PAEDIATRIC DERMATOLOGY

VON WILLEBRAND DISEASE AND PROGRESSIVE PIGMENTARY PURPURA : A RARE COINCIDENCE

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Background: Von Willebrand disease (VWD) is the most common inherited bleeding disorder. It results from a quantitative or qualitative deficiency of von Willebrand factor (VWF) – a large multimeric protein which is required for platelet adhesion and serves as a factor VIII (FVIII) carrier. We describe a case of a 13 year old boy with one year of history of episodic petechiae on the lower extremities revealing VWD type 1.

Observation: A 13-year-old boy presented with a 12 month history of episodic asymptomatic reddish-brown hyperpigmented macules and patches on the ankle region and the dorsum of feet bilaterally which appeared spontaneously. Early lesions showed coalescence of numerous petechiae into reddish macules. Later lesions showed only brownish hyperpigmentation. There was no pruritus. No lesions were palpable and mucous membranes were unaffected. He had been previously healthy and had taken no medications. Physical examination was unremarkable except for the skin findings. Family history was positive for bleeding disorder. Laboratory studies showed a normal platelet count and prothrombin time (PT) but a prolongation of the activated partial thromboplastin time (aPTT). Further analysis revealed a low factor VIII level of 20% (normal 60-150%), he had a low ristocetin cofactor activity of 14% (normal 50-150%), consistent with the diagnosis of VWD.

Key message: Willebrand disease is a constitutional haemorrhagic disease of autosomal dominant transmission. Mucocutaneous or postoperative haemorrhages are readily revealing. The revelation of the disease by a pigmented purpura (as was the case of our patient) is exceptional.