Response to a letter to the editor concerning “Giant Cell Reparative Granuloma of the Scapula: report of a case and literature review”

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We read with interest the letter to the editor regarding our manuscript on giant cell reparative granuloma (GCRG) of the scapula. The author commented that some relevant aspects have been omitted in the description and discussion of the case, making the paper unclear for readers.

It assumes the possibility that our case should be considered a solid form of aneurysmal bone cyst (s-ABC). Bertoni et al. [1], Oda et al. [2], and Ilaslan et al. [3] reported lesions with similar morphology in the long bones and designated them as solid ABC or extragnathic GCRG. To the best of our knowledge and based on the recent published literature, these two entities have been included in the group of giant cell lesions of bone. No clear criteria have been identified to distinguish the GCRG that occurred in the extremities (not in gnathic area or in small bones of the hands and feet) with s-ABC. The importance of recent discovery about USP6 mutations in ABC, GCRG, and other entities has not been omitted. In fact, ABC is now widely accepted as a neoplasm owing to recent findings of recurrent clonal chromosomal alterations. USP6 (also called TRE17/ubiquitin-specific protease 6) is a deubiquitinase that is involved in the arrest of maturation of the osteoblasts. The related gene is localized to the short arm of chromosome 17 (17p13). The result of the chromosomal anomaly is a hybrid gene, mostly identified as t(16;17)(q22;p13) CDH11/USP6. As we previously reported, genetic abnormalities of chromosome 17 have been widely described in s-ABC or GCRG, even if other infrequent chromosomal rearrangements have also been reported [4]. The solid variant of ABC has been shown to harbor similar genetic events as the classic cystic lesions, as well in GCRG of the extremities [5]. Based on genetic evaluation, Agaram et al recommended that only the terminology of GCRG of the hands

and feet should be abandoned and replaced instead with either ABC or solid variant of ABC depending on the radiologic findings [5]. The USP6 rearrangement on FISH, if present, can distinguish s-ABC/GCRG from other lesions with secondary ABC formation, metastases, or brown tumors [6]. In our case, we did not perform the FISH analysis to find rearrangements of the USP6 gene.

As stated in our original manuscript, this entity has a wide range of clinical and radiographic presentation and requires adequate staging and biopsy to avoid diagnostic errors and potential mismanagement. Evaluation of UPS6 mutations could help in further histologic definition, even if up to 80% of both s-ABC and GCRG cases with genetic abnormalities reported in literature have shown some alteration of chromosome 17 [4].

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