



A Case of Sphenoid Wing Dysplasia

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INTRODUCTION

Neurofibromatosis type 1 (NF1) is an autosomal dominant disorder that with an incidence of one in 2500.¹ Common manifestations include Lisch nodules, peripheral neurofibromas, optic gliomas, prominent corneal nerves, and café-au-lait spots.² Another known complication is sphenoid wing dysplasia, affecting 10-20% of NF1 patients.³

CASE PRESENTATION

- 4-year-old female with a history of NF1 found to have pulsatile proptosis and hypoglobus.
- MRI revealed left cavernous sinus enhancing mass (suspected arachnoid cyst), a left upper eyelid mass, and enlarged orbital fissure.
- Multidisciplinary surgical team including ENT, Neurosurgery, and Oculoplastics planned for combined surgery using 3D printed models. Image 1
- Neurosurgery performed a left frontotemporal orbital craniotomy and durotomy/durectomy resection of the arachnoid cyst.
- ENT performed orbital reconstruction and repositioning with medialization of left orbit via a temporalis flap.
- Oculoplastics performed orbitotomy with resection of an enlarging plexiform neurofibroma. Image 2
- Following surgery, the patient has been started on Selumetinib to prevent regrowth of the resected neurofibroma.

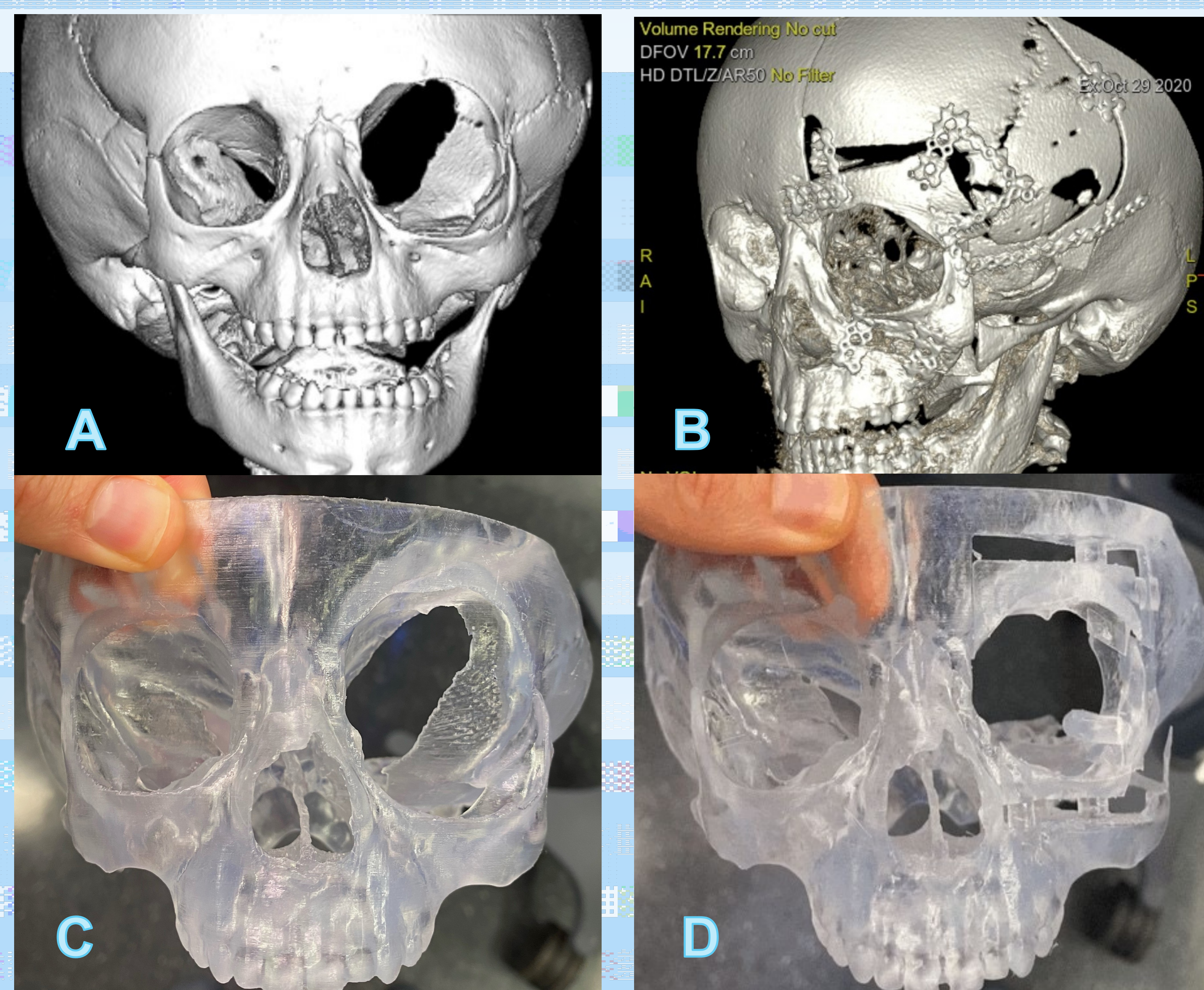


Image 1: A) CT 3D reconstruction demonstrating an enlarged superior orbital fissure, enlarged orbit and enlarged left middle cranial fossa with thinning of the corresponding calvarium and orbital walls. B) CT 3D reconstruction following surgical orbital repositioning. C) 3D printed model based on (A). D) 3D Printed model of the planned post-operative changes.

DISCUSSION

- A review of the literature reveals a review article regarding the usefulness of 3D printed models in maxillofacial surgery. The article reviews 2889 cases in which 3D printing was utilized. The primary benefits were precision in surgery and reduction of surgical time. The primary disadvantages were cost and manufacturing time.⁴
- Differential diagnosis of pulsatile proptosis includes sphenoid wing dysplasia (in NF1), carotidocavernous fistula, arteriovenous malformation, trauma (orbital roof fracture), and arachnoid cyst.⁵⁻⁸
- Selumetinib is a MEK1/2 inhibitor shown to prevent regrowth of plexiform neurofibromas and to reduce the size of inoperable tumors in 70% of cases.⁹



Image 2: Resected orbital plexiform neurofibroma.

CONCLUSION

Our patient had both sphenoid dysplasia and an arachnoid cyst. This complicated case required a multidisciplinary approach for the safest functional and cosmetic result. 3D printing of anatomical models based on the patient may allow for improved surgical precision and reduction of surgical time.

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