Am J Otolaryngol 40 (2019) 779–782

Contents lists available at ScienceDirect

Am J Otolaryngol

journal homepage: www.elsevier.com/locate/amjoto

The use of 3D printing in shared decision making for a juvenile aggressive ossifying fibroma in a pediatric patient

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ARTICLE INFO

Keywords:
Pediatric palate tumor
Juvenile aggressive ossifying fibroma
Juvenile ossifying fibroma
Ossifying fibroma

ABSTRACT

Juvenile aggressive ossifying fibromas (JAOF) are rare, typically benign pediatric tumors that are locally aggressive and have high recurrence rates. A 7-year old male presented with a palatal mass and a 3D printed model was created and used as a visual aide to highlight the importance of management in terms of functional, cosmetic, and disease-free outcomes with the family. The patient ultimately underwent successful enucleation with final pathology consistent with JAOF. To our knowledge, this is the first description of the use of 3D printing to help in the shared decision-making process for the treatment of this aggressive tumor.

1. Introduction

Juvenile aggressive ossifying fibroma (JAOF) is a rare, benign, but aggressive and recurring tumor that can lead to multiple surgeries and ultimately maxillofacial defects and deformities [1]. JAOF typically occurs in young children, age < 15 and are locally aggressive and destructive with report recurrence rates up to 30–50% [2]. Controversy exists as to the proper management of these locally aggressive benign tumors. Surgical resection remains the mainstay of treatment, but the extent of surgery remains unclear ranging from enucleation to local resection to radical excision [3–5]. As each treatment modality has its pros and cons that not only effect the patient, but their family as well, shared decision making becomes an important aspect in the JAOF patient's care.

The question becomes, how do we incorporate the patient and their families in the decision-making process? Part of the problem lies in helping the patient and their families to understand the complex nature of disease. This is particularly a major issue in surgical diseases. Visualizing the resulting defects associated with surgical resection and reconstruction options is a difficult task even for the surgeon themselves – how do we help those without a medical background to understand as well? The advent of 3D printing has provided an avenue in which both surgeons and patients can both learn and interact in meaningful ways to understand and plan treatment options. One study showed that 3D printing helped to facilitate a better understanding of complex anatomy compared to the use of computed tomography or computed tomography and digital reconstruction alone [9]. Furthermore, the use of 3D printed models in patients with glioma helped them to better understand their disease and situation and made it easier for them to ask questions to their physician and support their decision making about their treatment options [10]. Facilitating a patient and their families understanding of the disease helps in creating an active and interactive shared decision-making environment. Here we describe the first application of 3D printing in the shared decision-making and management of a juvenile aggressive ossifying fibroma and its successful management with surgical resection.

2. Case presentation

A 7-year-old male with no relevant past medical history was referred our otolaryngology clinic for evaluation of a palate lesion. He was seen by his pediatrician the week prior for new right sided ear pain

Abbreviation: JAOF, Juvenile aggressive ossifying fibroma

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https://doi.org/10.1016/j.amjoto.2019.07.001
Received 21 May 2019

Fig. 1. Preoperative 4.4 cm × 2.7 cm asymptomatic right hard palate mass.

and was found to have a right sided serous effusion. Additionally, a 4.4 cm × 2.7 cm asymptomatic right hard palate lesion was discovered. The palate mass was a solid, well circumscribed lesion extending into the nasal cavity and resulting in thinning of the involved hard palate bone (Fig. 1). On nasal endoscopy, there was leftward deviation of the inferior septum with a submucosal lesion involving the right inferior-posterior nasal cavity. The lesion superiorly displaced the right inferior turbinate. A contrast enhanced CT of the face was performed. CT showed an expansile, hypodense lesion of the right side of the hard palate extending into the right nasal cavity (Fig. 2A). There was scalloping of the contours of the hard palate and medial pterygoid plate without frank destruction. The lesion measured 4.4 cm × 2.4 cm × 3.1 cm (anterior-posterior by mediolateral by craniocaudal) and resulted in superior displacement of the right inferior turbinate. The lesion extended to the region of the torus tubarius resulting in opacification of the right middle ear cavity and mastoid air cells. MRI of the face lesion showed that the lesion was homogeneously T1-weighted hypointense (Fig. 2B) and showed heterogenous intense enhancement (Fig. 2C) following administration of intravenous contrast material. There was no diffusion restriction within the lesion. On the fluid-sensitive STIR sequence (Fig. 2D), there were multiple small nodular foci of hypointensity within the lesion with linear-curvilinear hyperintensities about them. The imaging characteristics were consistent with a non-aggressive, locally expansile lesion given lack of frank osseous destruction and presence of scalloping. The relative T2 hypointensity of much of the lesion suggests fibrous components within it. The preoperative differential diagnosis included fibrous tumors, such as fibrous dysplasia and ossifying fibroma.

An awake fine needle aspirate into the center of the lesion was easily taken with a 25-gauge needle. Initial FNA showed small fragments of cytologically bland spindle cells with scattered multinucleate osteoclastic giant cells without ossification or calcifications. These findings raised the possibility of a central giant cell granuloma. (Fig. 3; PAP stain; X400).

The CT images were segmented in Carestream PACS System and exported as a stereolithography (.STL) file. The tumor was removed and both 3D models were then printed using white resin on a Formlabs Form2 printer with a 0.05 mm layer resolution (Fig. 4A). As well the model was printed with the tumor colored green on an XYZ da vinci color 3D printer. The two models shown in Fig. 4B–C allow for better visualization of the tumor for both the physician and the patient/parents. The model with the tumor removed can also be used to plan any implants (i.e. obturator), pre-surgically.

During discussions with the family, the 3D printed model of the tumor was used as an extensive visual aide in describing the different treatment options as well as the resulting anatomical changes that would occur with each modality. The primary discussion focused on the risks and benefits of enucleation or tumor debulking versus that of a partial maxillectomy with negative resection margins and reconstruction. It was explained to the parents, with help from the 3D printed model, how enucleation would focus on removing the tumor with the margins limited to the capsule of the tumor itself, leaving as much regular anatomy intact with a defect involving only the palate without need for reconstruction. But this would come with a higher risk of recurrence given unclear negative margins as well as risk for development of an oronasal fistula and velopharyngeal insufficiency. This was contrasted to the more aggressive partial maxillectomy with negative margins that would result in a larger defect with need for likely local or free flap reconstruction with benefits of lower likelihood of recurrence but longer recovery period and risk for both cosmetic and functional deficits. The parents decided to proceed with tumor enucleation after weighing pros and cons of each modality with the surgical team.

The patient then underwent tumor enucleation of his palatal tumor with preservation of the nasal mucosa and palatal mucosa overlying the tumor. Frozen sections of submucosa taken intraoperatively were confirmed to be minor salivary gland tissue only. Once the mucosa was raised, the eggshell-thin bony hard palate abutting the tumor was encountered and the submucosa was raised away from the periosteum and bone. Once the nasal side was reached, the tumor was lifted off the bony periosteum of the maxilla and floor of nose and the tumor was removed. Once all gross tumor and periosteum was removed from the patient, Kenalog was injected into the remaining nasal mucosa and soft tissue. The palatal flaps were then re-aligned to close the defect. An obturator was placed to facilitate oral intake and velopharyngeal insufficiency in the postoperative period. The patient did develop a small oronasal fistula post-operatively but had good functional status with oral intake and speech with the obturator in place and the defect granulating in without signs of recurrence.

Surgical resection of the mass showed a cellular cytologically bland spindle cell proliferation with a somewhat whorled arrangement of cells, multiple foci of ossification, areas of interconnecting osteoid (Fig. 5A; H&E stain; X100) and foci of osteoclastic giant cells lining irregular cleft-like spaces (Fig. 5B; H&E stain; X100). The findings are that of a juvenile ossifying fibroma.

3. Discussion

The use of shared decision making has played a vital role in the development of our current healthcare practices, emphasizing the joint decision making and relationship between physicians and patients as the cornerstone to patient-centered care [11]. It has been shown that patients want to have more information in regards to their own healthcare and have more control and play a larger role in making decisions related to their illnesses and treatments [12]. In the surgical field, the difficulty lies in how to present enough information in such a way that patients and their families can understand the implications of their disease and the surgical management, particularly when it comes to visualizing how the body will change secondary to the procedure. The use of 3D printing has created an avenue in which providers can visually show patients how the disease affects the body and the ultimate changes that will occur secondary to different treatment options. One

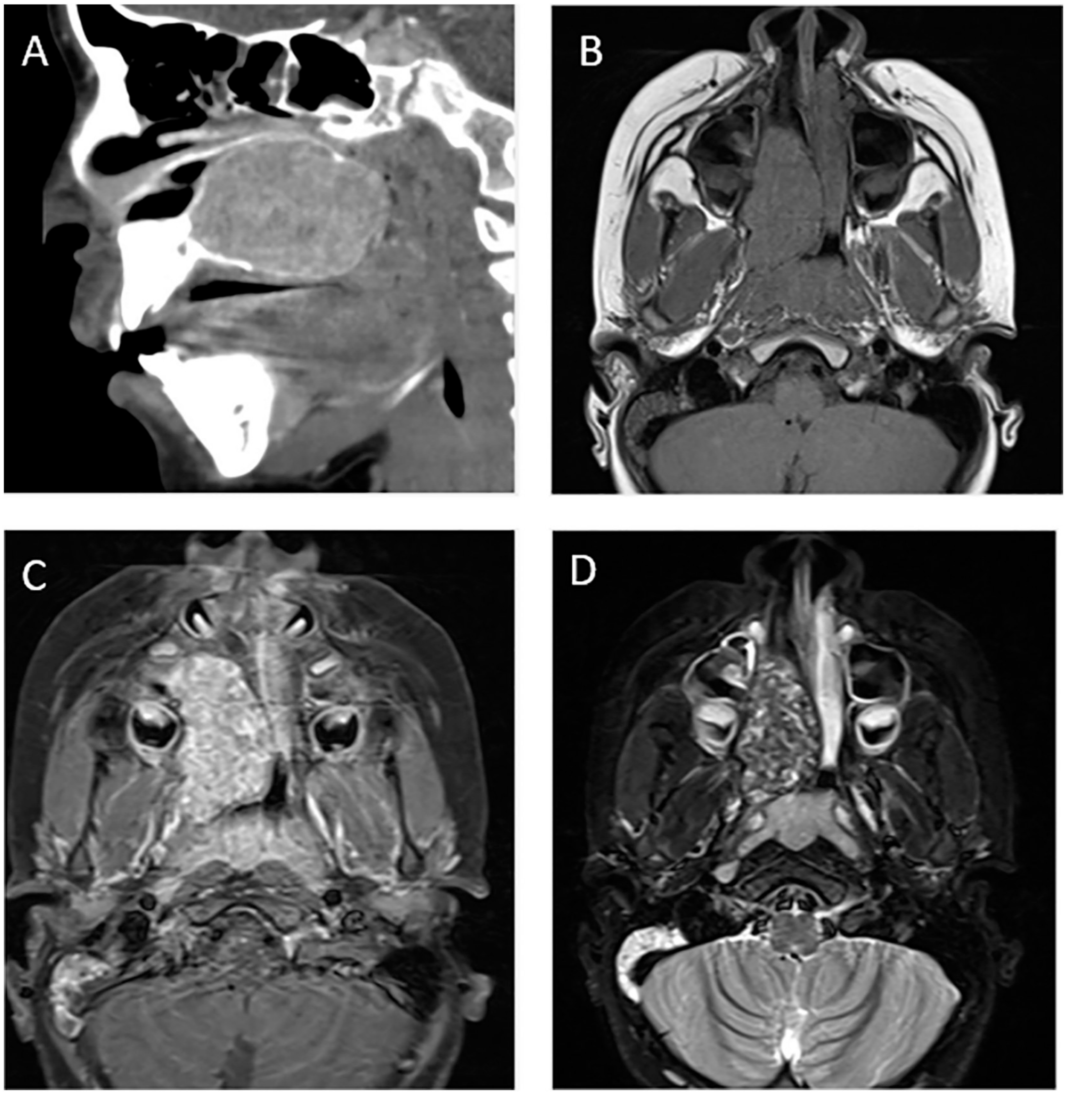

Fig. 2. A–D: Representative imaging of the palatal mass. A) Sagittal CT scan, B) T1-weighted MRI, C) Axial T1-weighted MRI with contrast, D) Axial fluid-sensitive STIR sequence MRI.

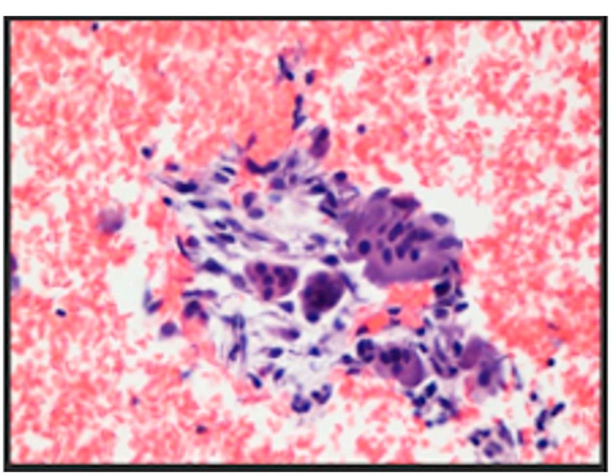

Fig. 3. Fine-needle aspirate of the palatal lesion.

study using 3D printing showed that a patient undergoing cranioplasty revision for osteogenesis imperfecta felt that the 3D printed model preoperatively helped her to better understand the proposed surgical plan and increased her comfort level prior to undergoing the surgery [13]. The same was noted when 3D printed models were created for patients with gliomas and kidney tumors that helped give the patients better understanding of their disease [10, 14]. By increasing patient knowledge with the use of visual tools such as high-resolution 3D printed models, we can potentially strengthen the communication and understanding to help improve shared decision making between the healthcare provider and patient.

In our case, this was particularly impactful in that the shared decision-making process involved a pediatric patient that could have drastically different post-operative outcomes depending on the decided treatment modality. Knowing that JAOF is a benign but aggressive disease, the 3D model allowed us to explain to the parents the current impact of the disease on the craniofacial structure of the maxilla and describe the potential resulting defects that would result secondary to different surgeries. Without the visual aide, understanding the difference in functional and cosmetic outcome between enucleation and partial maxillectomy may be difficult for patients to comprehend without the underlying medical or anatomical knowledge. The use of 3D printing in this case bridges that gap so that the family could see how each surgery was different while weighing the pros and cons related to both functional and cosmetic outcomes against chance for disease recurrence. The family felt that the use of the visual aide helped

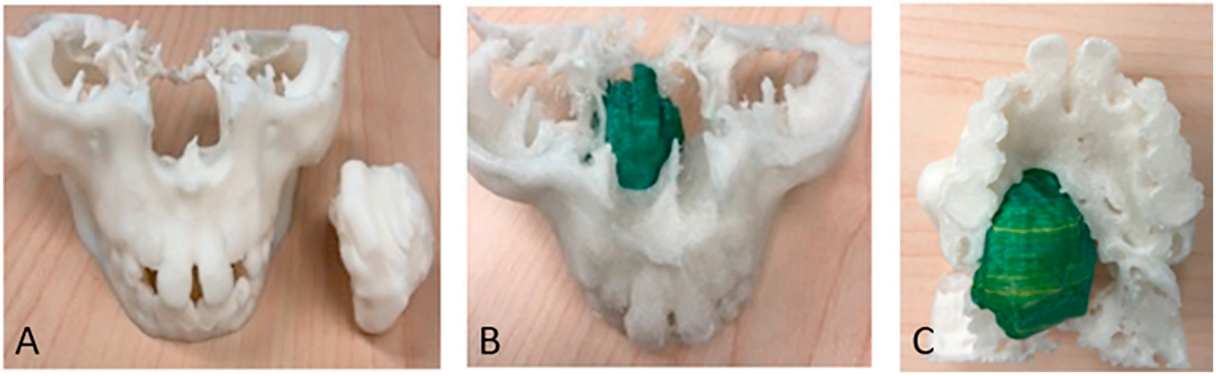

Fig. 4. A–C: 3D printed model of the lesion. A) General model with mass removed, B) Green marks anatomical positioning of the mass, C) Intraoral view of the green marked palatal mass. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

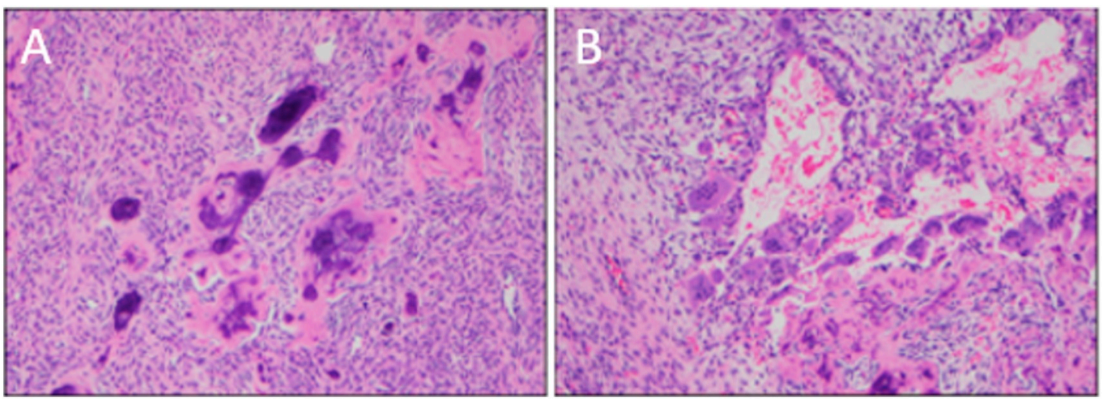

Fig. 5. A and B: H&E stained pathological specimens.

to better their understanding of the patient's disease and helped them come to a joint informed decision in which they were comfortable.

The applications of 3D printing in healthcare continue to evolve. As higher resolution imaging continues to progress, the realism and fine details afforded by 3D printed models will continue to improve. Though this is one case report detailing the use of a 3D printed model in helping in educating and shared decision making for a patient with JAOF, further research is needed to clarify the utility of the use of 3D printed models in patient education and shared decision making prior to decisions on surgical treatment. As 3D printing technology becomes more readily available to medical centers and as healthcare continues to evolve into a more patient-centered, shared decision-making model, the use of 3D printed models will provide an invaluable tool.

Here we report, what is to our knowledge, the first description of the successful management a juvenile ossifying fibroma with the use of 3D printing guidance for shared decision making. As we continue to use 3D printed models as aides in educating patients and their families prior to surgery, we will help to further strengthen the physician-patient relationship and patient-centered healthcare model.

Financial disclosure

The authors have no financial relationships relevant to this article to disclose.

IRB

This study was granted IRB exemption by the Northwell Health Human Research Protection Program.

Declaration of Competing Interest

The authors have no conflicts of interest relevant to this article to disclose.

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