Facial Desmoid Tumor in a 2-Year-Old

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A 2-year-old male presented to the emergency department with a rapidly growing right facial mass. Imaging revealed a 6 × 4 cm enhancing mass centered within the right parapharyngeal space with extensive local tissue compression and osseous remodeling (Figure 1). The patient underwent transoral biopsy that demonstrated both spindle and rhabdoid cells with mitotic figures. Immunostaining was positive for desmin, actin, and β-catenin, suggesting a desmoid tumor.

Given the extent of the tumor and its location, the patient was initiated on a weekly regimen of vincristine and methotrexate due to concerns for serious cosmetic and functional morbidity with surgery. Over the next 4 weeks, there was an interval increase in size by ~25%, primarily at the biopsy site, and the patient became increasingly symptomatic with intermittent bleeding and episodes of upper airway obstruction. This prompted surgical intervention by a multidisciplinary team comprised of oral-maxillofacial surgery (OMFS), otolaryngology—head and neck surgery, and microvascular reconstructive surgery (OMFS). Reconstruction was discussed at length, and a decision was made to defer definitive bony reconstruction because of growth concerns and lack of an adequate anatomic replacement for the mandibular ramus at this age. The reconstructive team was available in the instance that the oral soft tissue defect would require a free tissue to close.

A model of the tumor in situ was created using 3-dimensional reformatted images, which was then used to prebend a reconstruction plate for surgery. Complete resection was subsequently achieved by composite mandibulectomy via combined transcervical and lip-split approach. After endoscopic evaluation of the airway, a tracheostomy was placed. The tumor was first approached at its lower border via a transcervical incision, which allowed for identification of cranial nerves and vascular structures. Next, the anterior and inferolateral aspects of the tumor were dissected, maintaining a musculoperiosteal margin over the tumor. The distal mandibulotomy was then performed at approximately mid-body, allowing lateralization of the mandible, and the right parapharyngeal space was widely exposed giving access to the ipsilateral skull base. The tumor was then mobilized from its superior attachments to the skull base and pterygoid plates through a maxillary vestibular extension of the anterior ramus incision. Given the limited vertical growth of the pediatric lower and mid-face, the entire vertical compartment of the mandible was easily accessible through this approach, eliminating the need to use the coronal extension. The defect was temporarily reconstructed using the previously mentioned mandibular reconstruction plate and condylar prosthesis (Figure 2). Intraoral closure was obtained.

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Figure 1. Preoperative imaging demonstrating extent of tumor. Axial computed tomography (CT: A) and coronal T2-weighted magnetic resonance imaging (MRI: B). Three-dimensional reformatted imaging (C) and model (D) used for surgical planning. (*) represents mass; arrow marks thinning of mandibular ramus secondary to tumor.
and the buccal fat pad was allowed to prolapse into the dead space, suspended deep to the oral closure.

At completion of surgery, the patient’s face was symmetric with complete oral competence (Figure 3). The postoperative course was uncomplicated with no evidence of recurrence at 1 year. His maximal incisal opening and speech are excellent. He continues to do well, is taking an oral diet, and is pending decannulation in the near future.

Desmoid fibromatosis is a benign but locally aggressive neoplasm originating from musculoaponeurotic structures throughout the body. These tumors are rare and account for 0.03% of all neoplasms. Approximately 25% occur in children younger than 15 years, with 15% arising in the head and neck. The most commonly affected site is the mandible, followed by the submandibular area, neck, tongue, and paranasal sinuses.1

A detailed review of the literature identified only 19 reports of desmoid tumors in the pediatric head and neck, highlighting the rarity of these lesions and the lack of consistent data regarding their management. Surgical resection is the treatment modality of choice. However, given the tendency of the tumor cells to interdigitate with muscle fibers, complete surgical excision is difficult. In one retrospective review of 97 cases, of patients who underwent primary surgery, 16% experienced a local recurrence.1 Margin status affects recurrence with 30% of those with positive margins experiencing a recurrence compared to 2% of those with negative margins,3 the majority of which presented in the first 2 years.2-4 As a primary modality, radiation is rarely used in pediatric patients. A report on a series of pediatric patients treated with radiation alone documented local recurrences in 10 of 13 patients, with 8 patients experiencing severe radiation-associated complications.5 Chemotherapy may be tried for slower growing tumors, where methotrexate and vinblastine are favored due to fewer cytotoxic effects.6 Any form of intervention for these tumors has to take into consideration cosmetic deformity, functional morbidity, and long-term consequences, especially in the pediatric patient. In adults, adjuvant radiation may decrease recurrence in cases with microscopic residual disease. In children, the morbidity of chemoradiation often outweighs the potential benefit; thus, there is a paucity of clear recommendations for these adjuvant treatments.

This report emphasizes a multidisciplinary approach to the treatment of a complex parapharyngeal space tumor in a child. The patient was followed by genetics, pediatric, and oncology services from time of initial diagnosis through the postoperative period. He underwent a major surgery where 3 surgical teams worked in tandem to secure the airway, extirpate the large tumor, and reconstruct the mandible to preserve function and cosmesis. The surgery was performed expeditiously, with each surgical team assigned a defined role. Preoperative planning with reconstruction models shortened time spent under anesthesia and also allowed for more accurate family counseling. Continued close follow-up for local recurrence is critical, and for pediatric patients—the additional ongoing concern of facial growth will need to be addressed into late adolescence. The complex anatomy of the head and neck makes complete resection of these neoplasms without significant morbidity challenging, and at times impossible. We describe a successful approach to the management of an aggressive tumor with an excellent functional and cosmetic outcome.

Declaration of Conflicting Interests
The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Figure 2. Intraoperative photographs demonstrating modified Blair incision extending into the transcervical crease (A, black arrows) and placement of prefabricated reconstruction bar (B, white arrows).

Figure 3. Postoperative photographs, frontal (A) and side views (B) with well-approximated lip-split incision (white arrows).
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