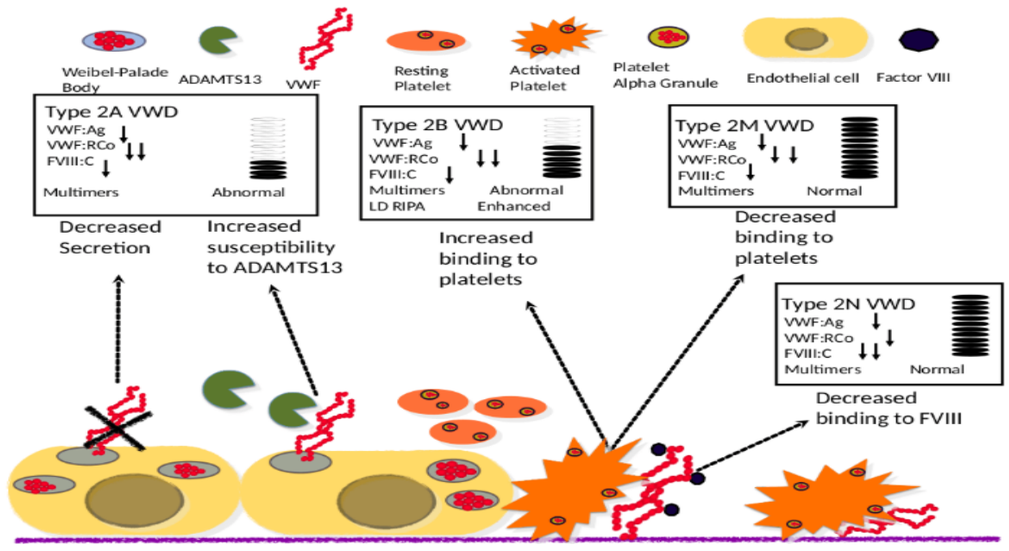

Figure 2: Qualitative defects of von Willebrand factor in VWD type 2

4-CONCLUSION

Type 2M von Willebrand disease should be considered in children with unexplained bleeding and a family history, as accurate diagnosis is key to appropriate treatment and long-term care.

Références bibliographiques
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