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Retiform Purpura Associated With Protein S Deficiency☆

Púrpura retiforme asociada a déficit de proteína S

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A 67-year-old woman with a past history of hypertension and ischemic stroke presented reticulated purpuric lesions with a base covered by necrotic slough (Fig. 1A). The lesions had appeared on both legs over a period of 3 weeks. There was no history of catheterization. The lesions were clinically compatible with retiform purpura, and a number of tests were performed based on the fact that this condition can be secondary to vascular occlusion or a lesion of the blood vessel walls: blood tests including autoimmunity, thrombophilia studies, and phosphorus and calcium metabolism; skin biopsy; transthoracic echocardiography; and blood cultures. Biopsy revealed vascular thrombosis and occlusion with secondary dermoepidermal ischemia, with no signs of vasculitis. The only abnormal finding in the other tests was a low free and functional protein S, suggesting a type I or type III hereditary deficit. During her admission, the lesions ulcerated and erythema developed, associated with increased local temperature and an exudate (Fig. 1B). Antibiotic therapy and heparin anticoagulation were therefore started, with a good response and a progressive improvement. Protein S acts as an antithrombotic factor, inhibiting factors Va and VIIIa; its deficit therefore produces a state of hypercoagulability. Given the high probability of hereditary thrombophilia, we discharged our patient on anticoagulant treatment with acenocoumarol to prevent further thrombotic episodes and undertook study of her family, which excluded further cases among her relatives. We draw attention to the importance of identifying retiform purpura in order to diagnose the underlying cause and start treatment to prevent potentially serious consequences.

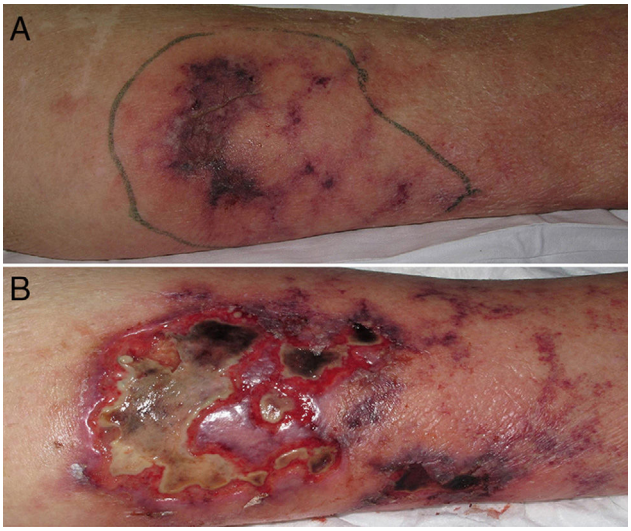

Figure 1

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