Sialoblastoma: A rare submandibular gland neoplasm

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Abstract
A sialoblastoma is a rare congenital epithelial tumor that arises in a major salivary gland. To our knowledge, only 24 cases of sialoblastoma have been previously reported in the English-language literature. We report a new case, that of a 15-month-old boy who presented with a submandibular mass. Surgical excision of the mass was undertaken. Intraoperatively, the mass appeared to be adjacent to the submandibular gland, but it had not invaded it. The mass was excised, and the submandibular gland was left in place. Pathology identified the tumor as a sialoblastoma. However, pathology also revealed that residual tumor was present at the surgical margin. The patient was returned to the operating room for excision of the left submandibular gland and the level I lymph nodes. Following revision surgery, the surgical margins were negative. The patient remained disease-free at the 1-year follow-up. Despite the need for revision surgery, this case provides support for the idea that surgery alone is sufficient for curative treatment.

Introduction
Fewer than 5% of all salivary gland tumors occur in children, and fewer than 0.25% are found in children younger than 10 years.1,2 Salivary gland tumors account for 8% of all pediatric head and neck tumors.3 Most tumors of the salivary gland are nonepithelial masses, usually hemangiommas or lymphangiomas.4 Some 50% of all solid salivary gland tumors in children are neoplastic.5 Sialoblastomas are rare congenital epithelial tumors of the major salivary glands. To the best of our knowledge, only 24 cases have been previously reported in the English-language literature.2,4,6-9 Of the 23 cases in which the tumor’s site of origin was identified, 18 occurred in the parotid gland and 5 in the submandibular gland. In 1966, Vawter and Tefft described the first cases of a salivary gland tumor of epithelial origin, which they called an embryoma.10 Since then, various terms have been used to describe this tumor, including congenital basal cell adenoma, basaloïd adenoma, and congenital hybrid basal cell adenoma–adenoid cystic carcinoma.6 In 1988, Taylor suggested the term sialoblastoma to convey both the dysontogenic character and salivary gland origin of these tumors.11

Case report
A 15-month-old boy was brought to our hospital with a 3-month history of left-sided neck swelling. The patient’s mother denied fevers, chills, weight loss, other masses, exposure to tuberculosis, exposure to cats, and recent illness. Physical examination revealed that the swelling had been caused by a firm, nontender, nonerythematous, 2-cm mass in the area of the submandibular gland. Cranial nerve examination demonstrated no deficits. Computed tomography (CT) of the neck with intravenous contrast revealed that a 3 × 2-cm homogenous soft-tissue mass, with minimal rim enhancement, had arisen from the submandibular gland (figure 1).

The patient was taken to the operating room for excision of the mass. Intraoperatively, the mass appeared to be adjacent to the submandibular gland, but it had not invaded it. The mass was excised, and the submandibular gland was left in place.

Pathologic examination revealed a well-demarcated cellular neoplasm that was surrounded by a thin, fibrous pseudocapsule (figure 2, A). The tumor was made up of primitive basaloid cells arranged in nests and trabeculae that were separated by a fibromyxoid stroma. Many of the tumor nests contained multiple cystic spaces that were filled with basophilic mucoid material, which gave rise to a cribriform appearance. The cells were relatively monomorphic. They had high nucleus-to-cytoplasm ratios and round-to-oval nuclei with irregular nuclear membranes that featured folds and grooves. The nucleolar chromatin was finely dispersed, and the nucleoli were inconspicuous. However, a few scattered cells contained larger pleomorphic nuclei and more prominent nucleoli; their cytoplasm was scant and mildly eosinophilic. The tumor cells at the periphery...
of the nests and trabeculae were arranged in a vaguely palisading pattern. Mitotic figures were easily seen, as mitotic rates were as high as 5 per 10 high-power (×40) fields. A few ductular structures scattered through the tumor were lined with cuboidal-to-columnar epithelial cells with lower nucleus-to-cytoplasm ratios. Focal perineural invasion by the tumor nests was present (figure 2, B). These findings were consistent with a diagnosis of sialoblastoma.

Pathology also revealed that tumor was present at the surgical margin. The combination of a positive margin, the presence of perineural invasion on histopathology, the neoplasm’s locally aggressive behavior, and the possibility that it might metastasize to regional lymph nodes led us to perform a second surgical procedure. The second operation included excision of the left submandibular gland and the level I lymph nodes. Histopathology following this procedure revealed no neoplasm.

Surveillance T2-weighted magnetic resonance imaging (MRI) 1 year later revealed the presence of a hyperintense 2.2 × 1.6-cm soft-tissue lesion at the site previously occupied by the left submandibular gland and the excised lesion. In response to this finding, the child underwent a third procedure to biopsy the area of concern. Histopathologic examination of the biopsy specimen revealed that it was consistent with a benign reactive lymph node.

Discussion
Sialoblastomas are locally invasive and have a propensity for recurrence, but no case of fatal metastasis has been reported. In fact, only 1 case of regional lymph node involvement has been reported. The earliest sialoblastoma has been identified is at 37 weeks of gestation; the diagnosis was made by intrauterine ultrasonography. After birth, sialoblastomas are best detected by MRI and CT. Luna reported that MRI of a sialoblastoma in a 21-month-old child revealed that the lesion was isointense with muscle on T1-weighted imaging, that it had a high-to-intermediate signal intensity similar to that of fat on T2-weighted imaging, and that it enhanced sparsely and nonhomogenously.

Histologically, sialoblastoma can be diagnosed with simple hematoxylin and eosin (H&E) staining; specimens characteristically demonstrate solid nests of epithelial cells that are separated from a nearly fibrous and fibrofribrous tissue. The epithelial cells may be basoloid with scant-to-normal cytoplasm and either solitary or very few nucleoli. In addition, ducts and sometimes acini with occasional central necrosis are seen in the surrounding stroma. Acini have been reported to stain positive for mucin by the diastase/periodic acid–Schiff method. Vascular invasion has been noted in at least 1 report. Immunohistochemistry has demonstrated cytokeratin in the ductal components, vimentin in both the ductal structures and solid nests, and S-100 protein, which confirms the presence of myoepithelial cells.

It is widely held that primary excision with negative margins is sufficient for cure with these tumors. Although distant spread has not been reported, local recurrence has occurred in at least 5 cases. In addition, there has been 1 case (nonfatal) of regional lymph node metastasis. Only 3 cases have warranted postoperative radiation and chemotherapy. The first two were described...
in the original report by Vawter and Tefft, and the other involved a child who had experienced four recurrences that required additional resections, including one of the orbital floor and maxilla. In most reports in the literature, follow-up has not exceeded 2 years, although Vawter and Tefft described 1 patient who was disease-free 9 years postoperatively.

The case we describe herein provides further evidence that surgical excision without postoperative chemo- or radiotherapy is sufficient for curative treatment. Although our patient’s follow-up MRI at 1 year demonstrated an area of concern, he was in fact disease-free.

Acknowledgments

The authors thank Denesh Rakheja, MD, a pathology fellow at Children’s Medical Center of Dallas, for his help in describing the pathologic findings of this case, and Tim Booth, MD, a staff radiologist at the Children’s Medical Center of Dallas, for his assistance with reading the CTs and MRIs.

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