Chapter 7

Keratocystic Odontogenic Tumors – Clinical and Molecular Features

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Additional information is available at the end of the chapter

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1. Introduction

Keratocystic odontogenic tumors (KCOTs) are certainly among the most studied lesions in oral pathology, which is not a surprise considering their perplexing clinical behavior and complicated mechanism of pathogenesis. In fact, the specific KCOT features are the reason for numerous discussions regarding the true nature and classification of these lesions, which are still debated in the scientific community.Until recently these lesions were known as odontogenic keratocysts (OKCs), a term first used by Philipsen in 1956. In the beginning, the term was used to describe any jaw cyst in which keratin was formed. However, it became obvious that some other types of jaw cysts, such as radicular and residual cysts, may exhibit keratinization as well, leading to the conclusion that specific histological features of OKCs and not solely the presence of keratin, should be used to distinguish these lesions from other cysts of the jaws [1]. Researchers soon realized that OKCs show aggressive clinical behavior and high recurrence rates, features which are not typical for other odontogenic cysts [2]. Besides that, it has been noted that OKCs are among the most prominent feature of Nevoid Basal Cell Carcinoma Syndrome (NBCCS), also known as Gorlin-Goltz syndrome. Finally, numerous studies have shown that genetic factors are predominant in etiology of these lesions and that some mechanisms of pathogenesis, typical for neoplastic lesions, are also involved in formation of OKCs [3]. Therefore, in 2005 these lesions were reclassified as Keratocystic Odontogenic Tumors (KCOTs) and defined as benign, odontogenic, uni- or multicystic intraosseous tumors, with characteristic parakeratinized squamous epithelium lining, having a potential for aggressive and infiltrative growth [4]. However, since KCOTs also exhibit some cysts-like features, including response to decompression [5], the tumoral