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MANAGEMENT AND RECONSTRUCTION OF RECURRENT SIALOBLASTOMA AND ITS COMPLICATIONS IN A PEDIATRIC PATIENT: A MULTIDISCIPLINARY APPROACH

Case Study

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Conflict of Interest: None
Grant Support & Financial Support: None

ABSTRACT

Background: Sialoblastoma is an exceedingly rare congenital epithelial tumor of salivary gland origin, predominantly affecting infants. It exhibits aggressive behavior with potential for recurrence and requires a multidisciplinary approach for management. This case report highlights the challenges of diagnosing and treating recurrent sialoblastoma in a pediatric patient, emphasizing innovative reconstructive techniques.

Case Presentation: A 2-year-old South Asian boy presented with a progressively enlarging left cheek swelling since infancy. Initial imaging and cytology suggested a salivary gland neoplasm, leading to total parotidectomy. Histopathological analysis confirmed sialoblastoma. Despite adjuvant chemotherapy, local recurrence occurred, requiring further surgical intervention, including hemimandibulectomy. Postoperative radiotherapy was administered; however, complications such as wound dehiscence and tumor recurrence necessitated additional surgical and reconstructive measures.

Intervention and Outcomes: The patient underwent a multidisciplinary surgical approach involving lesion excision, removal of the exposed mandibular plate, and mandibular reconstruction using a pedicled osseomyocutaneous pectoralis major flap with the sixth rib. Six-month follow-up showed no recurrence, with satisfactory functional and cosmetic outcomes.

Conclusion: This case underscores the aggressive nature of sialoblastoma, the importance of complete tumor excision, and the utility of innovative surgical reconstruction techniques. Multidisciplinary management and vigilant long-term follow-up are crucial for improving outcomes in pediatric sialoblastoma.

Keyword: Sialoblastoma, Salivary Gland Neoplasms, Pediatric Tumors, Multidisciplinary Treatment, Mandibular Reconstruction, Chemotherapy, Adjuvant, Recurrence

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INTRODUCTION

Sialoblastoma is an exceedingly rare congenital tumor originating from the epithelial anlage of the salivary gland. Predominantly localized in the parotid gland, it may also involve submandibular glands, minor salivary glands, or even ectopic salivary gland tissue (1, 2). Once classified as benign, the growing body of evidence has led to its reclassification as malignant (3, 4). Given the rarity and complexity of sialoblastoma, a multidisciplinary team (MDT) approach is essential for accurate diagnosis and effective treatment. This report details the case of a 4-year-old boy with recurrent sialoblastoma who underwent surgical resection and reconstruction using a pectoralis major osteomyocutaneous flap, highlighting the challenges and outcomes of managing this condition.

CLINICAL SUMMARY

A 2-year-old South Asian boy presented to a tertiary care hospital with a progressively enlarging swelling on the left cheek, initially noticed at 5 months of age. On examination, the lesion measured 4.5 × 4.0 cm. Magnetic resonance (MR) imaging revealed a 5.0 × 4.5 × 4.0 cm lesion within the superficial and deep lobes of the left parotid gland, showing isodense signals on T1W and T2W sequences. Fine needle aspiration cytology (FNAC) confirmed a salivary gland neoplasm but was inconclusive regarding its specific type. Following MDT evaluation, the patient underwent a left total parotidectomy. Histopathological examination revealed a multilobulated and multinodular tumor composed of predominantly round to oval cells with uniform nuclei and scattered mitotic figures. Immunohistochemical analysis showed positivity for Cytokeratin CAM 5.2, favoring the diagnosis of sialoblastoma over epithelial-myoepithelial carcinoma.

Postoperatively, the patient received six cycles of chemotherapy with a VAC regimen comprising vincristine, cyclophosphamide, and actinomycin-D. After three chemotherapy cycles, a computed tomography (CT) scan identified a minimally enhancing 5.8 × 7.0 mm soft tissue nodule in the parotid region with calcific foci, as well as heterogeneously enhanced left submandibular gland involvement. MDT discussions led to continued chemotherapy with reassessment planned post-treatment. However, a follow-up CT scan revealed persistent and slightly increased lesion size (6.0 × 7.0 mm) and low-volume cervical lymphadenopathy. Subsequent imaging at 3-month intervals demonstrated interval progression, with the lesion reaching 12.0 × 7.4 mm and bilateral cervical lymphadenopathy. A positron emission tomography (PET) scan revealed a soft tissue density with mild FDG avidity (SUV max 1.9) adjacent to the ramus of the left mandible. No hypermetabolic cervical lymph nodes were noted.

The patient was lost to follow-up and presented 5 months later with a large facial swelling. Imaging showed a 5.6 × 3.9 × 4.0 cm mass involving the left masseter and pterygoid muscles, with destruction of the ramus of the left mandible and extension into the pterygopalatine and infratemporal fossae. MDT evaluation led to a second surgery involving left hemimandibulectomy with stabilization using a titanium mandibular plate. Histopathology confirmed residual sialoblastoma involving the bone and lingual soft tissue. Postoperative management included 20 cycles of radiotherapy. The postoperative course was complicated by wound dehiscence, leading to exposure of the mandibular plate. Follow-up CT imaging demonstrated a poorly delineated left parapharyngeal mass (36.8 × 25.6 mm) with necrotic areas extending into the temporal fossa, indicative of tumor recurrence.

MULTIDISCIPLINARY MANAGEMENT AND RECONSTRUCTION

The patient presented to the maxillofacial team with an exposed mandibular plate and parapharyngeal mass. MDT discussions involving maxillofacial and plastic surgery teams resulted in a collaborative operative plan. Surgical intervention included resection of the lesion, removal of the mandibular plate, and mandibular reconstruction using a pedicled osteomyocutaneous pectoralis major flap incorporating the sixth rib. Histopathological evaluation of the resected specimen showed no residual malignancy.

OUTCOMES AND FOLLOW-UP

At the 6-month postoperative follow-up, the patient demonstrated no evidence of recurrence or wound dehiscence. The reconstructed mandible was stable, with the titanium implant functioning well. Cosmetic outcomes were deemed satisfactory, with significant improvement in facial symmetry and functionality.

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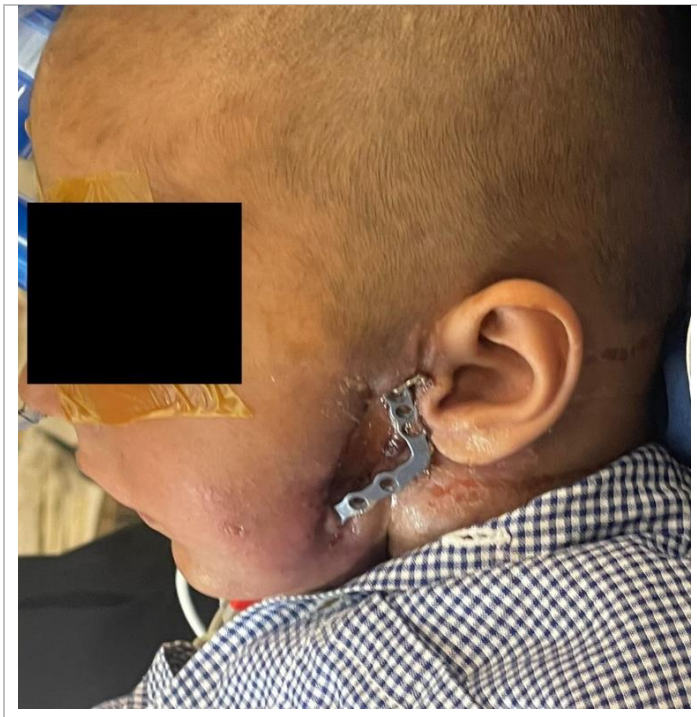
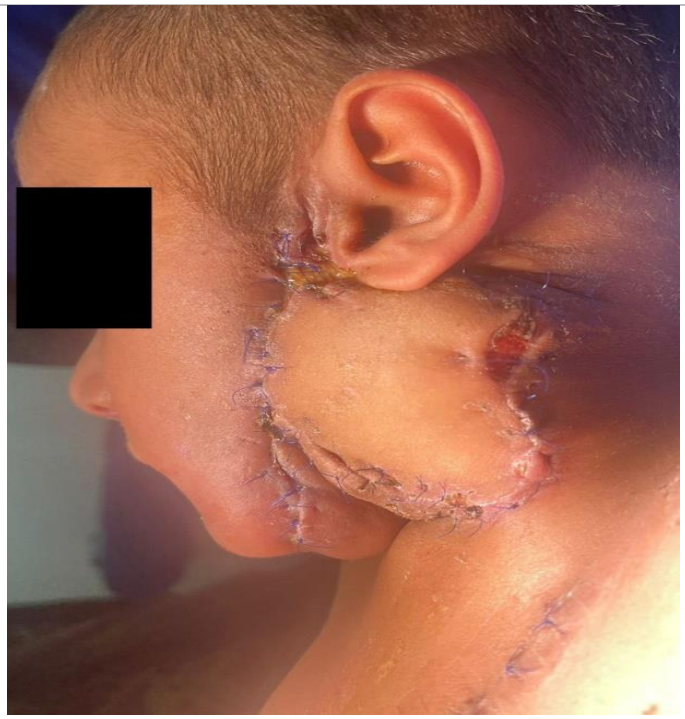
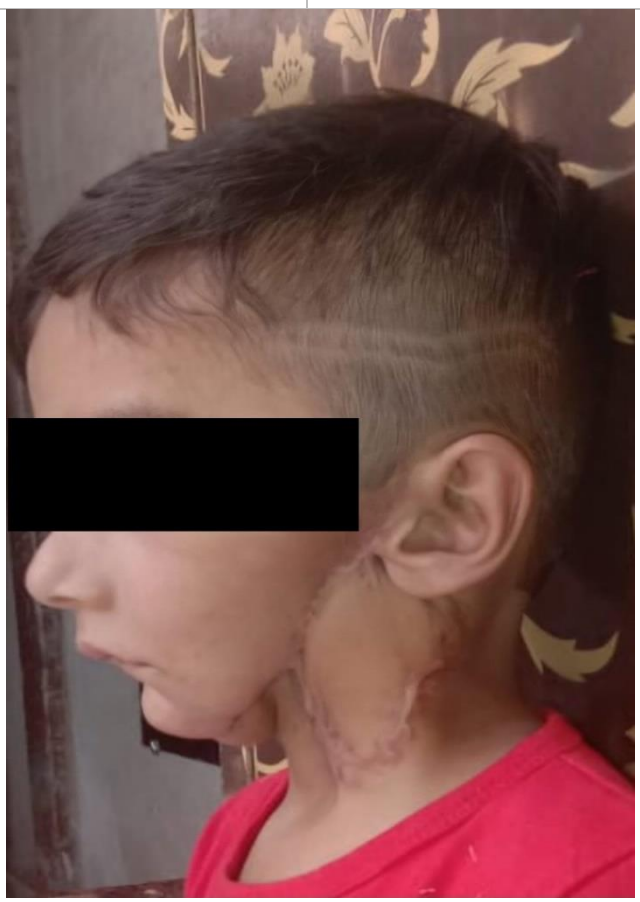


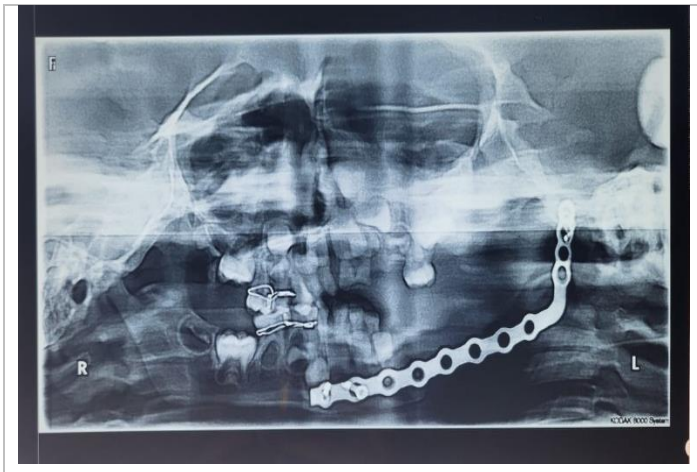
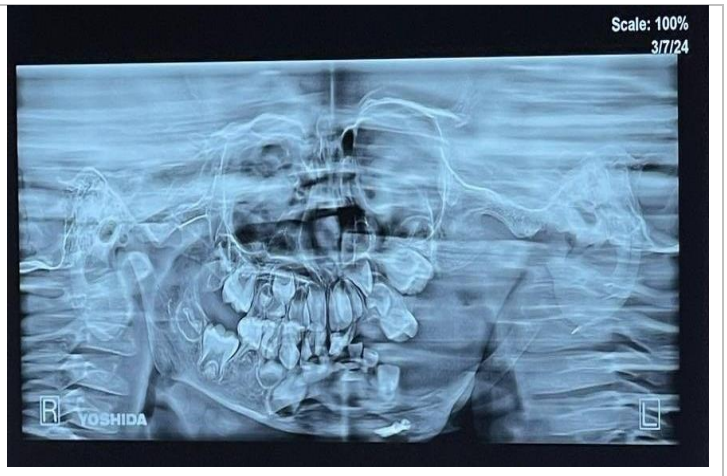
Pre-operative picture



Post-operative Picture



6 months follow up picture

	
Pre-operative OPG	Post-operative OPG

DISCUSSION

Primary salivary gland tumors in infancy and childhood are exceedingly rare, with non-epithelial tumors being more frequently encountered (5, 4). Sialoblastoma, a rare epithelial neoplasm of salivary gland origin, primarily affects the parotid and submandibular glands but can also involve minor salivary glands (6). Most cases are diagnosed shortly after birth, with the median age at diagnosis reported to be 9.8 months (7). The clinical presentation typically includes a slow-growing, painless subcutaneous mass, making early detection challenging. Microscopically, sialoblastoma is characterized by primitive histological features resembling early salivary gland tissue. These tumors consist of basaloid epithelial cells arranged in nests or cords, often with variable stromal components ranging from myxoid to fibrous. Approximately one-third of cases exhibit a cribriform growth pattern, where elongated and branching ductal structures are embedded within basaloid cell clusters (5, 2). Macroscopically, these tumors often appear as multilobulated, firm, tan-pink to yellow masses (1, 2). The differential diagnosis includes adenoid cystic carcinoma and basal cell adenoma, with sialoblastoma distinguished by its propensity for invasive behavior, including vascular, perineural, and skeletal muscle invasion, which sets it apart from benign salivary tumors (3).

Currently, there are no standardized treatment guidelines for sialoblastoma due to its rarity. However, surgical excision with tumor-free margins is considered the cornerstone of treatment, as enucleation alone is inadequate even for apparently well-circumscribed tumors (3). The roles of radiotherapy and chemotherapy remain controversial. Radiotherapy may be considered in cases of incomplete excision, though its use in pediatric populations is limited by the risk of long-term adverse effects (8, 3). Chemotherapy is typically reserved for cases with extensive or metastatic disease, recurrence, or incomplete surgical excision, with neoadjuvant chemotherapy being a potential option for locally invasive tumors. Long-term follow-up using imaging modalities such as CT and MRI is crucial for monitoring recurrence or metastasis. A multidisciplinary team (MDT) approach is essential to ensure optimal management and minimize complications. In the present case, the patient underwent initial surgical resection without confirmation of negative margins, followed by six cycles of adjuvant chemotherapy. Local recurrence shortly after treatment completion highlights the aggressive nature of sialoblastoma and underscores the importance of achieving complete tumor excision. The delayed involvement of an MDT likely contributed to suboptimal initial management. Subsequent recurrence necessitated a second surgical intervention, including hemimandibulectomy and reconstruction.

The early age of onset, with symptoms presenting at five months, makes this case particularly unique. The initial biopsy results were inconclusive, reflecting the diagnostic challenges of sialoblastoma. Definitive diagnosis relied on comprehensive histopathological evaluation and immunohistochemistry, emphasizing the need for meticulous diagnostic processes. Despite undergoing chemotherapy and radiotherapy, disease progression was observed, reiterating the tumor's aggressive behavior and propensity for recurrence. The innovative use of a pedicled osseomyocutaneous pectoralis major flap, incorporating the sixth rib for mandibular reconstruction, addresses both functional and cosmetic challenges. This technique effectively managed extensive tissue involvement and provided a durable reconstructive solution for the non-healing wound in the parotid region. Overall, this case underscores the complexities associated with sialoblastoma management, including early onset, diagnostic difficulties, aggressive disease behavior, and the need for individualized, multifaceted treatment strategies. Surgical resection remains the primary modality for achieving disease control, even in cases of recurrence and metastasis. The patient's long-term remission following surgical resection and innovative reconstruction highlights the importance of tailored management approaches and ongoing surveillance in pediatric patients with sialoblastoma.

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CONCLUSION

Sialoblastoma is a rare and aggressive pediatric salivary gland tumor requiring a multidisciplinary approach. Early diagnosis, complete surgical excision, and individualized treatment strategies are critical. Innovative reconstructive techniques enhance outcomes in complex cases. Vigilant long-term follow-up is essential to monitor recurrence and ensure optimal functional and cosmetic recovery in affected patients.

Author	Contribution
Aiza Bint e Shafqat	Conceptualization, Methodology, Formal Analysis, Writing - Original Draft, Validation, Supervision
Ehtesham-Ul-Haq	Methodology, Investigation, Data Curation, Writing - Review & Editing
Sana Rasheed	Investigation, Data Curation, Formal Analysis, Software
Ahmed Ibrahim	Software, Validation, Writing - Original Draft
Muhammad Ali Hassan	Formal Analysis, Writing - Review & Editing
Muhammad Abdullah Yousaf	Writing - Review & Editing, Assistance with Data Curation

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