A man with two syndromes

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Received: 30 January 2014 / Accepted: 13 February 2014 / Published online: 27 February 2014
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A 40-year-old man was referred to the emergency department because of a low platelet count on a routine complete blood count (CBC). He had an hemangioma on his forearm together with a “congenital deformity” with no other medical problem including hematologic disorder in the past. On physical examination he had an evident hypertrophy of the right side of the trunk and the right upper limb. There was a biopsy mark on his right forearm (Panel A). He had no other vascular or hypertrophic changes in the remainder of his body parts. The physical examination was otherwise normal including the neurologic assessment. The family history was negative. On laboratory study, the platelets were 77,000/ μ L (Normal 150,000–450,000/ μ L) hemoglobin 12.0 g/dL (Normal 13.5–17.5 g/dL), and white blood cells (WBC) 8.1×10^3 / μ L (Normal 3,500–10,500/ μ L). Coagulation profile showed prothrombin time (PT) 11 s (Normal 9–12 s), activated partial thromboplastin time (aPTT) 30 s (Normal 29.0–36 s), D-dimer 370 μ g/L (Normal 0–450 μ gm/L), and fibrinogen 1.9 g/L (Normal 1.5–4 g/L). The other laboratory findings were within normal limits.

The patient presentation is consistent with the diagnosis of the Kasabach-Merritt syndrome in Klippel-Trenaunay syndrome (KTS). KTS is defined as a triad of capillary malformations, congenital varicose veins, and bony and soft tissue hypertrophies. Although most patients demonstrate all three signs of the clinical syndrome, in a case series, 37 % had two of them. Hemangioma, which can be evaluated by Doppler ultrasonography, is seen in 98 % of patients, followed by venous malformations and limb hypertrophy. The hemangioma is typically of the nevus flammeus type, but cavernous hemangiomas or lymphangiomas may also occur [1]. Hypertrophy may be appreciated at birth. It usually progresses during the first years of life. KTS generally affects a single extremity, although cases of multiple affected limbs have been reported. The

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leg is the most common site followed by the arms, the trunk, and rarely the head and the neck [2].

Kasabach-Merritt syndrome is thrombocytopenia and consumptive coagulopathy associated with hemangiomas [3]. The mechanism of thrombocytopenia is unknown, but it is proposed that abnormal endothelial cells within the hemangioma destroy platelets with activation of the coagulation cascade [4]. The management involves hastening hemangioma regression, interfering with platelet trapping within the lesion, and supporting the patient with transfusions. Embolization, radiotherapy, corticosteroids, and alpha interferon have been successfully used in many patients [5]. Since the thrombocytopenia was mild, the patient was discharged with low-dose steroid, and followed up uneventfully.

Conflict of interest None.

References

1. Fishman SJ, Mulliken JB (1993) Hemangiomas and vascular malformations of infancy and childhood. *Pediatr Clin North Am* 40(6):1177
2. Akcali C et al (2008) A case of Klippel-Trenaunay syndrome involving only upper limbs. *Giornale italiano di dermatologia e venereologia: organo ufficiale, Societa italiana di dermatologia e sifilografia* 143(4):267–269
3. Kasabach HH, Merritt KK (1940) Capillary hemangioma with extensive purpura: report of a case. *Am J Dis Child* 59(5):1063–1070
4. Hillman RS, Phillips LL (1967) Clotting-fibrinolysis in a cavernous hemangioma. *Arch Pediatr Adolesc Med* 113(6):649
5. Akyüz C et al (2003) Successful treatment with interferon alfa in infiltrating angiolipoma: a case presenting with Kasabach-Merritt syndrome. *Arch Dis Child* 88(1):67–68