Reconsidering Surgery in Pediatric Head and Neck Desmoid Tumors

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BACKGROUND

Desmoid fibromatosis and desmoplastic fibroma are rare benign lesions characterized by their locally invasive nature and absence of metastatic potential. While these tumors are infrequent in children, they are often found in the head and neck region. Most relevant series in the head/neck present heterogenous treatment approaches across multiple centers over large time periods. Due to a reported 30-50% recurrence rate irrespective of surgical margins and the morbidity of complex head and neck resections in children, treatment paradigms have varied widely with recent emphasis on non-operative management and targeted therapy. We report our relatively large surgical experience in pediatric head and neck desmoid tumors treated by a single team over a 6-year period with an emphasis on surgical decision making.

PURPOSE

This review aims to present our relatively large surgical experience in pediatric head and neck desmoid tumors over a 6-year period, with a focus on surgical decision making.

METHODS

We conducted a retrospective analysis of pediatric patients diagnosed with head and neck desmoid tumors who underwent surgical intervention at our institution by a single pediatric head and neck tumor team from 2016 to 2022. Data on patient demographics, tumor characteristics, surgical procedures, adjuvant therapies, and outcomes were collected and analyzed

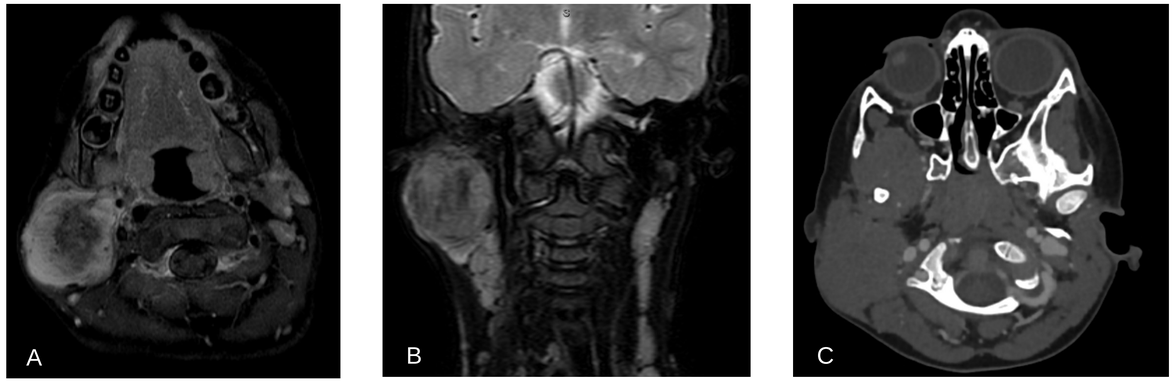

Fig 1: Patient 1 Imaging Characteristics. . A. Axial T1 and B. coronal fat-suppressed T2 MRI with contrast, show heterogeneous lesion centered in the right posterior triangle between the right parotid gland, sternocleidomastoid muscle and parapharyngeal space, measuring 4.1 x 3.1 x 4 cm (AP, transverse and craniocaudal dimensions). C. Axial post-contrast CT, show lesion displaces the right parotid gland anteriorly and deforms the posterior aspect of the right mandible at the level ramus with a thin rim of surrounding fat with low attenuation.

Table.1 Summary of patient characteristics

Patient No.	Age (Yr)& Gender	Tumor site/Size	CTNNB1 (Beta-Catenin) mutation	Treatment			Recurrence	Follow up (Yr) (outcome)	Comment
				Pre op	Surgical Margins	Post op			
1	3/F	Parotid extend to Parapharyngeal space / 4cm	Positive	CT (DD)	Positive	CT (DD)	No	1.2 with NED	
2	2/M	Scalp / 4 cm	Positive	No	Negative	No	No	2.2	
3	2/F	Mandible / 4.5 cm	Positive	No	Positive	No	No	6.41	
4	3/F	Mandible / 3.5 cm	Positive	CT (VLB/MTX)	Positive	No	Yes; twice after 7 month and after 15 months both required CT and surgical resection	3.8 from last surgical resection	CT before 2nd resection with DD. CT before 3rd resection with Hydroxyurea
5	1.5/F	Mandible / 6 cm	Positive	No	Negative	No	No	4.4	
6	18/F	Infratemporal Fossa and temporal bone / 6.5 cm	Positive	CT (DD)	Positive	No	No	1	CT started with VLB/MTX with progression
7	16/M	Mandible / 5 cm	Positive	CT (DD)	Positive	No	No	2	CT with Hydroxyurea With progression
8	2/F	Mandible / 5 cm	Positive	CT (DD)	Positive	No	No	1.4	
9	1/M	Maxilla / 4.7 cm	Positive	CT (DD)	Positive	No	No	1	CT started with MTX/VLB progression
10	8/F	Mandible / 4 cm	Negative	No	Negative	No	No	2.3	

CT: Chemotherapy, DD: Decarbazine and Doxorubicin), MTX/VLB: Methotrexate/ Vinblastine

RESULTS

In this study, a total of 10 patients (6F:4M) were treated for desmoid tumors with an average age of 5.62 years (1.1 to 17.7 years). The tumors sites included 5 in the mandible, 3 in the mandible and infratemporal fossa, 1 in the scalp, and 1 in the maxilla. Four underwent primary surgery with tumors isolated to the mandible body/ramus or scalp. The remaining 6 patients received neoadjuvant chemotherapy, either after an initial observation period or as part of a planned treatment strategy, followed by surgery. During the average follow-up period of 2.59 years (range 1.03 to 6.41, SD 1.7y), only 1 patient (10% of the total) experienced recurrences. The recurrences occurred at 7 months and again at 2 years, in separate subsites.

CONCLUSION

The management of pediatric head and neck desmoid tumors presents considerable difficulties given their invasive nature, tendency for recurrence, and the absence of a clear consensus on optimal treatment approaches. However, by employing oncologic surgical principles and ensuring a consistent and experienced medical team, it is possible to achieve lower recurrence rates compared to previous studies. It is important to view surgery as part of a comprehensive multidisciplinary treatment plan, integrating adjuvant therapies aimed at enhancing resectability while minimizing potential complications.

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