

# Disorders and Tumors of the Salivary Glands in Children



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## KEYWORDS

- Pediatric ear/nose/throat • Salivary glands • Pediatric tumors head and neck • Hemangioma

## KEY POINTS

- Salivary gland neoplasms are rare in children.
- An asymptomatic swelling in the periauricular region is the most common presenting complaint in older children.
- Approximately 50% of salivary gland lesions are malignant, dictating a thorough diagnostic evaluation by a head and neck surgeon.
- Surgical excision is the primary treatment of lesions.
- Histopathologic findings determine prognosis.

## INTRODUCTION

Disorders of the salivary glands can be broadly divided into neoplastic and nonneoplastic disease. Further divisions can be made, as salivary gland lesions are associated with a wide spectrum of etiology (**Table 1**). The age of the patient narrows the potential diagnoses, with vascular lesion often presenting at birth or within the first year of life,<sup>1</sup> whereas solid tumors, with notable exceptions, are more likely to occur in older children.<sup>2,3</sup> Inflammatory disease usually has a relatively rapid onset in contrast to neoplastic or congenital processes, which have more insidious onsets.<sup>4</sup>

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<b>Table 1</b>	
<b>Differential diagnosis</b>	
<b>Nonneoplastic lesions</b>	
Congenital/developmental	First branchial arch anomalies
Inflammatory and infection	Acute bacterial sialadenitis Viral: mumps, cytomegalovirus, Coxsackie A or B, or parainfluenza virus Human immunodeficiency virus (HIV)-associated salivary glands Recurrent parotitis in children (RPC)
Granulomatous disease	Mycobacterial disease Actinomycosis Sarcoidosis
<b>Necrotizing sialometaplasia</b>	
Autoimmune	Sjögren syndrome
Cysts	Ranula Mucocele
<b>Neoplastic tumors</b>	
Benign epithelial	Pleomorphic adenoma
Benign mesenchymal	Hemangioma Lymphangioma
Malignant-epithelial	Mucoepidermoid carcinoma Acinic cell carcinoma Adenoid cystic carcinoma Sialoblastoma
Mesenchymal	Rhabdomyosarcoma

Tenderness of the affected area is most often associated with inflammatory disease. Persistent, firm, painless, well-circumscribed lesions should be thoroughly investigated with imaging such as ultrasonography, computed tomography (CT), and MRI, and occasionally with fine-needle aspiration in older children.<sup>5</sup> In a large review of 9993 salivary gland lesions, 430 were in children, accounting for 4.3% of the total.<sup>6</sup> Of the lesions in children, 262 were nonneoplastic and 168 others were considered tumors. This review discusses neoplastic lesions of the salivary glands in children, and malignant epithelial tumors in particular.

## **NEOPLASTIC TUMORS**

### ***Benign Epithelial Tumors***

#### ***Pleomorphic adenoma***

Pleomorphic adenomas (PAs) account for approximately half of all epithelial tumors of the salivary glands and more than 90% of benign salivary gland epithelial tumors.<sup>2,6–20</sup> Composite analysis of several studies reveals that most (62%; range 56%–77%) PAs occur in the parotid gland, with the submandibular gland (26%; range 11%–40%) and the minor salivary glands (12%; range 0%–21%) being less frequent sites of origin.<sup>6,8,9,16,18,21–23</sup> The female to male ratio is approximately 1.4:1.<sup>22</sup> The typical presentation of a PA is a slow-growing, painless, and firm mass with an average duration of symptoms of approximately 12 months.<sup>17,22</sup> On ultrasonography, CT, and MRI, PAs are most commonly well defined. On CT, small calcifications can be seen and on MRI, PAs are typically very bright on T2-weighted images and show homogeneous or heterogeneous enhancement (**Fig. 1**). On gross inspection, PAs are well circumscribed

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