Neurofibromatosis type 1 and subungual glomus tumors: A noteworthy association

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To the Editor: Neurofibromatosis type 1 (NF1) is an important diagnosis to make, particularly in its pretumor stage.[1,2] We are grateful to Lipner and Scher[3] for noting that the association between glomus tumors and NF1 that we mentioned might have been emphasized more.[4,5] We concur that glomus tumors may cause significant morbidity and represent an important clue to the diagnosis of this neurocutaneous syndrome.[1-3] These benign tumors are typically found on the fingertips, originate from the glomus body, produce a symptomatic triad of pinpoint tenderness, pain, and cold hypersensitivity in some patients, and be evident as longitudinal erythronychia or as a bluish nodule seen through the nail plate.[3]

We agree that the presence of glomus tumors merits consideration of NF1. The salient question is whether glomus tumors should be incorporated into the diagnostic criteria for NF1. In our article, we proposed modified diagnostic criteria and noted the inconsistency of considering freckling in the axillary or inguinal regions as a separate diagnostic criterion when these "freckles" are, in fact, simply small café-au-lait spots, themselves, in large size, being 1 of 7 diagnostic criteria, with 2 of 7 required to render a diagnosis of NF1.

Lipner and Scher[3] have persuaded us that glomus tumors should have been included in our Table 2 ("Other Manifestations of NF1") along with rhabdomyosarcomas and gastrointestinal stromal tumors.

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