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LETTER TO THE EDITOR

Extensive venous malformation: an alternative diagnosis to Klippel–Trenaunay syndrome

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To the Editor:

I read with interest the case report by Holak and Pagel [1] in which the authors presented a young adult female patient with "cavernous hemangiomas" of the face, neck, chest, upper and lower extremities, and colon. In addition, the patient demonstrated signs of airway obstruction and lower gastrointestinal bleeding. Laboratory findings included mild anemia, leukopenia, mild thrombocytopenia (platelet count = 122×10^9/L), slightly elevated prothrombin time, severe hypofibrinogenemia and elevated fibrin degradation products. A chest radiograph demonstrated multiple phlebolith. The authors concluded that these features are compatible with Klippel–Trenaunay syndrome (KTS) associated with Kasabach–Merritt coagulopathy. Reviewing the constellation of clinical, laboratory and imaging features documented by the authors, I disagree with the diagnosis given to this patient for the following reasons:

1. The vascular anomalies in KTS are of the slow-flow type, including: (i) capillary malformation (port wine stain), (ii) lymphatic malformation, and (iii) venous malformations (CLVM) in an overgrown limb [2]. No clinical, photographic, or imaging documentation was provided by the authors regarding the presence of any capillary or lymphatic malformations. Overgrowth of the extremity in KTS is predominantly extrafascial and composed mainly of subcutaneous fatty tissue. This classic feature was not noted in this patient.
2. Unfortunately, the terminology used in the published literature on vascular anomalies is often poorly standardized [3]. The arbitrary use of certain terms (such as hemangioma; a benign tumor) as a generic diagnosis referring to a large group of heterogeneous vascular malformations and tumors creates an unnecessary diagnostic confusion. The authors referred to the multiple vascular lesions as "cavernous hemangiomas." Using the simplified binary classification proposed by Mulliken and Glowacki [4] and adopted by the International Society for the Study of Vascular Anomalies (ISSVA), these lesions should appropriately be called "venous malformations." In this classification, "infantile hemangioma" strictly refers to the common benign vascular tumors of infancy.

   In contrast to the mass-like lesions described by the authors, venous anomalies in KTS are of the "phlebectasia" type, such as the marginal venous system (anatomically related to the capillary stain), pelvic phlebectasia, and megacava.
3. Kasabach–Merritt phenomenon (KMP) is a specific consumptive coagulopathy with severe thrombocytopenia and hypofibrinogenemia typically associated with kaposiform hemangioendothelioma and tufted angioma [5]. The coagulopathic abnormalities described in the case report are likely to represent a milder form of consumptive coagulopathy known as localized intravascular coagulation (LIC) commonly encountered in patients with large venous malformations. LIC is characterized by elevated D-dimers, hypofibrinogenemia, and normal or occasionally slightly decreased platelet count [6]. The natural history and management of these two distinct forms of coagulopathy are very different.

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4. The rare combination of multiple, often numerous, focal venous malformations affecting the soft tissue and the gastrointestinal tract resulting in chronic gastrointestinal bleeding and anemia is known as blue rubber bleb nevus syndrome, or Bean syndrome. This disorder should be considered in the differential diagnosis of the reported case.

In their introduction to the case report, the authors correctly warned against "imprecise clinical definitions" of apparently related but distinct vascular anomalies, which may lead to the use of incorrect terminology and potential mistreatment. However, I believe that the diagnosis and the terms used in this case report are yet another example of such nosologic and diagnostic imprecision.

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