

# Nonodontogenic Cysts of the Jaws and Treatment in the Pediatric Population



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## KEYWORDS

• Pediatric • Nonodontogenic • Cysts • Jaws

## KEY POINTS

- Nonodontogenic cysts within the jaws are not a common presentation, especially in the pediatric population.
- It is well documented that cysts within the pediatric population tend to be developmental and odontogenic in nature.
- Although nonodontogenic cysts of the jaws are relatively uncommon, it is imperative the clinician understand their clinical behavior and management because failure to do so can result in increased patient morbidity.
- The nonodontogenic cysts of the jaws that are most often encountered are the central giant cell granuloma, traumatic bone cavity, aneurysmal bone cyst, nasopalatine duct cyst, and nasolabial cyst.

## INTRODUCTION

Nonodontogenic cysts within the jaws are not a common finding in either the adult or pediatric patient population. Shear and Speight<sup>1</sup> conducted a retrospective review of 2616 cysts (in the general population) within the jaws and found 80.1% to be radicular cysts, dentigerous cysts, or odontogenic keratocyst/keratocystic odontogenic tumor. Manor and coworkers<sup>2</sup> found 95 of 322 cysts within the jaws to be in the pediatric (<16) population, with 82% of these being odontogenic. It is well documented that cysts within the pediatric population tend to be developmental and odontogenic in nature.<sup>3,4</sup> Although nonodontogenic cysts of the jaws are relatively uncommon, it is imperative the clinician understand their clinical behavior and management, because failure to do so can result in significantly increased patient morbidity.

The nonodontogenic cysts of the jaws most often encountered are the central giant cell granuloma (CGCG), traumatic bone cyst (TBC), aneurysmal bone cyst (ABC), nasolabial cyst, and nasopalatine duct cyst. This article focuses on the background, clinical findings, radiographic features, histopathologic features, and treatment of these nonodontogenic cysts of the jaws.

## CENTRAL GIANT CELL GRANULOMA *Background*

This is a benign lesion of unknown cause, possibly from hemorrhage into bone with incomplete healing. It was first described in 1953 by Jaffe who stated that this was a “giant-cell reparative granuloma.”<sup>1</sup> The use of the term “reparative” has since been abandoned but may still be seen on older pathology reports, or those written by non-oral and

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maxillofacial pathologists. The World Health Organization classifies this lesion as a benign idiopathic lesion. It is also considered a reactive lesion that is nonneoplastic in nature, but it can show behavior similar to benign neoplasms.<sup>5</sup> In the 2009 Cochrane review, Suárez-Roa and coworkers<sup>6</sup> described this entity as a rare benign tumor of the jaws with an unknown cause accounting for up to 7% of jaw tumors. Its incidence in the general population is 0.00011% (1 in 900,000).<sup>7</sup> Indeed, Chuong and coworkers<sup>8</sup> first described two variants, nonaggressive and aggressive, with the former being more common. To be classified as aggressive a lesion must have one of the following characteristics: pain, paresthesia, cortical plate perforation, rapid growth, root resorption, or a high rate of recurrence following surgical curettage.<sup>8</sup> Additionally, CGCGs have been shown to be related to cherubism, hyperparathyroidism, neurofibromatosis type 1, and Noonan syndrome.

### **Clinical Features**

This lesion is more common in children and young adults younger than age 30, but can occur at any age.<sup>9</sup> There is a female predilection of 2 to 1. The lesion is more common in the mandible and maxilla, but involvement of other facial bones has been reported.<sup>5</sup> The mandible is affected approximately 70% of the time, with a relatively even distribution in the anterior to posterior regions.<sup>10</sup> Conversely, it is more common in the anterior maxilla, and may cross the midline. Nonaggressive lesions are typically asymptomatic, painless, and slow-growing, and the aggressive lesions present with one or more of the symptoms listed previously. These aggressive lesions are more often found in children and are greater than 5 cm in diameter (Figs. 1 and 2).<sup>3</sup>

### **Radiographic Features**

The appearance of these lesions varies greatly, and depends on the criteria that define them as aggressive or nonaggressive. Aggressive lesions often cause displacement of teeth, resorption of roots, and perforation of the cortical plate. They



**Fig. 2.** Intraoral view of CGCG of left maxilla in a 9 year old.

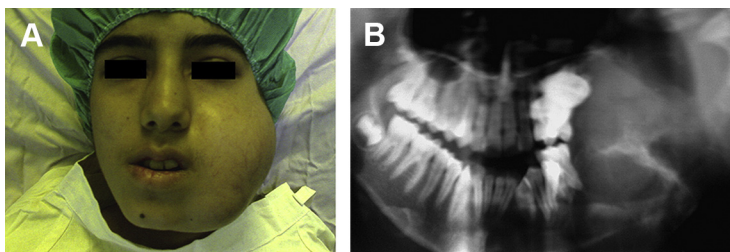
can be unilocular or multilocular, appearing as well-defined, usually with scalloped margins or as an irregular radiolucency. The term “soap bubble” appearance is often used to describe these lesions (see Figs. 1 and 5).

### **Histopathology**

A highly cellular, uniform fibroblastic stroma with aggregates of multinucleated giant cells is the diagnostic characteristic of the CGCG. The giant cells may vary in appearance, and extravasated red blood cells are commonly encountered (Fig. 3).<sup>4</sup>

### **Treatment**

All patients with an initial diagnosis of CGCG must be investigated further to rule out other diseases, particularly hyperparathyroidism. The first diagnostic adjunct must be a blood test to ascertain serum calcium and parathyroid hormone levels. In true CGCG, these laboratory values are normal. If the calcium and parathyroid hormone levels are elevated, the CGCG diagnosis may represent a brown tumor of hyperparathyroidism. Management of the parathyroid disorder results in resolution of the CGCG. Conventional surgical management of CGCG consists of enucleation and curettage, excision, or en bloc resection with



**Fig. 1.** (A) Locally aggressive CGCG in a 13 year old. (B) Panoramic radiograph. (From Shirani G. Management of a locally invasive central giant cell granuloma (CGCG) of mandible: report of an extraordinary large case. *J Craniomaxillofac Surg* 2009;37:531; with permission.)

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