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LETTER TO THE EDITOR

In reply: extensive venous malformation: an alternative diagnosis to Klippel–Trénaunay syndrome

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In reply:

We thank Dr. Alomari for his interest in our recent report [1]. The alternative diagnosis of extensive venous malformation that he proposes for our patient is certainly quite plausible. Our patient's original diagnosis of Klippel–Trénaunay syndrome was established during infancy based on the classic triad of capillary, lymphatic, and venous malformations in an enlarged lower extremity [2]. The patient received much of her continuing medical care at the Mayo Clinic and our affiliated children's hospital, where her consultants concurred with the original diagnosis. The patient subsequently developed additional vascular malformations as described over the intervening two decades after her original diagnosis was made. Her hematology team established the presumptive diagnosis of Kasabach–Merritt syndrome after the patient sustained an intracerebral hemorrhage during infancy. The patient also suffered from chronic intermittent lower gastrointestinal hemorrhage as a consequence of the presence of colonic vascular malformations and this chronic coagulopathy. As Dr. Alomari correctly notes, Kasabach–Merritt syndrome may be associated with Kaposiform hemangioendothelioma and tufted angioma [3], but Kasabach–Merritt syndrome may affect nearly one-half of patients with Klippel–Trénaunay syndrome independently of these specific pathologic abnormalities [4]. Thus, we feel confident that her original diagnosis of Klippel–Trénaunay syndrome complicated by chronic Kasabach–Merritt coagulopathy was correct. We specifically asked our patient for permission to photograph her peripheral and upper airway vascular malformations, but she adamantly refused because other physicians and health care providers had repeatedly photographed her in the past and she was very self-conscious about her appearance. Indeed, concerns about body image are very common in patients with Klippel–Trénaunay syndrome [5], and we accordingly respected her wishes. Regardless of the original diagnosis, we chose to present this difficult case because of its specific challenges, including the presence of vascular malformations in the upper airway, the potential for neuraxial vascular malformations within the dermatomal distribution of cutaneous lesions, and the presence of a chronic coagulopathy affecting both anesthetic and surgical management.

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