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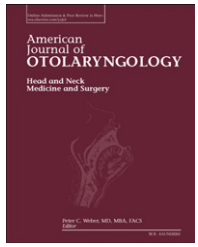

Pediatric desmoid fibromatosis of the parapharyngeal space: A case report and review of literature☆,☆☆

Zhong Zheng, MD[a,*], *Adrienne C. Jordan, MD*[b], *Alyssa M. Hackett, MD*[a], *Raymond L. Chai, MD*[a,c]

[a] *Department of Otolaryngology, New York Eye and Ear Infirmary of Mount Sinai, New York, NY, USA*
[b] *Department of Pathology, New York Eye and Ear Infirmary of Mount Sinai, New York, NY, USA*
[c] *Institute of Head, Neck and Thyroid Cancer, Mount Sinai Beth Israel, New York, NY, USA*

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ABSTRACT

Desmoid fibromatosis, or aggressive fibromatosis, is a benign but locally infiltrative fibroblastic neoplasm arising from fascial or musculoaponeurotic tissues. Although lacking metastatic potential, head and neck fibromatosis can have significant functional or cosmetic morbidities. 7%–15% of all desmoid tumors are seen in the head and neck region, 57% of which occur in the pediatric population. The incidence of pediatric desmoid tumor peaks around age 8. Treatment of choice is complete surgical resection; however, local recurrence is common. We present a case of a 14-month-old male with an 8-cm desmoid tumor in the right parapharyngeal space and provide an overview of diagnosis and management of pediatric head and neck fibromatosis. This is the largest desmoid tumor of the parapharyngeal space in the youngest patient described in the literature.

1. Introduction

Desmoid fibromatosis, also known as aggressive fibromatosis, is a neoplastic monoclonal proliferation of fibroblasts, with an incidence of 2 to 4 per million per year [1]. The incidence peaks at 8 years of age, as well as in the third or fourth decades of life [2]. It is most commonly classified as intra-abdominal, abdominal wall, or extra-abdominal. Intra-abdominal fibromatosis is associated with familial adenomatous polyposis (FAP) and Gardner's syndrome. Head and neck desmoid fibromatosis represents 7%–15% of all cases [1]. It has a tendency to be locally invasive but does not metastasize. Nevertheless, it can be associated with significant functional and cosmetic morbidities, attributed to the intricate anatomy of the head and neck region.

The most common presenting symptom of head and neck fibromatosis is an enlarging painless mass [5]. The mandible is the most commonly affected location, followed by the

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* *Corresponding author at:* Department of Otolaryngology, New York Eye and Ear Infirmary of Mount Sinai, 310 East 14th Street, New York, NY 10003, USA. Tel.: +1 212 979 4545.

E-mail addresses: zzheng@nyee.edu (Z. Zheng), adjordan@nyee.edu (A.C. Jordan), ahackett@nyee.edu (A.M. Hackett), rchai@chpnet.org (R.L. Chai).

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submandibular area, neck, tongue, and paranasal sinuses, respectively [3,9]. Two cases of parapharyngeal fibromatosis have been described in the literature [2,5]. CT and MRI are imaging modalities of choice and are valuable in assessing extent of disease and involvement of vital structures. Imaging findings are variable based on cellularity and collagen content of lesions [1,6]. Definitive diagnosis often requires incisional biopsies, as fine needle aspirations and core biopsies are frequently inconclusive [3]. Histologically, lesions comprise uniform-appearing spindle cells with intervening collagenous stroma and without dysplastic features. Positive β-catenin staining is characteristic in fibromatosis, but it is not disease specific [1,6]. Treatment of choice is surgical excision with clear margins; however, local recurrence is common, ranging from 13% to 47% [1]. Complete resection is often challenging due to involvement of major neurovascular structures of the head and neck and a tendency for lesions to spread along fascial planes without a capsule. The association between positive margin status and local recurrence has been debated in the literature [1–3,5,8,12]. Preservation of function and avoiding unnecessary morbidity should be given a high priority. Adjuvant radiotherapy or chemotherapy may be valuable in controlling microscopic or macroscopic residual disease [1–3,5]; however, interpretation of data is limited by heterogeneity and rarity of the disease.

2. Case presentation

A 14-month-old male presented with a 1-month history of a rapidly enlarging painless right neck mass, preceded by blunt cervical trauma from a 2-feet fall. The patient was evaluated by his pediatrician and treated with amoxicillin without improvement prior to initial presentation. On exam, a firm and fixed mass was noted over the right angle of the mandible, without any associated cervical lymphadenopathy or facial nerve dysfunction. Initial ultrasound revealed a 5.5 × 3.3 × 4.4 cm heterogeneous but solid appearing mass with high internal vascularity and ill-defined borders. On CT scan, a 5.5 × 4.2 × 3.3 cm mass was seen inseparable from the right parotid gland, abutting the stylomastoid foramen and traversing the stylomandibular tunnel. The mass was mildly hyperintense to skeletal muscle but hypointense to the parotid gland. Mass effect was demonstrated on the cartilaginous external auditory canal, the condylar head of the mandible anterosuperiorly and carotid sheath structures posteriorly. There was no carotid encasement or cervical lymphadenopathy (Fig. 1).

The patient underwent incisional biopsy and histology revealed a bland spindle cell neoplasm with collagenous stroma. The specimen stained positive for beta-catenin and Vimentin but was negative for SMA, S-100, and Desmin (Fig. 2). Preliminary diagnoses included nodular fasciitis versus fibromatosis, and the pathological specimen was sent to an outside institution for further review. There was significant interval growth after initial incisional biopsy and the decision was made to proceed with total parotidectomy and parapharyngeal space dissection for definitive resection. Intraoperatively, the mass was found to encase the main trunk of the facial nerve at the stylomastoid foramen. After identifying all distal branches, the facial nerve was meticulously

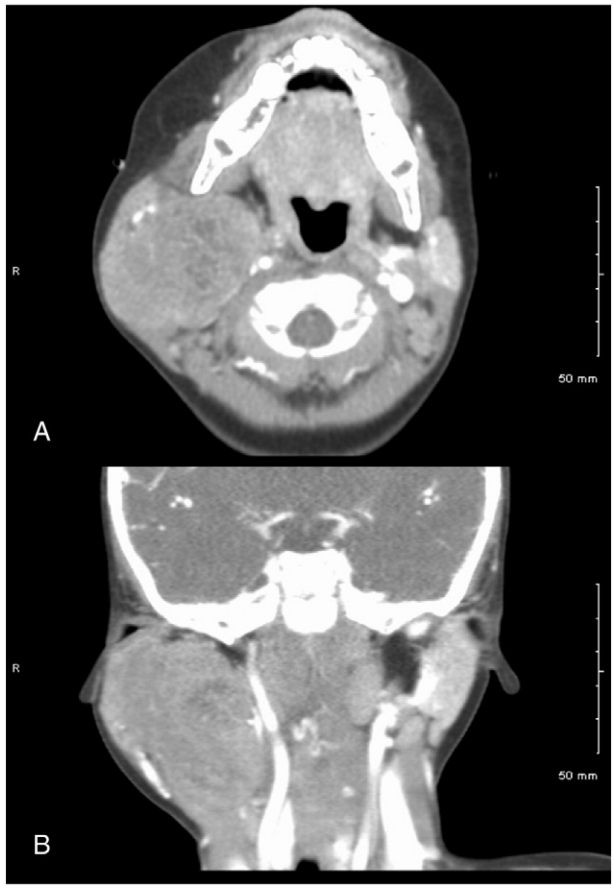

Fig. 1 – Axial (A) and coronal (B) CT images of the right parapharyngeal mass. The mass is slightly hyperintense to muscle and hypointense to the parotid gland, traversing the stylomandibular tunnel into the right parapharyngeal space. The carotid artery is displaced medially.

dissected to the main trunk in a retrograde fashion. A small cuff of tumor was left to preserve facial nerve continuity. A near-total gross resection of tumor was performed (Fig. 3). Electrophysiological response was observed with stimulation of the upper and lower divisions of the facial nerve distal to the pes anserinus, but not at the main trunk; however, the facial nerve was intact on gross inspection. A diagnosis of desmoid fibromatosis was confirmed on final pathology. In the immediate post-operative period, the patient had a complete facial nerve paralysis, likely secondary to neuropraxia. His facial nerve function recovered to a House–Brackmann Grade II with good eye closure 1 month post-operatively. The sole deficit noted at this time was residual marginal mandibular nerve weakness. The patient was referred for pediatric medical oncology evaluation to discuss possible adjuvant chemotherapy. Serial MRI scans are planned for continued surveillance.

3. Discussion

Head and neck desmoid fibromatosis is a rare soft tissue neoplasm of fibroblasts with a propensity for local recurrence. It represents 7%–15% of all cases and may be considered a distinct