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CASE REPORT

Craniofacial/Pediatric

Mandibular Myofibroma and Severe Trismus: A Complex Case and Review of Complications

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Summary: A female child was investigated for insidious onset of temporomandibular joint dysfunction and trismus in the setting of a mandibular myofibroma. Myofibromas, benign mesenchymal neoplasms composed of spindle cells, are rarely found in the oral cavity, most commonly in the mandible. These lesions are historically described as indolent with a high cure rate and minimal recurrence rates following surgical resection. The patient initially presented with concerns regarding snoring, retrognathia, and jaw ankylosis, as well as a history of trouble latching as an infant but without obvious physical deformities. Imaging revealed a large expansile lytic mass of the mandible, but no temporomandibular joint involvement; surgical biopsy evidenced myofibroma, and the lesion was resected. Over the course of disease, the lesion continued to expand, and the patient's maximal incisal opening continued to decrease despite conservative management with jaw physiotherapy; eventually she could not open her mouth despite the absence of joint involvement. Re-exploration along with formal jaw physiotherapy was achieved and optimal jaw opening was maintained. Myofibromas are rare benign desmoid tumors that can present anywhere in the body in solitary and multicentric forms, and previously did not present significant challenges to surgical and medical management. Tumors of the mandible may present with trismus and soft tissue ankylosis, which can mimic temporomandibular joint dysfunction in the absence of joint involvement. Physical therapy, rehabilitation, and soft tissue contracture release are key to management and improving outcomes in oral cancer patients, regardless of tumor pathology. *(Plast Reconstr Surg Glob Open 2022;10:e4380; doi: 10.1097/GOX.0000000000004380; Published online 14 June 2022.)*

Myofibromas constitute benign mesenchymal neoplasms composed of spindle-cells.[1] Myofibromas may present as isolated lesions or as part of multicentric disease of soft tissue and viscera referred to as myofibromatosis.[2] While a majority of these lesions are described in the superficial tissues of the head and in skeletal muscle and aponeuroses, a minority of these lesions are found in the oral cavity, most commonly in the mandible.[3] The current literature describes these lesions as indolent with a high cure rate and minimal recurrence rate following surgical resection.[4,5] In this article, we report an isolated myofibroma of the mandible causing significant temporomandibular dysfunction and trismus in a pediatric patient.

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CASE REPORT

The patient is a White girl and was 4 years old during initial presentation in 2018. She presented at our institution with concerns regarding snoring, retrognathia, and jaw ankylosis possibly due to soft tissue pathology, as well as a history of trouble latching as an infant. She had no prior surgical history.

On physical examination, the patient was found to have a retrognathic mandible with an overjet of approximately 10 mm, mentalis strain, associated Class II malocclusion, and a maximal incisal opening of 20 mm. There was no obvious mass on physical examination. Initial computed tomography imaging showed a 3.6 × 3 × 4 cm large expansile lytic mass involving the right mandibular ramus (Fig. 1). The mass was producing significant thinning of the mandibular cortex nearly circumferentially but spared

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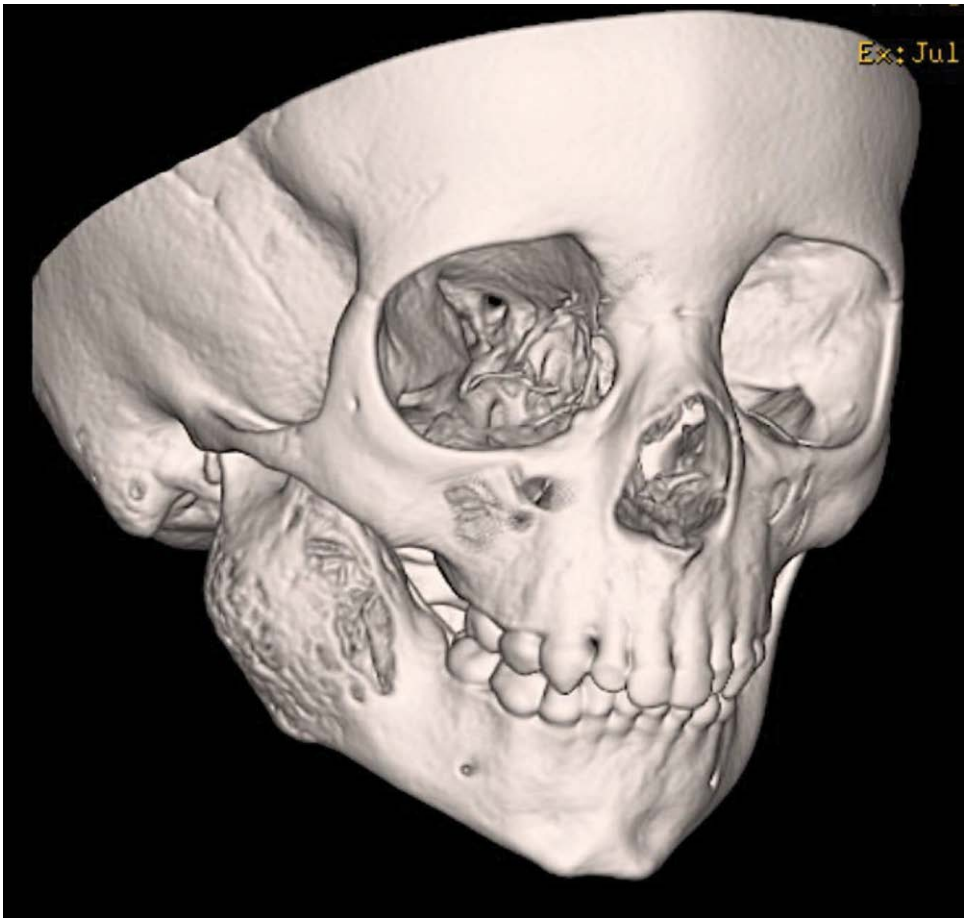

Fig. 1. Preoperative cranio-maxillofacial three-dimensional computed tomography reconstruction of the lesion.

the condyle. The patient then underwent TMJ manipulation and surgical biopsy, which showed a benign spindle cell neoplasm that was positive for smooth muscle actin, and negative for myogenin, desmin, CD34, S-100 protein, and beta-catenin. Shortly after she underwent complete resection, surgical pathology again evidenced a benign spindle cell neoplasm that was most consistent with myofibroma (Fig. 2).

Post-treatment surveillance showed a 3 × 2 × 2 cm lesion in the mandibular ramus, now with erosion and thinning of the remainder of the right mandibular condyle but no involvement of the condylar articular surface or glenoid fossa (Fig. 3). Of note, the patient's family endorsed minimal efficacy of TheraBite to increase patient's jaw opening. After initial recurrence, family discussions with surgical teams, oncologists, and tumor board were made to reach an agreement on expectant management of the lesion before re-operation, based on the benign tumor pathology.

In 2019, the patient had a repeat maxillofacial CT with 3D reconstruction, which showed that the lesion was stable. The patient returned later this year unable to open her mouth more than 10 mm. Magnetic resonance imaging of the patient showed progression of the lesion to 3 × 2 × 4 cm and extension into the condyle but not into the condylar head or articular surface, as well as extension into the coronoid process. There was no obvious bony or soft tissue destruction outside the mandible.

In 2020, the patient returned unable to open her mouth. The patient was referred to oncology before further surgical management. Pediatric oncology opted for genetic testing before treatment initiation, which found that the lesion was negative for platelet-derived growth factor receptor B and NOTCH3 mutations. Chemotherapy was declined in favor of conservative management with TheraBite to increase mouth opening. However at this stage, the patient's family reported minimal efficacy and compliance with the device.

In 2021, the patient returned with worsening of her clinical condition and could not open her mouth more than 5 mm. Repeat imaging showed normal temporomandibular joints and mild reduction in tumor size but now with pseudoarticulation of the lesion with a broad sclerotic styloid process from the inferior aspect of the skull. We proceeded with re-operation with a right coronoidectomy, soft tissue release and manipulation of the temporomandibular joint, exploration of the masticator space, and lysis and steroid injection of scar bands. We achieved jaw opening to 25 mm.

The 1 month postoperative visit showed a consistent 25 mm jaw opening. The patient was scheduled for formal physical therapy along with Therabite use at home. The

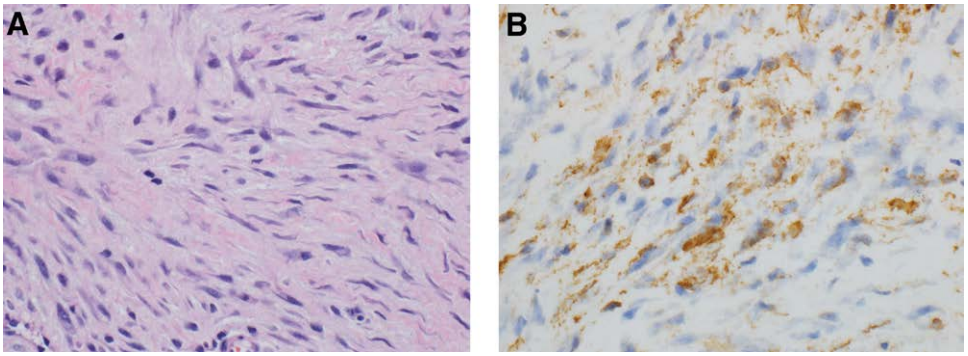

Fig. 2. Microscopic and histologic pathology slides of the lesion. A: H&E 40× view. B: smooth muscle actin staining 40× view.

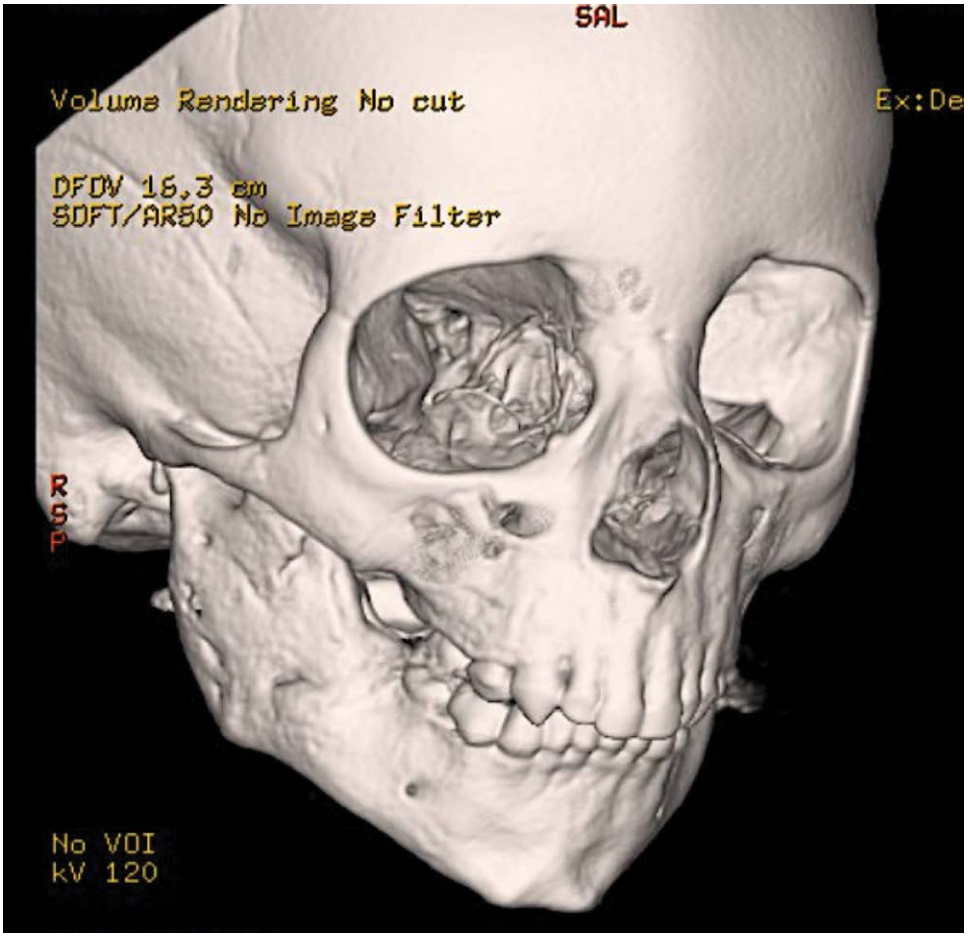

Fig. 3. Postoperative cranio-maxillofacial three-dimensional computed tomography reconstruction of the lesion.

patient continues to have three monthly visits to monitor the tumor and TMJ function.

DISCUSSION

Most reported cases of head and neck myofibromas show a benign growth pattern, and excellent cure rates with resection and minimal recurrence.[4,5] Our patient presentation suggests that this may not always be the case. Literature review shows that the only documented complication in patients with mandibular myofibromas is mental nerve hypesthesia in one patient, and this was likely present before surgical intervention.[6]

Etiology of trismus and restricted jaw range of motion in our case may be multifactorial. Mechanical obstruction at the zygomatic arch and/or coronoid process due to tumor mass effect may be contributory. As the patient's disease progressed, local inflammation and destruction of the temporalis muscle may be indicative of soft tissue

ankylosis. Another plausible explanation may be stiffness from disuse of the temporomandibular joint over the progressive course of the disease, which highlights the importance of jaw physiotherapy.

Trismus in oral cancer is a well-documented complication, and is usually secondary to chemotherapy and/or radiation therapy, which our patient did not have.[7] Extension of the lesion into the coronoid process was the basis of proceeding with coronoidectomy, which has been documented as a plausible solution to trismus and soft tissue ankylosis.[8] Regardless of operative management, data suggest that jaw physiotherapy is the cornerstone for optimizing jaw opening in oral cancer patients with trismus.[9,10] Noncompliance with TheraBite therapy in our patient suggests this as well. Additional research is needed on development of targeted medical therapies for myofibroma, and early intervention with jaw physiotherapy as well as an individualized patient approach may be key to optimize outcomes in surgical and nonsurgical cases.

In conclusion, myofibromas are rare benign desmoid tumors that can present anywhere in the body in solitary and multicentric forms. Tumors of the mandible may present with trismus and soft tissue ankylosis, which can mimic TMJ dysfunction in the absence of joint involvement. Physical therapy, rehabilitation, and soft tissue contracture release are key to management and improving outcomes in oral cancer patients, regardless of tumor pathology.

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