LETTER TO THE EDITOR



Regarding the Use of the Term "Cementum" in Fibro-Osseous Lesions of the Craniofacial Skeleton

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I read with great interest Drs'. E. J. Raubenheimer and C. E. Noffke letter to the editor titled: *Regarding the use of the term "cementum" in pathologic proliferation* and would like to make a few comments regarding this controversial subject. To start with it is important to point out that our understanding of the basic cytogenetic and molecular genetic mechanisms involved in the development of fibro-osseous lesions of the jaws is very limited. Terms used to identify these entities are based on their morphologic and clinical features and not on pathogenetic or molecular basis [1, 2].

In the new edition of the WHO Classification of Head and Neck tumors (2017), ossifying fibromas were classified into odontogenic and nonodontogenic types [3]. The odontogenic ossifying fibroma is termed cemento-ossifying fibroma (COF). Two types of nonodontogenic fibromas are described; juvenile trabecular ossifying fibroma (JTOF) and juvenile psammomatoid ossifying fibroma (JPOF). The term COF is preferred for the odontogenic variant because they are believed to arise from the periodontal ligament and are characterized by production of bone and cementum-like calcified structures. The latter shows distinct morphology. It is typically hypocellular, basophilic, curvilinear structures that may coalesce forming ginger-root-like appearance. The cementum-like structures commonly manifest peripherally radiating collagen fibers reminiscent of Sharpey's fibers of the periodontal membrane [1, 2]. Under polarized light microscopy the cementum like tissue shows a characteristic quilted pattern. No such tissue is detected in extragnathic bone lesions. It important to state that the psammomatoid calcified particles seen in extragnathic JPOF are distinct and resemble bone more than cementum.

Cementum-like calcified tissue is also detected in cemento-osseous dysplasia of the jaws [4]. These lesions are classified according to their anatomic location and clinical

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presentation into periapical (associated with the apices of mandibular incisors), florid (multifocal, multiquadrant) and focal (single). The use of the descriptive term periapical is useful clinically and is utilized to distinguish it from other periapical pathologies.

Lastly, there is a group of very rare form of jaw fibroosseous lesions which is associated with genetic abnormality. It includes familial gigantiform cementoma (FGC) and gnathodiaphyseal dysplasia (GDD) [5, 6]. These are characterized by early onset multifocal, multiquadrant rapidly progressive expansive lesions that may be massive. Microscopically, the jaw lesions are indistinguishable from COF and COD. Unlike COD the jaw lesions are rapidly expansive and grow in a tumor-like fashion with rapid recurrence following corrective surgery.

GDD differs from FGC in that the jaw lesions in GDD are associated with extragnathic skeletal abnormalities, particularly of the tubular bones, that have been variably described as osteopenia, osteosclerosis and fragile bone. FGC may be inherited as autosomal dominant trait. However, sporadic cases are without known heritable features. GDD is also inherited in an autosomal dominant fashion. A mutated (GDD1) gene has been identified in multiple GDD families. The gene is also known as anoctamin 5 (ANO5) [6].

Whether the jaw lesions in FGC and GDD are neoplastic or dysplastic is a matter of speculation.

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