Cystic Lesions of the Jaw in Children
A 15-Year Experience

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IMPORTANCE Pediatric jaw cysts represent a rarely symptomatic clinical entity and are not well addressed in the otolaryngology literature. It is important that otolaryngologists should be familiar with these lesions, which can manifest as jaw swelling or as paranasal sinus abnormalities.

OBJECTIVE To review the clinical presentation, radiologic features, management, and outcomes of jaw cysts in children treated at a single academic institution.

DESIGN, SETTING, AND PARTICIPANTS Retrospective medical record review at a tertiary care children's hospital among patients 16 years and younger who were seen with a cystic jaw lesion.

MAIN OUTCOMES AND MEASURES Medical records were reviewed for data on symptoms, physical examination findings, imaging, pathology, interventions, and outcomes among children who were seen at the Department of Oral and Maxillofacial Surgery or the Department of Otorhinolaryngology–Head and Neck Surgery at the University of Maryland Medical Center between January 1997 and December 2012 and were diagnosed as having a jaw cyst.

RESULTS Fifty-seven patients were identified who were diagnosed as having a true cystic jaw lesion and whose complete medical records were available for review. The most common cystic lesions were keratocystic odontogenic tumors (n = 19) and dentigerous cysts (n = 17). Fifty-six percent (32 of 57) of all cystic lesions were asymptomatic on presentation and were identified by imaging. The second most common presentation was local swelling (n = 15), followed by dental irregularities (n = 6). All patients, except for 1 with an eruption cyst, required surgical intervention, including biopsy, enucleation, curettage, or ostectomy, with reconstruction as indicated. Keratocystic odontogenic tumors tended to require more treatment (median, 2 procedures) for metachronous lesions or recurrence.

CONCLUSIONS AND RELEVANCE Pediatric jaw cysts are unusual, and data are scarce regarding their presentation and management. Many of these cysts are asymptomatic and are identified incidentally on orthopantomography. Keratocystic odontogenic tumors were the most common lesion seen in our series, followed by dentigerous cysts. Surgical intervention is required in most patients with a cystic lesion of the jaw.
Cystic lesions of the jaw are rarely symptomatic in the pediatric age group, and most tend to be of odontogenic origin. Their predilection to these bony structures is owing to the presence of residual epithelial nests during and after odontogenesis. Most lesions tend to be benign, although some can be locally aggressive and cause cosmetic and functional impairment. They are classified as odontogenic, nonodontogenic, or pseudocysts. Most cysts in the craniofacial skeleton occur in the mandible and maxilla and have an epithelial lining of odontogenic origin. They tend to manifest deep within the bone, where these epithelial remnants are located. When closer to the surface, they can produce a smooth indentation or cavity within the underlying bone that is seen on radiology. Radicular cysts are the most common jaw cysts occurring in the general population and are inflammatory rather than developmental in origin.1-3 However, in the pediatric population, developmental dentigerous cysts predominate and tend to occur around impacted or unerupted teeth.4 This development occurs because newly erupted permanent teeth are unlikely to have been subjected to infectious decay or trauma that is responsible for cyst formation due to inflammation.5,6 Although some of these lesions can be identified with regular dental examination, it is often difficult to differentiate them based on the clinical presentation and radiologic features alone. The radiographic appearances of several benign but occasionally aggressive lesions of childhood are similar, while requiring vastly different treatments. Keratocystic odontogenic tumors (KOTs) and ameloblastomas are highly proliferative lesions that can result in substantial osseous destruction without proper surgical management.7-10 Although rare, an ever-present potential of malignancy also necessitates a thorough evaluation of pathological specimens at the time of definitive jaw cyst management.

There is little information in the otolaryngology literature regarding the prevalence, clinical presentation, and management of jaw cysts in the pediatric age group, with most studies found in the dental literature. Otolaryngologists should be familiar with these lesions, which can be seen on imaging as facial swelling or as a mass in the maxillary sinus. We present our experience with a series of pediatric patients managed at a tertiary care children’s hospital over a period of 15 years, along with a brief review of the literature.

### Methods

The study was approved by the Institutional Review Board of the University of Maryland, Baltimore. A retrospective medical record review was performed among patients 16 years and younger who were seen at the Department of Oral and Maxillofacial Surgery or the Department of Otorhinolaryngology-Head and Neck Surgery at the University of Maryland Medical Center between January 1997 and December 2012 and had a final diagnosis of a jaw cyst, maxillary or mandibular cyst, or dental cyst. This group was then stratified to evaluate only those with benign cystic lesions on final pathology. Medical records were analyzed for patient sex, age, pathological diagnosis, initial signs and symptoms, radiologic features, interventions, and outcomes.

### Results

Over the 15-year period, 57 patients were identified who were diagnosed as having a true cystic lesion involving the mandible or maxilla and whose complete medical records were available for review at our institution. The mean (SD) patient age was 10.5 (3.2) years, with a total of 58 independent lesions (one child had 2 separate cysts). There was an approximate 1:1 ratio of boys to girls. Most of the cysts were KOTs (n = 19) or dentigerous cysts (n = 17). Demographic data and location of the cysts are summarized in Table 1. In total, 32 of 57 patients were asymptomatic on presentation, with 24 cysts being discovered on routine orthopantomography and 1 found incidentally on lateral neck radiography. The next most common symptoms were local swelling (n = 15) and dental irregularities (n = 6). The mandible alone was the primary site of the cyst in 34 individuals, whereas the maxilla alone was involved in 16 individuals. The remainder had lesions in both locations. All but one individual required at least 1 surgical procedure for diagnosis or definitive treatment. The initial symptoms by diagnosis, treatment, and outcomes are summarized in Table 2.

Keratocystic odontogenic tumors were the most common pathological finding, with a 0.9:1 ratio of boys to girls. This

### Table 1. Demographic Data and Location of Cystic Lesions*

<table>
<thead>
<tr>
<th>Final Diagnosis</th>
<th>No. of Patients (N = 57)</th>
<th>Sex</th>
<th>Age, y</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Male</td>
<td>Female</td>
<td>&lt;10</td>
</tr>
<tr>
<td>Keratocystic odontogenic tumor</td>
<td>19</td>
<td>9</td>
<td>10</td>
<td>7</td>
</tr>
<tr>
<td>Dentigerous cyst</td>
<td>17</td>
<td>12</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>Traumatic bone cyst</td>
<td>10</td>
<td>3</td>
<td>7</td>
<td>2</td>
</tr>
<tr>
<td>Radicular cyst</td>
<td>6</td>
<td>1</td>
<td>5</td>
<td>2</td>
</tr>
<tr>
<td>Calcifying odontogenic cyst</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Eruption cyst</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Nasopalatine duct cyst</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Aneurysmal bone cyst</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Odontogenic cyst</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

*One individual had 2 simultaneous diagnoses on final pathology.
pathology tended to be found in older patients (mean [SD] age, 11.3 [3.5] years). These lesions were more often symptomatic on presentation, with patients experiencing pressure and pain associated with their jaw mass. However, KOTs in 7 of 19 individuals (37%) were discovered incidentally on orthopantomography. Both the mandible and maxilla were frequently involved, with 6 individuals having simultaneous lesions with similar radiologic features at each location, as shown in Figure 1. Keratocystic odontogenic tumors required the most extensive operative management, with a median of 2 procedures per patient, and approximately half of the patients (11 of 19) required extensive dental extractions and bony ostectomies. Overall, 10 of 19 patients underwent staged procedures, with 7 of them having complete resolution and no sign of recurrence at the last follow-up visit. Ultimately, 2 of the 3 remaining individuals were diagnosed as having basal cell nevus syndrome (BCNS) and required multiple procedures for recurrent disease. No recurrence occurred in 6 of 9 individuals undergoing single-stage enucleations. In patients with synchronous lesions, BCNS was strongly suspected, and they were referred for a genetic evaluation. In total, 7 individuals were ultimately diagnosed as having BCNS.

Dentigerous cysts were the second most common pathological finding, with a 2.4:1 ratio of boys to girls, and occurred in younger patients (mean [SD] age, 9.3 [3.5] years). Eight patients with these cysts were initially seen with swelling or pain, and 6 patients were asymptomatic and were diagnosed incidentally on orthopantomography. On imaging, the cysts appeared as well-circumscribed radiolucencies surrounding the crowns of teeth, as shown in Figure 1. Basal Cell Nevus Syndrome

<table>
<thead>
<tr>
<th>Final Diagnosis</th>
<th>No. of Patients (N = 57)</th>
<th>Imaging</th>
<th>Initial Symptoms</th>
<th>Definitive Therapy</th>
<th>Median No. of Procedures</th>
<th>Outcome</th>
<th>No. of Patients With Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Keratocystic odontogenic tumor</td>
<td>19</td>
<td>OP (n = 18) CT (n = 12) MRI (n = 1)</td>
<td>Swelling, pain, facial and sinus pressure</td>
<td>Enucleation vs marsupialization plus affected tooth extraction with or without peripheral ostectomy</td>
<td>2</td>
<td>Multiple procedures for resolution</td>
<td>9</td>
</tr>
<tr>
<td>Dentigerous cyst</td>
<td>17</td>
<td>OP (n = 14) CT (n = 6) MRI (n = 1)</td>
<td>Swelling, incidental finding on OP; pain, failure of tooth eruption</td>
<td>Enucleation vs marsupialization plus affected tooth extraction</td>
<td>1</td>
<td>Complete resolution</td>
<td>1</td>
</tr>
<tr>
<td>Traumatic bone cyst</td>
<td>10</td>
<td>OP (n = 8) CT (n = 6) MRI (n = 2)</td>
<td>Asymptomatic, incidental finding</td>
<td>Incisional biopsy with or without curettage</td>
<td>1</td>
<td>Complete resolution</td>
<td>0</td>
</tr>
<tr>
<td>Radicular cyst</td>
<td>6</td>
<td>OP (n = 6) CT (n = 2) MRI (n = 0)</td>
<td>Swelling, aberrant dental growth, asymptomatic</td>
<td>Incisional biopsy plus tooth extraction</td>
<td>1</td>
<td>Decrease in size plus resolution</td>
<td>0</td>
</tr>
<tr>
<td>Calcifying odontogenic cyst</td>
<td>1</td>
<td>OP (n = 1) CT (n = 0) MRI (n = 0)</td>
<td>Asymptomatic, incidental finding</td>
<td>Enucleation and curettage plus tooth extraction</td>
<td>2</td>
<td>Complete resolution</td>
<td>0</td>
</tr>
<tr>
<td>Eruption cyst</td>
<td>1</td>
<td>OP (n = 1) CT (n = 0) MRI (n = 0)</td>
<td>Asymptomatic, incidental finding</td>
<td>Nonoperative</td>
<td>0</td>
<td>Complete resolution</td>
<td>0</td>
</tr>
<tr>
<td>Nasopalatine duct cyst</td>
<td>1</td>
<td>OP (n = 1) CT (n = 0) MRI (n = 0)</td>
<td>Pain, firm swelling</td>
<td>Enucleation plus curettage</td>
<td>2</td>
<td>Complete resolution</td>
<td>0</td>
</tr>
<tr>
<td>Aneurysmal bone cyst</td>
<td>1</td>
<td>OP (n = 1) CT (n = 1) MRI (n = 0)</td>
<td>Pain, swelling, hypoesthesia</td>
<td>Extraction, hemimandibulectomy, cryotherapy, plus reconstruction</td>
<td>7</td>
<td>Multiple reconstructive procedures</td>
<td>1</td>
</tr>
<tr>
<td>Odontogenic cyst</td>
<td>1</td>
<td>OP (n = 1) CT (n = 0) MRI (n = 0)</td>
<td>Painless swelling of lip</td>
<td>Enucleation</td>
<td>1</td>
<td>Complete resolution</td>
<td>0</td>
</tr>
</tbody>
</table>

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging; OP, orthopantomography.

* One individual had 2 simultaneous diagnoses on final pathology.

Figure 1. Basal Cell Nevus Syndrome

Coronal view bone window computed tomography shows mandibular and maxillary synchronous unilocular radiolucent lesions. Note the bone perforation at the superior margin of the mandibular lesion. R indicates right; L, left.
The primary sites were the mandible in 11 patients and the maxilla in 5 patients, with 1 patient having simultaneous cysts in the mandible and maxilla. Five patients underwent initial marsupialization, while 6 patients underwent initial enucleation. Nine patients required extraction of teeth during their initial procedure. All patients had complete resolution, although 3 required an additional procedure that included orthodontic assisted extrusion. Complete resolution of these lesions was the norm.

The third and fourth most common lesions were traumatic bone cysts and radicular cysts, respectively. Traumatic bone cysts (n = 10) were exclusively localized to the mandible and had a 3:7 ratio of boys to girls. The mean (SD) age of patients with these lesions was 11.9 (2.4) years, which was approximately 1.4 years older than the mean age of the entire cohort. All of these lesions were asymptomatic on presentation and were discovered on imaging. All individuals had operative intervention, with most undergoing biopsy with curettage of the cyst cavity. Two patients required retreatment with curettage for complete resolution of their cysts.

There were 6 radicular cysts, 5 of which occurred in girls, with an overall mean (SD) patient age of 10.3 (2.0) years. Radicular cysts were found twice as often in the maxilla as in the mandible. Three of the cysts were asymptomatic, with the rest manifesting as swelling and dental disease. All patients underwent biopsy, with 4 individuals requiring dental extraction of associated teeth.

The remaining cystic lesions occurred in single patients only and included a calcifying odontogenic cyst, an eruption cyst, a nasopalatine duct cyst, an aneurysmal bone cyst, and an odontogenic cyst (not otherwise specified). All were incidentally discovered.

Discussion

Most jaw cysts in our series of pediatric patients were developmental rather than inflammatory. This finding is to be expected in a pediatric population, among whom considerable growth and development occur. Specifically, the most common cysts were KOTs, comprising 33% (19 of 57) of true cystic lesions. Although occurring frequently among the pediatric population, there was a higher proportion of KOTs in our series than is reported in the literature, likely because our institution is a tertiary care center. The lesions can be aggressive and require complex surgical intervention and long-term follow-up care. Individuals with these cysts often experience recurrence, which is characteristic of this pathology because of residual epithelial tissue after any treatment other than resection. Early appearance of KOTs, more frequent recurrence, and the development of new primary lesions are common among patients with BCNS. Treatment of these children goes well beyond management of the tumor and selective dental extractions, often including jaw reconstruction and comprehensive dental rehabilitation at skeletal maturity.

Dentigerous cysts, which accounted for 30% (17 of 57) of lesions in our study, are the most common jaw cysts in the pediatric population according to the published literature. Herein, these lesions were primarily confined to the mandible and were posteriorly located in association with the molar teeth in 10 of the cysts. We observed a low incidence of radicular (periapical) cysts compared with studies among other pediatric and general populations, perhaps because these types of lesions typically do not require tertiary medical care and are managed in community dental settings.

Most important, 56% (32 of 57) of all cystic lesions in our series were asymptomatic on presentation, which is why many of these lesions go unnoticed in the general population. In particular, KOTs have a tendency to spread through the marrow space or expand into the maxillary sinus rather than resulting in the bony expansion and palpable lumps noted in other intraosseous jaw pathology. This asymptomatic nature of cystic lesions highlights why many of them are first identified on imaging screens. Orthopantomography is a sensitive imaging modality that can delineate differences among disease processes based on the appearance of a lesion, which can be detected by an astute diagnostician. It is an ideal method for intraosseous lesion screening, with incidental finding on imaging more common than patient report of any symptoms of pain, swelling, or dental abnormalities (due to tooth impaction or gingival changes). In some instances, computed tomography (CT) may be used for operative planning and provides the most detailed information about the cortical thickness and structural integrity of the residual jaw, which must be considered before extirpation and reconstruction. The mandible was the predominant location of cystic lesions in our series, although this predilection has varied among published studies. The osseous location of jaw cysts has been amenable to dental imaging software programs, which have proved beneficial for evaluating cortical bone involvement and mandibular anatomy. In our study, CT was used only for larger lesions with suspected
extension to surrounding osseous and soft-tissue structures and posterior maxillary lesions. More recently, cone beam CT has been used when possible because of its 10-fold reduced radiation exposure compared with body CT. When this imaging modality is not an option, pediatric protocols exist for conventional CT in which substantial weight-based dose attenuation can be accomplished without loss of diagnostic accuracy.

Despite the radiation exposure that is inherent with CT and the ever-increasing parental concern in this regard, this imaging is far superior for planning purposes because jaw cyst lesions are located in bone and tend to deform it in unique patterns. Computed tomography can be used for extirpative and reconstructive planning, accurately evaluates residual surrounding bone, and is well tolerated by young children. In addition, with pediatric CT protocols and office-based cone beam CT scanners, imaging can be obtained with radiation exposure that is little more than that of dental radiographs. Magnetic resonance imaging can be a prolonged study and thus difficult to perform in children without sedation or general anesthesia, while yielding less practical information. Although magnetic resonance imaging has a role in the diagnosis of jaw cysts, its usefulness is not as well defined as that of CT. Magnetic resonance imaging has been shown to be useful in differentiating between pathologies (eg, ameloblastoma vs KOT), but a tissue diagnosis is still required. Immune markers, such as anticytokeratin antibodies, show promise to delineate disease processes, especially among more potentially destructive conditions, such as KOTs. However, they do not have a role in guiding treatment at present.

Cystic lesions of the jaw represent diseases in which most patients undergo invasive procedures for a curative intent. In most cases, this surgery is successful, leading to resolution of the disease process. In the pediatric population, among whom osseous and dental structures are continuing to develop and mature, the extent of resection requires careful planning, without compromising complete excision of the disease. Even with aggressive therapy, KOTs in our study showed a tendency to recur, and this finding is consistent with the literature, which highlights their obstinate nature, with poor outcomes and a propensity to develop new lesions. Individuals in our study with multiple synchronous KOTs often had a final diagnosis of BCNS (Gorlin syndrome). Because of the genetic basis of this condition, these patients were more likely to have multiple lesions, often in both the maxilla and mandible, where de novo lesions can sometimes be difficult to differentiate from recurrence. This condition predisposes patients to more procedures, and in our series, a diagnosis of multiple lesions required a median of 2 procedures per patient.

In general, treatment varied based on the size and extent of cystic lesions. We typically stage lesions larger than 2 cm with primary drainage or grommet placement, followed by enucleation, curettage, and ostectomy as indicated. Lesions 2 cm or smaller undergo definitive enucleation and curettage as the initial procedure. Staging procedures with marsupialization to reduce the size of the cyst and thicken the lining is a viable option and has shown some promise in the literature to reduce recurrence. Studies have shown that the most effective treatment for KOTs includes combined surgical enucleation and adjuvant application of Carnoy solution to the cyst cavity, with reported recurrence rates as low as 5%. However, Carnoy solution in its original formulation (with chloroform) is not approved for use in humans by the Food and Drug Administration, and the use of the modified solution without chloroform is no more effective than conventional surgical treatment. The only current treatment that is equivalent to the use of the original Carnoy solution is resection. However, given the benign nature of the disease, surgery likely constitutes overtreatment in many cases. Regarding reconstructive efforts, some surgeons advocate bone grafting for these lesions and defects, although the use of such materials has not been proved beneficial for healing in the pediatric population, even in larger cystic lesions. In addition, given the 30% recurrence rate of jaw cysts, it is advisable to avoid grafting of any kind until recurrence is no longer likely.

This investigation was a retrospective study and has inherent limitations, including inconsistencies in the medical record description of some lesions and nonuniform follow-up care. Our study largely reflected the referral patterns in our geographic area to surgeons with known expertise in certain disease entities, and the findings may not have external validity. This work was intended to familiarize otolaryngologists with benign pediatric jaw cysts and to suggest a broad-based approach to management. Because of the diversity of lesions encountered, relative infrequency, and differing recurrence rates, it was difficult to formulate a standardized treatment. The outcomes represent our group’s philosophy regarding surgical management of jaw cysts, which continues to evolve as newer diagnostic and therapeutic options become available.

Conclusions

Pediatric jaw cysts are rarely symptomatic. Small lesions tend to be underrecognized because of the lack of symptoms and may be identified on routine dental imaging. Most of our patients had developmental odontogenic cysts, and this finding is in agreement with the literature on jaw cysts. Keratocystic odontogenic tumors were the most common lesion observed in our series, followed by dentigerous cysts. Patients with recurrent KOTs are more likely to have a diagnosis of BCNS (Gorlin syndrome) by genetic testing. Surgical intervention was required for most patients in our series. Keratocystic odontogenic tumors were more likely to recur than other jaw cysts and require additional procedures for cure. The approach to management of cystic lesions of the jaw in children should consider future growth and development of the face.
Cystic Lesions of the Jaw in Children

Original Investigation Research

References


