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Diagnostic and Therapeutic Approach to Sialoblastoma of Submandibular Gland: A Case Report

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Salivary gland tumors are rare in childhood and account for 3% to 5% of all tumors in children.[1-7] Congenital salivary epithelial tumors are characterized by some as medical curiosities.[8-12] In 1966, Vawter and Tefft[13] reported 2 cases in neonates and used the term *embryoma* to describe them. Since then, a variety of names have been used to describe histologically similar or identical tumors that, retrospectively, are virtually indistinguishable from one another.[1] In 1988, Taylor[14] suggested the term *sialoblastoma* to describe these lesions because it conveyed the dysontogenetic character and the site of the tumor in a single name; this has become the preferred term. Because of its rarity, the biologic behavior of this lesion is incompletely defined. Early surgery is the recommended treatment.[5-10]

This article presents a rare case of sialoblastoma of the submandibular gland that occurred in a 6-month-old girl, including the natural history of the tumor, its morphology, and its management.

Report of a Case

A female infant was born at full term and without complications. At the time of her first "well child" checkup, she was noted to have a right submandibular mass that measured approximately 3 × 3 cm (Fig 1A). At the age of 6 months, the mass had enlarged slightly but remained nontender. Ultrasonographic scan of the area showed mixed pattern nodular areas with well-defined margins and a predominantly hypoechoic pattern (Fig 1B).

During surgery, the mass was found to be well-encapsulated but not distinct from the submandibular gland. Both were excised, and a number of enlarged lymph nodes were removed (Fig 2A). The nodular circumscribed mass (Fig 2B) was diagnosed as a sialoblastoma. Histologically, tissue sections showed a tumor made up of cellular islands of anastomosing epithelial cells, cribriform sheets, and trabeculae embedded in a fibromyxoid stroma (Fig 3). Abundant tortuous duct formations and distinct foci of disorganized acinar differentiation were observed. In most cellular islands, small epithelial ductular structures were lined by cuboidal cells and open lumina with secretions (Fig 4).

Immunohistochemically, ductular structures were positive for low-weight cytokeratins, whereas the basaloid-type cells of the islands were protein S-100 positive, with a peripheral rim that was positive for actin. No further therapy was provided. The child is doing well and showed no evidence of recurrence 12 months after surgical resection.

Discussion

Only a few cases of perinatal epithelial salivary gland tumor have been reported.[6,12-16] Among those in which the gland of origin was identified, a total of 4 tumors arose in the submandibular gland. This report describes a case of sialoblastoma of the submandibular gland.

Since the time of Vawter and Tefft (1966), a variety of terms have been used to describe perinatal epithelial salivary tumors, including *congenital basal cell adenoma, basaloid adenoma, congenital hybrid basal cell adenoma,* and *adenoid cystic carcinoma.*[8-16] In a retrospective review, Hsueh and Gonzalez-Crussi[10] considered that these tumors were histologically similar or identical. The "salivary gland unit" theory of Batsakis hypothesizes that neoplasia within salivary glands results from the activation and proliferation of resting multipotent reserve cells.[8,9] Taylor[14] attributed the histogenesis of congenital tumors to the disordered proliferation of blastemous cells rather than to the activation of resting reserve cells, suggesting that the term *sialoblastoma* conveys both the dysontogenic character and the salivary origin of these tumors.

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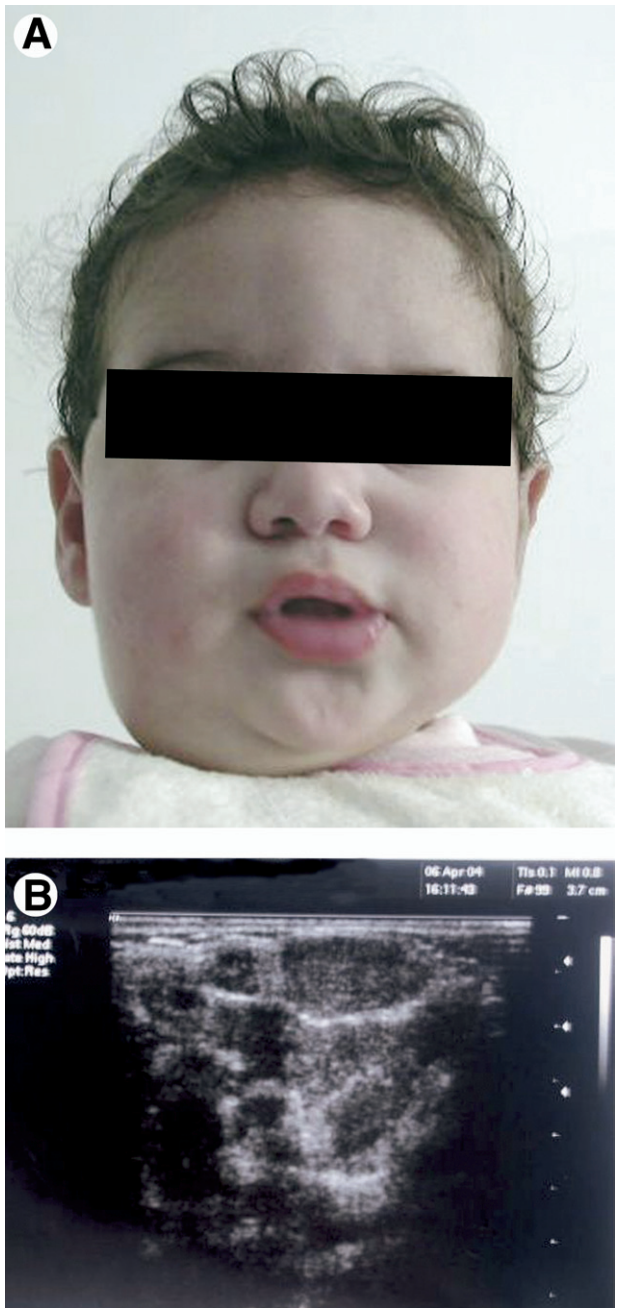

FIGURE 1. *A* and *B*, A female child was noted to have a right submandibular mass that measured approximately 3 × 3 cm. Ultrasonography of the area showed nodular areas with well-defined margins characterized by a mixed pattern, with prevalence of a hypoechoic pattern.

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Batsakis has classified perinatal salivary gland tumors into the following 4 categories[8,9]:

1. Histologically benign tumors comparable with adult counterparts (eg, pleomorphic and monomorphic adenomas).
2. Hamartomatous tumors.
3. Tumors that are histologically reminiscent of the embryonic epithelial anlagen of major salivary glands at various stages of development (eg, sialoblastoma).
4. Tumors that are biologically and histologically malignant.

Batsakis also proposed histologic criteria for the assessment of malignancy in sialoblastoma; these included "invasion of nerves or vascular spaces and ancillary findings of necrosis of cells with cytological atypia beyond that expected or presumed for an embryonic epithelium."[8]

However, these histologic determinations should be considered in the clinical context of tumor stage

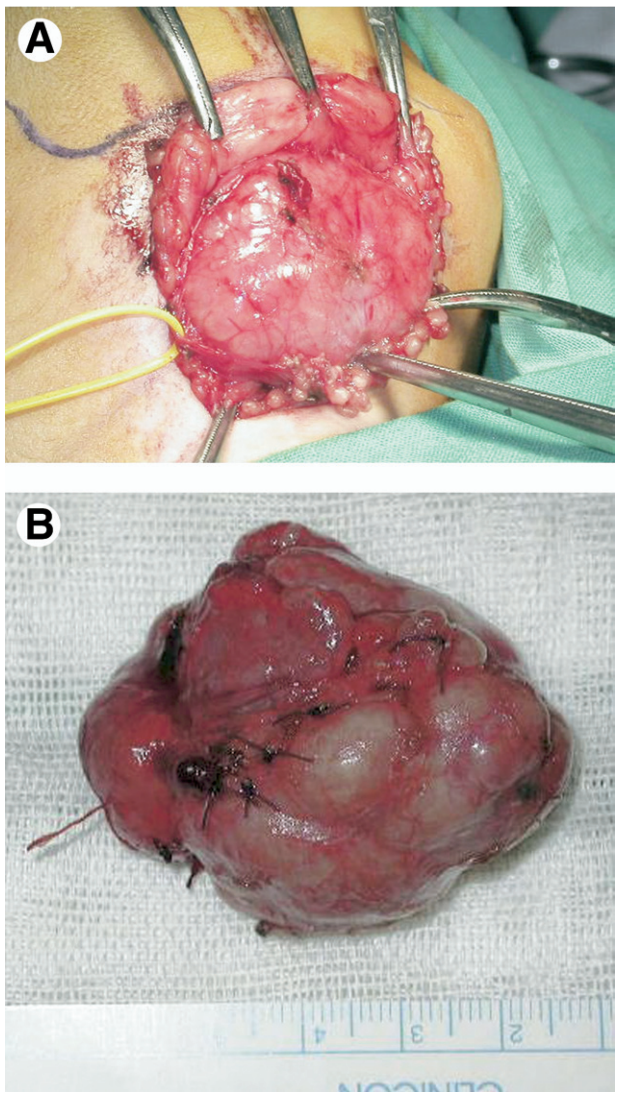

FIGURE 2. *A*, The mass was dissected free and removed carefully. The nodular circumscribed mass (*B*) was found to be well-encapsulated.

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and effectiveness of resection. As Brandwein et al pointed out,[12] the distinction between benign and malignant sialoblastomas may not be as well-defined as was thought originally.

Examination of the admittedly limited number of reported patients with sialoblastoma reveals that local recurrence is of primary concern. Of 24 reported cases, including those referred to here, no deaths due to disseminated disease have occurred.[9-12] However, local recurrence has been documented in at least 5 patients, and in 1 case, regional lymph nodes were positive for metastasis.[10-12]

Surgery remains the treatment of choice without irradiation or chemotherapy. A review of the literature on sialoblastomas showed that a few cases recurred locally, 1 case had regional lymph node involvement, and in only 1, distant metastases were reported.[1-4,15,16]

Radiation therapy may be considered if lesions are not completely resectable, but the adverse effects of radiotherapy may be severe for growing facial structures.[16] Chemotherapy may be chosen when the tumor is not completely resectable or in cases of persistent or recurrent tumor.[16]

It is not clear from the limited literature why these lesions tend to have local recurrences; however, even though they are considered locally aggressive, they are not believed to be malignant. In our case, the tumor was excised with a narrow margin. No adjuvant treatment was given, and no recurrence has been noted after 18 months—a fact that confirms the notion that sialoblastoma rarely and slowly tends to metastasize. Reported nodal involvement in sialoblastoma does not necessarily indicate malignant behavior because benign salivary gland tumors of embryonic origin with nodal involvement have been reported.[13-15]

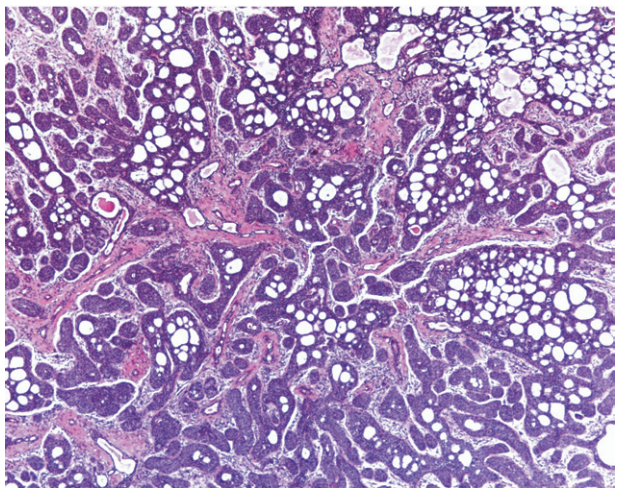

FIGURE 3. Anastomosing trabeculae and cribiform sheets composed of cytologically bland and basaloid cells in a fibromyxoid stroma.

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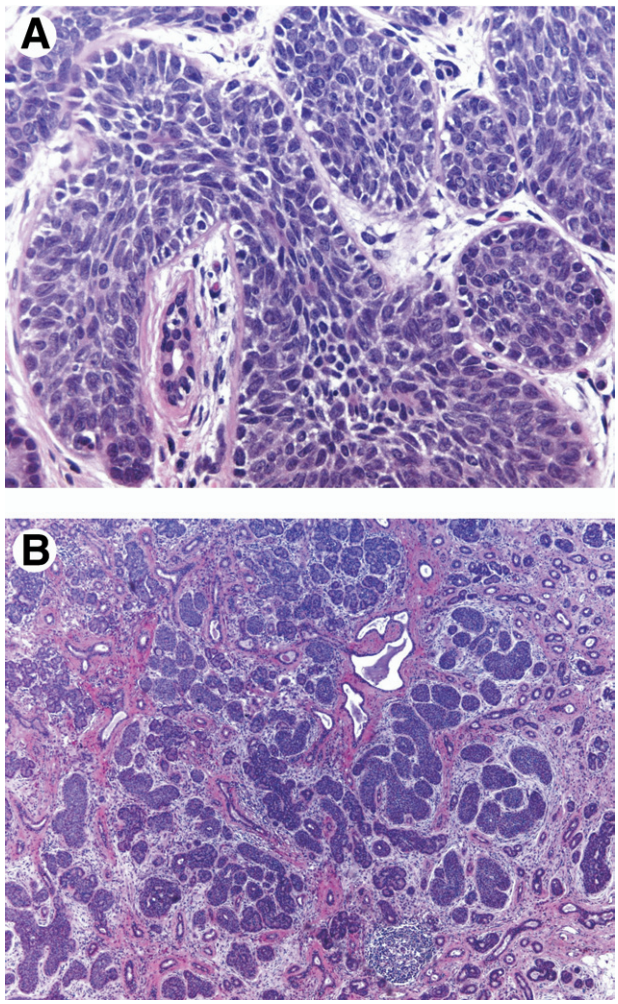

FIGURE 4. *A* and *B*, Tortuous duct formations and distinct foci of disorganized acinar differentiation. In most of these cellular islands, small epithelial ductular structures were lined by cuboidal cells and had open lumina with secretions.

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Sialoblastomas are very rare neonatal tumors of the salivary gland that are primarily parotid in origin. They are locally aggressive, rarely metastasize, and should be treated surgically first. Radiotherapy and chemotherapy are indicated only in selected cases.

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Maxillary Anterior Segmental Advancement of Hypoplastic Maxilla in Cleft Patients by Distraction Osteogenesis: Report of 2 Cases

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Maxillary hypoplasia is a common deformity in patients with cleft lip and palate. Le Fort I osteotomy is one of the routine procedures for treatment of maxillary hypoplasia in cleft palate patients. In recent years, rigid external and internal distraction systems have gained popularity for improvement of severe maxillary hypoplasia. However, these techniques have the risk of velopharyngeal insufficiency by increasing the nasopharyngeal distance. For this reason, advancement of anterior maxillary segment by distraction osteogenesis can be thought of as an alternative method that offers many advantages.

In cleft lip and palate patients, early surgical corrections may result in poor skeletal and dental growth in the transverse and sagittal planes, especially in the maxilla.[1] Maxillary advancement greater than 6 mm is often difficult to achieve in this group of patients because of maxillary scarring. A mean postoperative relapse of 20% to 25% has been documented with conventional orthognathic surgery.[2]

Distraction osteogenesis (DO) is a recent addition to the treatment modalities for reconstructing severe facial deformities. It was first used for correction of the craniofacial skeleton in the early 1990s.[3] Rachmiel et al[4] achieved maxillary advancement with DO in adult sheep, and an increasing number of studies have been reported about the advancement of the maxilla or midface region.[4,5] Polley and Figueroa used adjustable rigid external distraction (RED) devices to advance the maxilla in children with cleft lip and palate and reported successful results.[6] Today, RED systems and internal distractors are the most popular techniques for advancing the hypoplastic maxilla in cleft palate patients. Some limitations of the treatment are reported, such as exter-

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