Infantile Hemangioma or Kaposiform Hemangioendothelioma?

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Editor:

We read with interest the letter sent by Yoon et al (1) about transarterial embolization of a cervicofacial hemangioma associated with Kasabach–Merritt (KM) syndrome in a premature neonate. There are some facets of the letter that warrant further comments, namely the association of the KM phenomenon with vascular tumors and the use of proper consensus-approved terminology adopted by the International Society for the Study of Vascular Anomalies.

The authors reported a KM phenomenon associated with a tumor assumed to be an infantile hemangioma by pathologic diagnosis obtained from a punch biopsy (histologic images not shown). However, the clinical data reported and the figure illustrations are more likely of a kaposiform hemangioendothelioma (KHE) rather than an infantile hemangioma. KHE is congenital in 60% of cases (2) whereas infantile hemangiomas are not fully developed at birth. KHE manifests as an infiltrating soft-tissue tumor accompanied by purple skin discoloration (3), as illustrated in figure 1 of the letter of Yoon et al (1). At magnetic resonance imaging, KHE is a moderately intense T2-weighted tumor with ill-defined borders infiltrating the surrounding fat tissue and often extending into adjacent muscles (3), similarly to what is described in the letter and shown in figure 2 by Yoon et al (1). In contradistinction, infantile hemangiomas do not tend to infiltrate surrounding tissues.

Infantile hemangiomas and KHE present different structures at pathologic examination. The latter has a lobular architecture composed of endothelial cells forming vessels of variable size and peripheral spindle cells positive on D2-40 and negative on GLUT1 immunostaining. In contradistinction, infantile hemangioma has a distinctive pathology and is positive on glut 1 immunostaining (4). Unfortunately, these data was not provided in the letter from Yoon et al (1).

Several experienced authors have reported that the term KM should be reserved for clinically significant thrombocytopenia and coagulopathy associated exclusively with KHE or tufted angioma (5,6). It is now generally accepted that tufted angioma and KHE are synonymous vascular tumors with similar presenting symptoms and potential for KM syndrome (3). To avoid confusing this type of tumor with infantile hemangioma, the use of proper terminology is essential, as adopted by the International Society for the Study of Vascular Anomalies based on the classification of vascular anomalies by Mulliken and Glowacki in 1982 (7). The clinical, imaging, and pathologic features of KHE are different from those of infantile hemangioma and lead to the correct diagnosis. This is of utmost importance because the entities have different prognoses and are treated with different modalities (3,8).

Arterial embolization may be life-saving in the acute setting of a critically ill newborn with KHE associated with KM phenomenon because it may rapidly reduce tumor volume and improve coagulation disorders (8), but combined treatment with systemic drugs such as corticosteroids, vincristine, and rapamycin is recommended to achieve long-term cure (4,8). These comments do not undermine the embolization treatment performed by Yoon et al (1), but are intended as a contribution to better management of this unusual disease.

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