PST | PEDIATRIC SURGERY IN TROPICS

PAROTID GLAND MALIGNANCY IN PEDIATRICS: MUCOEPIDERMOID CARCINOMA

Matlhatse Phuti Maluleke, Elliot Motloung, Modise Zacharia Koto, Eliyah Aaron Sischy

Faculty of Medicine, Sefako Makgatho Health Science University, Department of Pediatric Surgery, Ga-Rankuwa

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Keywords	**Abstract**
Salivary gland, parotid, mucoepidermoid, carcinoma, total parotidectomy	**Introduction:** Salivary gland neoplasms are rare in children and adolescents, with an estimated incidence of ~4% in patients under 20 years of age. Malignant epithelial salivary gland neoplasms are particularly rare, most often presenting around 14 years of age, and with a female predominance. The parotid gland is the most frequently affected anatomical site. Low or intermediate grade mucoepidermoid carcinoma is the most common malignant subtype in this age group, followed by acinic cell carcinoma and adenoid cystic carcinoma, which together account for up to 80 to 90% of pediatric cases. Compared with adults, firm salivary gland swellings in children should raise a higher index of suspicion for malignancy. Additional concerning features include local pain and facial nerve palsy. Imaging with ultrasonography, magnetic resonance imaging (MRI), and occasionally contrast enhanced computed tomography (CECT) scan are essential for evaluating tumor extent and detecting metastasis. Complete surgical (R_0) resection remains the cornerstone of treatment, with histological grade, perineural invasion, and margin status serving as important prognostic factors. **Case Report:** We describe a 7-year-old girl who presented with a 4-month history of a progressive, painless right parotid gland mass.

	Presentation at this young age is exceptionally rare, as most pediatric salivary gland malignancies are reported closer to adolescence. Biopsy revealed an invasive neoplasm with squamous and mucinous differentiation, consistent with mucoepidermoid carcinoma. After correlation with radiological findings, the patient underwent a right total parotidectomy with lymph node dissection. Final histology confirmed a low grade mucoepidermoid carcinoma with clear (R_o) resection margins and no evidence of nodal metastasis. **Conclusion:** Pediatric salivary gland malignancies are rare but clinically significant, and prompt recognition is essential. Mucoepidermoid carcinoma is the most common malignant variant, and surgical excision provides excellent outcomes when complete (R_o) resection is achieved. This case highlights the unusual occurrence of mucoepidermoid carcinoma in a young child, underlining the importance of maintaining suspicion even at an early age. Only a few cases under the age of 10 have been documented in the literature, further emphasizing the rarity and clinical relevance of this presentation.
Abbreviations	TREP: Tumori Rari in Età Pediatrica MRI: Magnetic Resonance Imaging CECT: Contrast Enhanced Computed Tomography FNA: Fine needle aspiration SEER: Surveillance, Epidemiology, and End Results

INTRODUCTION

Salivary gland neoplasms are uncommon in adults and extremely rare in children, particularly in the prepubertal age group.[1] Pediatric salivary gland malignancies represent approximately 10% of all pediatric head and neck cancers,[2] yet only ~5% of all salivary gland neoplasms occur in this population.[3] Unlike in adults, the majority of pediatric cases are epithelial malignancies.[3] These neoplasms display wide histopathological diversity, creating challenges in surgical planning and in determining the role of adjuvant therapy.[4] Epidemiological data highlights the rarity of these neoplasms. The Surveillance, Epidemiology, and End Results (SEER) database estimates an annual incidence of 0.8 per million among individuals aged 0 to 19 years, a finding consistent with results from the Italian TREP (Tumori Rari in Età Pediatrica) project.[5,6] Among epithelial salivary gland neoplasms, mucoepidermoid carcinoma and adenoid cystic carcinoma are the most frequent histologic types in both adults and children.[7] Clinical and pathological features are largely similar across age groups.

The most common clinical presentation of these neoplasms is a painless, firm, noninflammatory, and slow growing mass. [8] Surgical resection remains the cornerstone of management. A Mayo Clinic series reported recurrence rates for mucoepidermoid carcinoma of 48% post simple enucleation, 31% post lateral parotidectomy, and 0% post total parotidectomy. [9] These findings support total parotidectomy as the preferred surgical approach for malignant parotid neoplasms in children. If the facial nerve is infiltrated by the neoplasm, surgical resection of the nerve is required to achieve an (R_o) resection margin, followed by appropriate rehabilitation. [10] Neck dissection in children follows similar principles as in adults, and it is indicated when abnormal lymph nodes are detected clinically or radiologically. Elective neck dissection may be appropriate for undifferentiated neoplasms, large lesions (T3 to T4), or in selected individual cases. [11,12]
Adjuvant radiotherapy is reserved for specific indications such as an (R_1) resection, perineural invasion, advanced disease stage, or poorly differentiated histology.[13] In pediatric patients, however, potential adverse effects of radiotherapy including impaired craniofacial growth, osteoradionecrosis, trismus, dental anomalies, visual complications, and radiation induced secondary malignancies necessitate cautious use.[14] Chemotherapy is generally limited to palliative situations or recurrence where surgery and radiotherapy are no longer feasible. Despite these challenges, mucoepidermoid carcinoma carries a relatively favorable prognosis. Even when arising as a secondary malignancy, for example, post treatment of lymphoma, reported 5-year survival rates approach 94%. [7] Prognostic factors include histopathological grade, perineural invasion, and margin status. Most children present with low grade neoplasms, and with complete surgical (R_0) resection, excellent long-term outcomes can be achieved regardless of the neoplasm site. [15]

CASE PRESENTATION

We report on the case of a 7-year-old girl who presented to our institution with a 4-month history of a progressive, painless right lateral neck swelling. She had no constitutional symptoms, dysphagia, or respiratory symptoms. Her past medical and surgical history was unremarkable, with no prior radiation exposure. She had no family history of malignancy or tuberculosis contact. Perinatal history was unremarkable. On clinical examination, she appeared well nourished and not chronically ill. Her anthropometric data was appropriate for age. Inspection revealed a visible right parotid region mass with no overlying skin

changes or restriction of neck movement. The mass was hard in consistency, irregular, non-tender, and fixed to the underlying structures. Cranial nerve functions (facial, glossopharyngeal, vagus, spinal accessory, and hypoglossal) were intact. She had palpable level V (posterior neck triangle) lymphadenopathy. Contrast enhanced computed tomography (CECT) scan of the neck demonstrated a solid mass originating from the right parotid gland. (Fig 1)

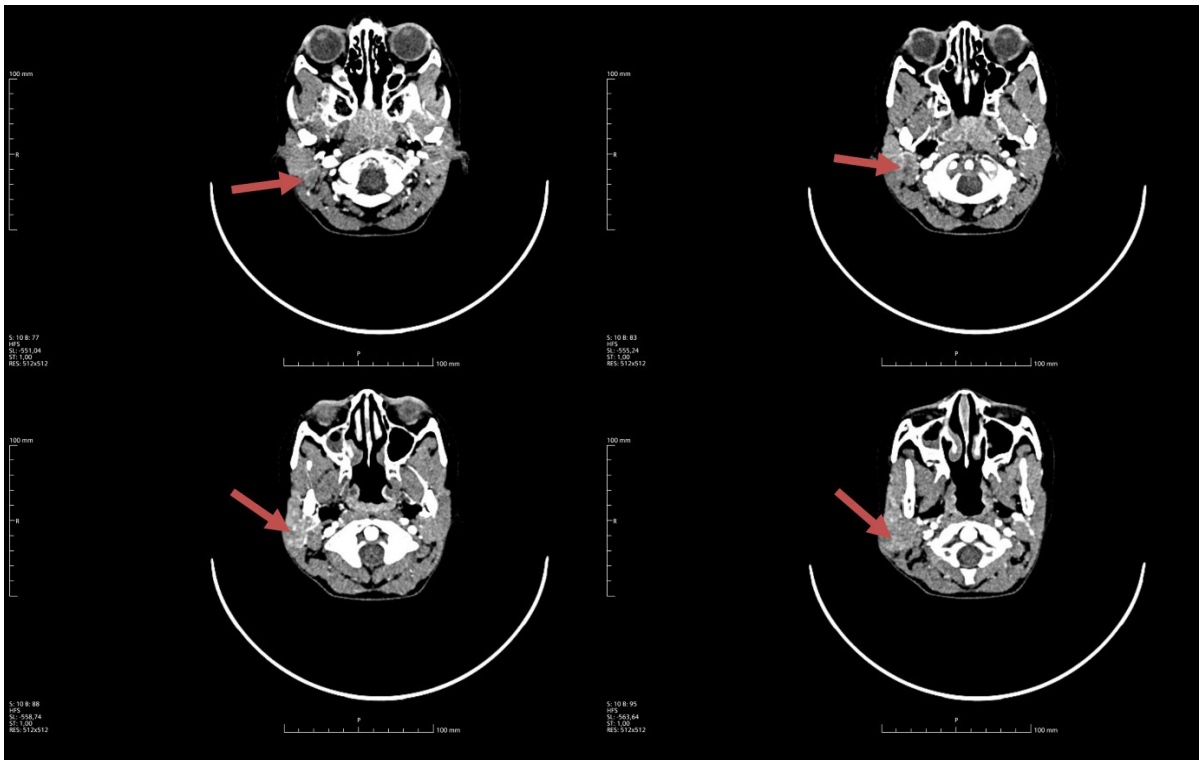

Fig 1. CECT scan of the neck demonstrating a solid right parotid gland mass

Histopathological examination of the biopsy tissue revealed an epithelial neoplasm with squamous and mucinous differentiation. The neoplasm was composed of infiltrative cohesive nests, trabeculae, and cords, with cystic glands containing intraluminal and intracytoplasmic mucin, as well as goblet cells. The neoplastic cells included a mixture of squamous, intermediate, and mucinous types, with mild to moderate pleomorphism and rare mitoses. No necrosis, perineural invasion, or lymph vascular invasion was identified. Lymph node biopsy demonstrated reactive changes only. Histopathological findings were consistent with a low grade mucoepidermoid carcinoma, classified as low-risk morphology

according to the Brandwein-Grensler grading system (cystic component = 1, neural invasion = 0, necrosis = 0, mitosis = 0, anaplasia = 0).

Following a multidisciplinary discussion of this case, a surgical resection was recommended. The family was thoroughly counselled, and an informed consent for surgical resection was obtained. The patient successfully underwent a right total parotidectomy with lymph node dissection. A lazy S preauricular cervical incision was made, and a superficial cervicofacial flap was elevated. (Fig 2) The facial nerve landmarks were identified and preserved, with careful blunt dissection providing access to the deep parotid lobe. (Fig 2) The neoplasm was meticulously resected without capsular breach. Her postoperative course was unremarkable, and her cranial nerve function remained intact. (Fig 3)

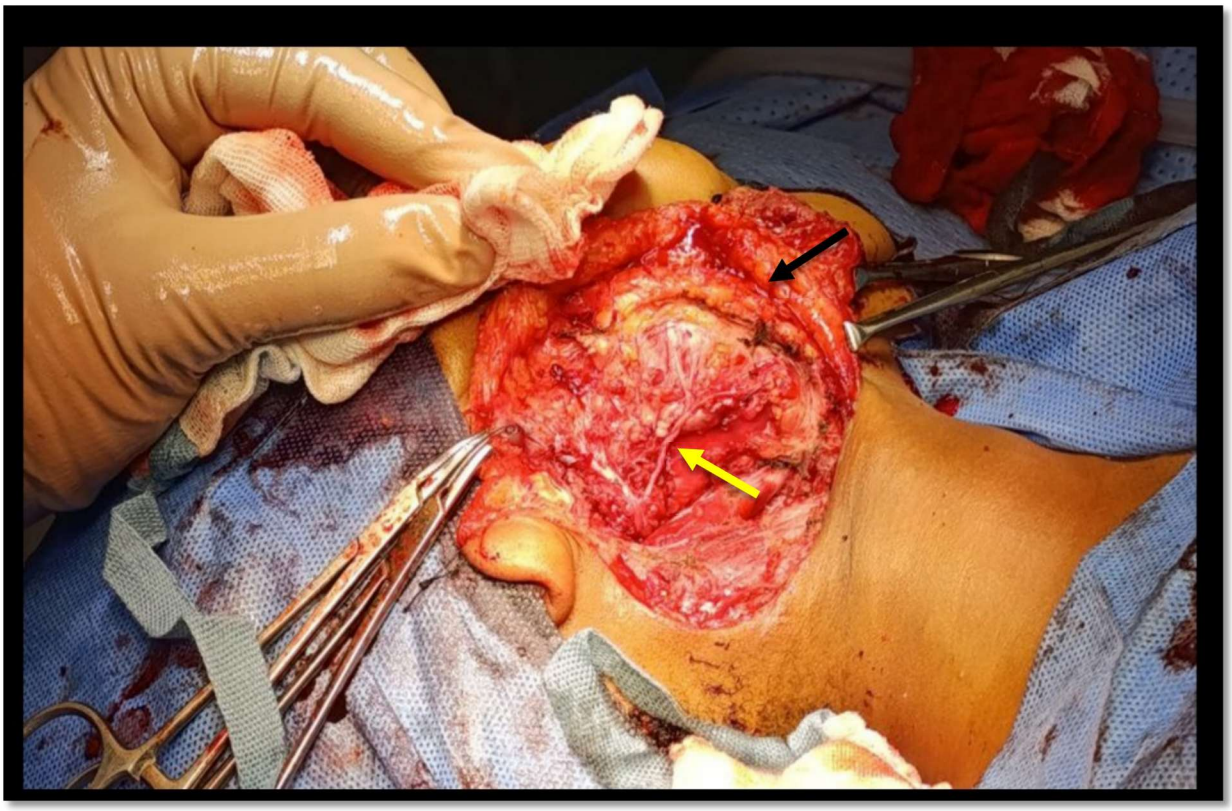

Fig 2. Demonstrating superficial cervicofacial skin flap (black arrow); and the facial nerve and its branches (yellow arrow)

Fig 3. Postoperative images at 1 month follow-up demonstrating that there are no clinical signs suggestive of an iatrogenic facial nerve injury

DISCUSSION

Malignant epithelial salivary gland neoplasms are rare in children, with the parotid gland being the most common site. The predominant histopathological subtypes in this age group are low or intermediate grade mucoepidermoid carcinoma, followed by acinic cell

carcinoma and adenoid cystic carcinoma. In children, a firm salivary gland swelling warrants a higher index of suspicion for malignancy than in adults, particularly when associated with pain or facial palsy. Diagnostic workup should include a thorough clinical examination, ultrasonography, magnetic resonance imaging (MRI), and in selected cases, CECT scan to assess the extent of the neoplasm and presence of metastasis. Fine needle aspiration (FNA) cytology remains a safe and minimally invasive diagnostic tool, though accuracy depends heavily on operator expertise and pathologist interpretation. The primary role of FNA is to distinguish benign from malignant lesions, with definitive subtyping deferred to excisional histopathology.

Prognosis in pediatric salivary gland malignancies is generally favorable. Adverse outcomes are associated with high grade neoplasms, positive surgical margins, and perineural invasion. Adjuvant radiotherapy can improve locoregional control but carries significant long-term risks in children, including growth disturbances, osteoradionecrosis, trismus, dental abnormalities, visual impairment, and secondary malignancies. Chemotherapy is typically reserved for recurrent or palliative settings. Our patient underwent a total parotidectomy with lymph node dissection following multidisciplinary discussion. An (R_0) resection was achieved, and histology confirmed a low grade mucoepidermoid carcinoma. Postoperative recovery was unremarkable, and the patient remains under surveillance. This case underscores the importance of early suspicion, comprehensive evaluation, and surgical management to achieve optimal outcomes in pediatric salivary gland malignancies.

CONCLUSION

Mucoepidermoid carcinoma is exceptionally rare in the pediatric population, particularly in children under 10 years of age. Surgical resection remains the cornerstone of treatment, with adjuvant radiotherapy considered in aggressive or high-grade cases. While the overall prognosis in young patients is favorable, clinicians must weigh the benefits of radiotherapy against potential long-term consequences, including craniofacial growth disturbances and the risk of radiation induced secondary malignancies. Survival outcomes in pediatric patients appear broadly comparable to those with mucoepidermoid carcinoma as a secondary neoplasm.

REFERENCES

1. Boukheris H, Curtis RE, Land CE, Dores GM. Incidence of carcinoma the major salivary glands according to the WHO classification, 92 to 2006: a population-based study in the United States. Cancer Epidemiol Biomarkers Prev 2009;18:2899–2906.
2. Heinrich Iro, Johannes Zenk. GMS Current Topics in Otorhinolaryngology - Head and Neck Surgery 2014, Vol. 13, ISSN 1865-1011.
3. Hicks J, Flaitz C. Mucoepidermoid carcinoma of salivary glands in children and adolescents: assessment of proliferation markers. Oral Oncol. 2000;36:454–60.
4. Lack EE, Upton MP. Histopathologic review of salivary gland tumors in childhood. Arch Otolaryngol Head Neck Surg.1988;114:898–906.
5. Pastore G, De Salvo GL, Bisogno G, et al. Evaluating access to pediatric cancer care centers of children and adolescents with rare tumors in Italy: the TREP project. Pediatr Blood Cancer. 2009;53:152–155.
6. Surveillance, Epidemiology, and End Results (SEER) Program (www.seer.cancer.gov) SEER*Stat Database: survival-SEER17 Regs Limited-Use, Nov 2008 Sub (1973–2006) <Katrina/Rita Population Adjustment> - Linked To County Attributes – Total U.S., 1969–2006 Counties, National Cancer Institute, DCCPS,Surveillance Research Program, Cancer Statistics Branch, released April 2009, based on the November 2008 submission.
7. Védrine, Pierre Olivier, et al. "Mucoepidermoid carcinoma of salivary glands in the pediatric age group: 18 clinical cases, including 11 second malignant neoplasms." *Head & Neck: Journal for the Sciences and Specialties of the Head and Neck* 28.9 (2006): 827-833.
8. Thorvaldsson SE, Beahrs OH, Woolner LB, Simons JN. Mucoepidermoid tumors of the major salivary glands. Am J Surg 1970;120:432–438.
9. Orvidas LJ, Kasperbauer JL, Lewis JE, Olsen KD, Lesnick TG. Pediatric parotid masses. Arch Otolaryngol Head Neck Surg. 2000 Feb;126(2):177-84. DOI: 10.1001/archotol.126.2.177.
10. Callender DL, Frankenthaler RA, Luna MA, Lee SS, Goepfert H. Salivary gland neoplasms in children. Arch Otolaryngol Head Neck Surg. 1992 May;118(5):472-6. DOI:10.1001/archotol.1992.01880050018003.
11. Orvidas LJ, Kasperbauer JL, Lewis JE, Olsen KD, Lesnick TG. Pediatric parotid masses. Arch Otolaryngol Head Neck Surg. 2000 Feb;126(2):177-84. DOI: 10.1001/archotol.126.2.177.
12. Ryan JT, El-Naggar AK, Huh W, Hanna EY, Weber RS, Kupferman ME. Primacy of surgery in the management of mucoepidermoid carcinoma in children. Head Neck. 2011 Dec;33(12):1769-73.DOI: 10.1002/hed.21675.
13. Sultan I, Rodriguez-Galindo C, Al-Sharabati S, Guzzo M, Casanova M, Ferrari A. Salivary gland carcinomas in children and adolescents: a population-based study, with comparison to adult cases. Head Neck. 2011 Oct;33(10):1476-81. DOI:10.1002/hed.21629.
14. Kupferman ME, de la Garza GO, Santillan AA, Williams MD,Varghese BT, Huh W, Roberts D, Weber RS. Outcomes of pediatric patients with malignancies of the major salivary glands. Ann Surg Oncol. 2010 Dec;17(12):3301-7. DOI: 10.1245/s10434-010-1165-2.

15. Ryan JT, El-Naggar AK, Huh W, Hanna EY, Weber RS, Kupferman ME. Primacy of surgery in the management of mucoepidermoid carcinoma in children. Head & neck. 2011 Dec;33(12):1769-73.

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