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CASE REPORT
Craniofacial/Pediatric

Total Resection of Pediatric Desmoid Tumor of the Left Neck with Utilization of 3D Virtual Surgical Planning

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Summary: A 13-year-old girl with a painful left neck mass was referred to our institution due to suspicions of malignancy. The patient reported pain that accompanied her frequent neck spasms. Computed tomography revealed a large, soft-tissue mass in the left neck, deep to the sternocleidomastoid. The lesion anteriorly displaced the internal carotid artery and both displaced and crushed the internal left jugular vein. Uniquely, a three-dimensional virtual reality model combining magnetic resonance imaging and computed tomography data was used to determine the lesion's resectability and visualize which structures would be encountered or require protection while ensuring total resection. During operation, we confirmed that the mass also laterally displaced the brachial plexus, cranial nerves X and XI, and spinal nerves C3–C5 (including the phrenic) of the cervical plexus. Postsurgical pathological analysis confirmed a diagnosis of desmoid tumor, also known as aggressive fibromatosis, whereas DNA sequencing revealed a *CTNNB1* mutation, a somatic genetic marker found in approximately 90% of desmoid tumor cases. When possible, the most widely used method for the treatment of desmoid tumors has been gross resection. Chemotherapy, radiotherapy, and local excision are also used in the treatment of fibromatoses when complete resection is judged infeasible. In this case, a complete surgical resection with tumor-free surgical margins was performed. A standard cervical approach with a modified posterolateral incision site was implemented to avoid a conspicuous anterior neck scar. No flap, nerve repair, or reconstruction was warranted. At 1 year of postsurgical follow-up, the patient showed minimal scarring and no signs of recurrence.
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Desmoid tumors are rare, nonmalignant lesions of fibroblastic proliferations within connective tissue.[1] Pediatric fibromatoses are often locally aggressive and invade into adjacent tissue walls and bone, making them difficult to fully expunge.[2,3] Still, some studies advocate for a "wait-and-see" approach that favors surveillance and conservative intervention dependent on tumor location, size, and growth rate.[2] Fibromatoses exhibit a tendency toward recurrence, with reports suggesting higher rates in patients who do not undergo complete excision, or for those for whom negative margins are impossible.[4] Pediatric anatomy, endocrine development, and continued growth present special surgical and therapeutic challenges. This case report describes the presentation and excision of a large, painful desmoid tumor in the left neck of a young girl. Strategic surgical planning was critical to navigating the vascular network and series of nerves affected by the tumor, and virtual modeling assisted in

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Fig. 1. A comparison of preoperative and postoperative patient images. A, Preoperative profile view of the patient taken 2 months after reported tumor development, demonstrating a large left neck mass. B, Postoperative profile view of the patient taken 5 months postoperatively, showing the successful absence of the large, painful, desmoid tumor.

determining the cervical approach and modified incision site chosen. One year postoperatively, the patient showed excellent recovery and is without recurrence.

CASE REPORT

A 13-year-old Hispanic girl presented to our clinic reporting a week-long history of neck spasms and a painful left neck mass (Fig. 1A, B). She denied prior infection, injury, or other symptoms. Computed tomography revealed a large, left neck, soft tissue mass deep to the sternocleidomastoid. The lesion dimensions measured 4.2×4.2×4.8 cm with encroachment upon the left side of the cervical airway and anterior displacement of the internal carotid artery and internal jugular vein (Fig. 2). Additionally, the patient experienced complete left jugular vein compression and displacement of the brachial plexus. Extension of the lesion marginated the left vertebral artery foramen at C3 and C4. Physical examination found multiple enlarged, less than 1.6-cm-sized, level-2 nodes within the cervical chain bilaterally.

Based on the diagnosis and radiological findings, a three dimensional (3D) virtual model of the desmoid tumor was used to plan a safe and precise surgical operation (Fig. 3). Scar minimalization was another major goal to maintain our patient's self-confidence. Thus, our team opted

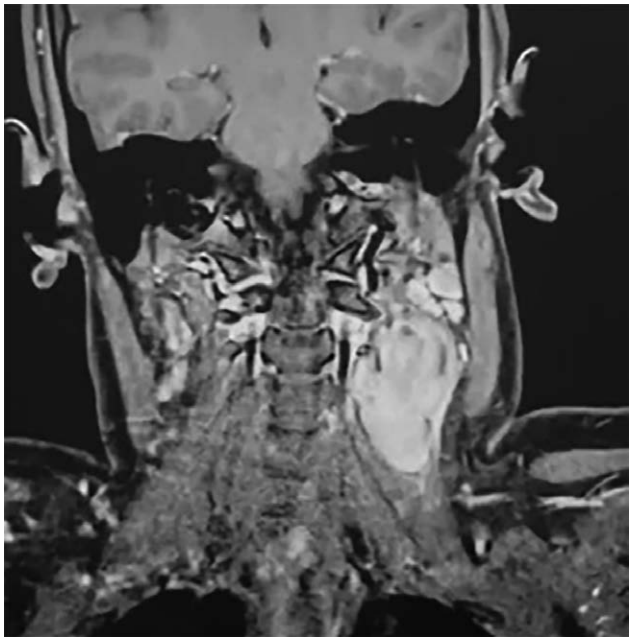

Fig. 2. Preoperative coronal MRI image showing the disruptive size of the large left neck mass deep to the sternocleidomastoid and flanking the C3 and C4 foramen.

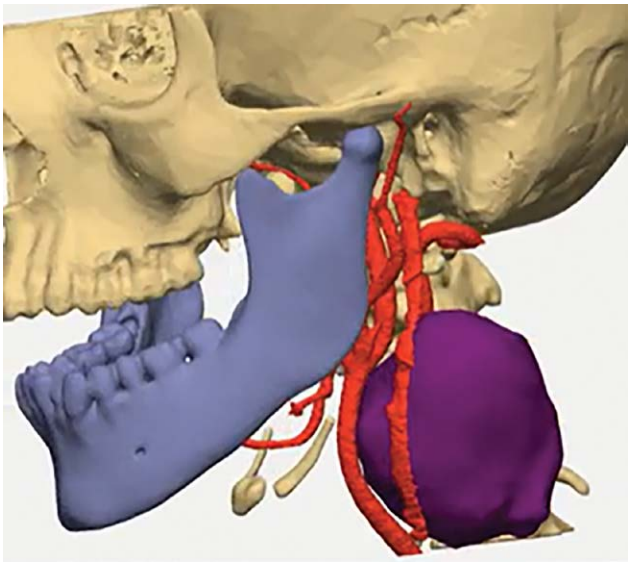

Fig. 3. Preoperative mapping of the patient's fibromatosis using a three-dimensional virtual reality model software by 3D Systems.

for a standard cervical approach with a modified infero-posterolateral incision site to reduce scar visualization and in sensitivity to this young woman's continuing development and body positivity. The excision was performed 10 days after imaging. Raising the subplatysmal flaps and laterally retracting the sternocleidomastoid revealed the tumor's superficial border. Intraoperative examination confirmed lateral displacement of the brachial plexus, phrenic, vagus, and spinal accessory nerves, along with internal jugular vein and internal carotid artery anterior displacement. Although the external jugular vein was lost, all other major structures remained intact, and no dead space was created, rendering reconstruction unnecessary. Extracapsular dissection enabled removal of the tumor as a whole specimen. Upon excision, the mass appeared dense and homogenous with a tan-pink, rubbery surface, and had changed in size to 6.2×6.0×4.0 cm from 4.2×4.2×4.8 cm in her magnetic resonance imaging 10 days prior.

Postoperatively, microscopic examination demonstrated a spindle cell neoplasm with high immunoreactivity for β-catenin, a desmoid tumor hallmark.[2] Negative family history alongside genetic testing confirming a *CTNNB1* mutation supported that the patient's lesion was somatic.[1] One year later, no muscular or nervous deficits were noted apart from slight numbness at the resection site. Neither signs of recurrence nor novel neoplasia have occurred.

DISCUSSION

Although benign, desmoid tumors can have destructive mass effects. In this adolescent's case, a large left neck mass displaced major neurovascular structures, causing her spasms and pain. Our team used a 3D virtual reality model for operative planning to navigate these delicate structures and leave an inconspicuous scar. The "3D Systems" software combines magnetic resonance imaging and computed tomography data to create a three-dimensional virtual reality model that can be manipulated digitally, helping us determine the lesion's resectability, visualize structures to be encountered or requiring protection, and achieve total resection with precise negative margins through the modified infero-posterolateral incision site. [**See Video 1 (online),** which shows a simulation of the patient's desmoid tumor in 3D Systems software, used to improve direct visualization of the target lesion and allow precise planning of the excision.] Beneficially, the model enabled planning of the entire resection in virtual space before the successful surgery, lending support to its utilization in other head and neck oncology and plastics cases.

Pediatric malignancies differ from those of adults due to their increased risk of long-term effects from cancer treatment. Because desmoid tumors are more aggressive in juveniles, early detection and total excision are key factors in foiling the potential damage of desmoid tumors' mass effects in the head and neck. Current literature indicates total resection (negative surgical margins) as the best intervention and recurrence prevention.[3,5] Still, desmoid head and neck tumors are not always resectable, and when they are, surgical procedures that functionally or aesthetically compromise a child's facial structure are not advisable.[5,6] Additionally, because recurrence is not exclusive to those who possess residual tumor, and no prospective studies have been conducted establishing predictive patterns of recurrence, current recommendations are to closely monitor patients postoperatively no matter the margins achieved.[2]

Other cases have approached surgical procedures conservatively with adjuvant chemotherapy and/or radiation regiments.[2,4,7] These decisions may be prompted by patients' individual presentations, but evidence is unclear regarding long-term consequences and efficacy of these modalities.[8] The literature shows that approximately 50% of desmoid tumor patients left with microscopically positive surgical margins experience recurrence, even when those patients preceded surgery with cytotoxic drugs.[9] The potential for harm, whether by disfigurement from overly wide surgical margins or by the risks conferred by exposure to DNA-damaging agents, may explain why some experts have advocated for a "wait-and-see" approach.[8,9] However, most argue that this strategy is more favorable for adult desmoids, which tend to be slower growing and allow for safer monitoring, than the more invasive pediatric desmoid tumor.[7,8,9]

Here, the clinical and diagnostic findings made surgical resection urgent and evident.

CONCLUSIONS

Our intervention applies modern virtual 3D surgical models during preoperative planning and adheres with the principles of total surgical resection with negative tumor margins, when possible, which our team found both possible and successful. We advocate for close, long-term follow-up to monitor for recurrence, and support mindfulness of the unique experiences and development of the pediatric patient when planning treatment and providing guidance throughout recovery.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

The patient provided written consent for the use of her image.

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