



Case Report

Transport of patients with giant disfiguring cranial tumors from Africa to the US for collaborative multidisciplinary treatment

Eric Nussbaum¹, Patrick Graupman², Martin Lacey³, Andrew Harrison⁴, Jodi Lowary¹, Jillienne Touchette⁵, Leslie Nussbaum¹

¹Department of Neurosurgery, National Brain Aneurysm and Tumor Center, Minneapolis, Minnesota, United States, Departments of ²Neurosurgery, ³Craniofacial and Plastic Surgery, Gillette Children's Specialty Healthcare, Saint Paul, Minnesota, United States, ⁴Department of Ophthalmology and Visual Neurosciences, University of Minnesota, United States, ⁵Superior Medical Experts, Minnesota, United States.

E-mail: *Eric Nussbaum - lnussbaum@comcast.net; Patrick Graupman - graupman@mac.com; Martin Lacey - martinslacey@gmail.com; Andrew Harrison - eyeliddoc@gmail.com; Jodi Lowary - jodi.lowary@allina.com; Jillienne Touchette - Touchettejc@gmail.com; Leslie Nussbaum - nussbaumla@gmail.com



*Corresponding author:

Eric Nussbaum,
Department of Neurosurgery,
National Brain Aneurysm and
Tumor Center, Minneapolis,
Minnesota, United States.

lnussbaum@comcast.net

Received : 18 December 2020

Accepted : 05 March 2021

Published : 19 April 2021

DOI

10.25259/SNI_923_2020

Quick Response Code:



ABSTRACT

Background: Giant disfiguring cranial tumors are exceptionally rare and develop over the course of many years, typically in patients who lack access to medical care. Here, we describe four patients who were flown to our center for treatment by a multidisciplinary surgical team, who had previously been turned down for treatment at multiple international centers in Africa, Europe, and the United States (US) due to complexity and financial concerns. The case series describes socioeconomic implications and the feasibility of offering such care to patients from outside the US.

Case Descriptions: Four patients with giant skull disfiguring tumors were flown internationally and treated by a surgical team consisting of a complex cranial neurosurgeon, a craniofacial reconstructive plastic surgeon, and an oculoplastic surgeon. All patients underwent aggressive surgical therapy with the aim of complete tumor removal and simultaneous cranial reconstruction. A patient with osteogenic sarcoma underwent two additional resections in 3 years, with delayed reconstruction. They returned home but ultimately succumbed to the disease. A patient with ossifying fibroma required two follow-up procedures for cosmetic reconstruction and sought asylum in the US, where they remain today. Two additional patients, one with a giant plexiform neurofibroma and one with a cerebellopontine angle meningioma, achieved good results and returned to Africa 1 month and 3 weeks after surgery, respectively.

Conclusion: Resection of giant disfiguring cranial tumors and reconstruction of the impacted region requires an experienced multidisciplinary team. These cases can be managed by transporting such patients from areas without access to medical care to specialized centers able to provide excellent care.

Keywords: Cranial reconstruction, Disfiguring cranial tumors, Tumor resection

INTRODUCTION

Giant disfiguring cranial tumors have become extremely rare due to advances in early tumor detection and generally only occur when patients do not have access to care during the early stages of tumor development.^[2,11,12] Specialized care is required in these cases due to the high risk of perioperative hemorrhage,^[2,7,11,12] difficulty of removing the large mass,^[7,8] and the potential need for reconstruction of the damaged skull bone and facial structures.^[2,7,11] Here,

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we describe four patients with giant (>20 cm) disfiguring cranial tumors who were turned down for surgery at multiple international locations due to medical complexity and financial concerns. They were flown to be treated by a multidisciplinary surgical team including neurosurgeons, craniofacial reconstructive surgeons, and oculoplastic specialists. A cross-continental arrangement allowed the hospital and surgeons to complete their services free of charge, waiving the treatment costs, which ranged from \$125,000 to \$250,000.

CASE REPORTS

Case 1

A 28-year-old male from Ethiopia presented with a large bilateral mass measuring over 20 cm in its largest dimension arising from the frontal aspect of the left side of his head. The mass had grown rapidly over the previous 2 years, resulting in headaches, nosebleeds, and left-sided blindness.

Imaging studies

A catheter angiogram showed portions of the mass that were mildly to moderately vascular. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a 24 cm × 20 cm × 18 cm extra-axial mass centered in the region of the left frontal bone [Figure 1]. The mass was partially solid, partially calcified, and partially cystic.

Operation

A bicoronal incision was made to the level of the zygoma and was carried down to the level of the tumor. The scalp flap was reflected forwards and backwards until we reached normal bone posteriorly and nasal brow anteriorly. Cutaneous and subcutaneous bleeding was controlled with bipolar electrocautery. Two holes were drilled through the skull on either side to normal dura and were connected posteriorly and anteriorly, crossing the sagittal sinus. The skull was cracked and lifted, along with the massive tumor mass, in one piece, weighing 17 lbs. Additional tumor was cleared out of the orbit and from the region of the sphenoid wing and temporal fossa. Substantial bleeding from the bone and dura was controlled, with 4 L of blood lost in total. The scalp and skull were closed over two large Jackson-Pratt drains and a portion of the redundant scalp flap was resected. Intraoperative photographs are presented in [Figure 2].

Pathological examination

Histopathological examination of the mass revealed a high-grade osteogenic sarcoma with telangiectatic features.

Postoperative course

The patient had no neurological defects or serious side effects and returned to Ethiopia where he underwent limited

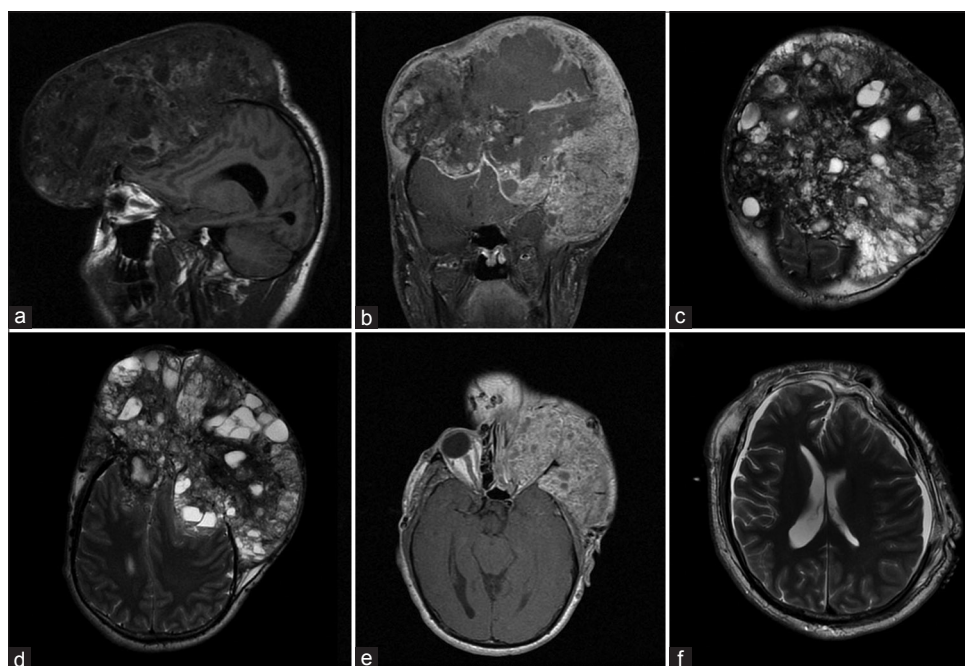


Figure 1: Preoperative radiographic images of disfiguring osteosarcoma tumor mass (Case 1). (a) Sagittal view, T1-weighted MRI showing disfiguring tumor obstructing the left eye. (b) Posteroanterior view, CT. (c and d) Axial view, T2-weighted MRI showing well-defined enhancing nodules within the mass. (e) Axial view, CT. (f) Axial view, T1-weighted MRI showing displacement of brain parenchyma.

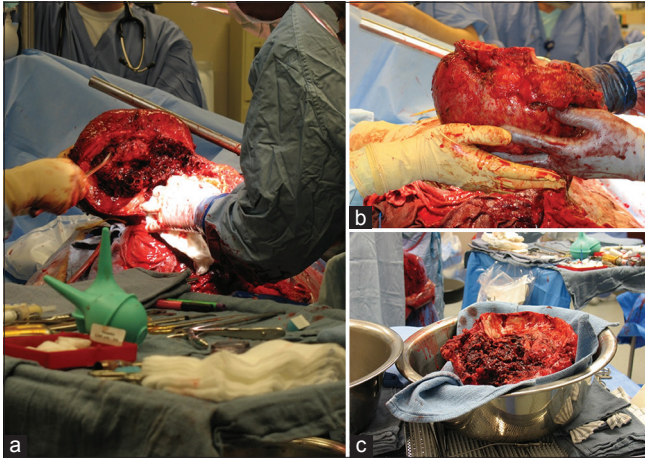


Figure 2: Intraoperative photographs showing (a) resection of the mass, (b) removal of the mass, and (c) the mass fully separated from the patient.

treatment with chemotherapy, but not radiation. At that time, a proton emission tomography (PET) scan had shown the mass to be locally contained without remote spread.

Second surgery

The patient returned 11 months later with a recurrent tumor involving the nasal cavity and orbits. The largest axial dimensions of the anterior component measured 7 × 12 cm. The patient opted to undergo a second operation. The bicoronal incision from the previous surgery was reopened, and the scalp was reflected anteriorly revealing the tumor over the right orbital region. The tumor was resected bluntly with bipolar cautery. Once the tumor was resected, dural repair was placed. The team's craniofacial reconstructive surgeon performed a mesh cranioplasty. The wound was closed with a Jackson-Pratt drain and nylon sutures, and the patient was taken to the recovery room in stable condition.

Third surgery

The patient returned 26 months later with another large recurrence of the tumor and opted for a third surgery. Before surgery, the patient underwent endovascular particle embolization of the right middle meningeal artery, right accessory meningeal artery, and anterior deep temporal artery, which were supplying the tumor. During surgery, the previous bicoronal incision was reopened. The scalp was scarred down to the underlying mesh. Tumor underlying the mesh and other posteriorly located tumor tissue were aggressively removed. The frontal bone, which had been largely replaced by tumor, was removed bilaterally down to the orbital rims. The orbital rims were removed, and orbits were decompressed aggressively to the optic and carotid canals. The sphenoid wing was drilled down bilaterally, as

it had been completely involved with the tumor on the right side. Surgeons continued down into the pterygopalatine fossa and also came down in the midline into the mid-face to the region of the ethmoid sinuses, which were completely filled with tumor tissue.

The patient experienced severe blood loss and intraoperative coagulopathy. This was addressed with activated Factor VII. Some tumor remained in the ethmoid sinus and behind the incision on the left side; this was not removed due to the significant blood loss that had already occurred. The craniofacial reconstructive surgeon using the existing titanium graft from the previous cranioplasty to reconstruct the defect.

The patient was treated with additional rounds of chemotherapy, but after returning to Ethiopia, he succumbed to his disease 9 months later.

Case 2

A 20-year-old female from Ethiopia presented with a giant skull-based tumor with a massive orbitocranial deformity distorting her facial anatomy and right eye.

Imaging studies

Diagnostic angiography, CT, and MRI revealed a giant, moderately hypervascular, multilobulated, and peripherally calcified expansile mass lesion which appeared to arise from the skull base and occupy the entire nasal cavity, maxillary sinus, and ethmoid sinus regions [Figure 3]. The intracranial extension of the tumor obliterated the sella turcica, bilaterally invaded the cavernous sinuses, and caused extensive elevation and splaying of the internal carotid arteries, anterior cerebral arteries, optic chiasm, and intracranial optic nerves. The mass measured approximately 10 cm × 8 cm × 11 cm. There was no evidence of edema or gliotic reaction in the adjacent brain parenchyma.

Operation

The patient underwent resection of the tumor through a subcranial transfacial approach. A bicoronal incision was carried down, preserving the galeal layer for possible use as a graft later. An incision was made over the face and nose to expose the front of the tumor. We broke through the thin bony shell and entered the multiloculated cystic tumor cavity. Fluid was drained and the operating microscope was brought into the field. This allowed us to work circumferentially to remove the entire solid tumor components all the way back to the circumferential bony shell. This left a massive cavity measuring over 15 cm in diameter. All soft tissues were removed and bone was drilled down. The lateral wall of the tumor was drilled, allowing for replacement of the globe to a more natural position. Bone was also drilled off the medial

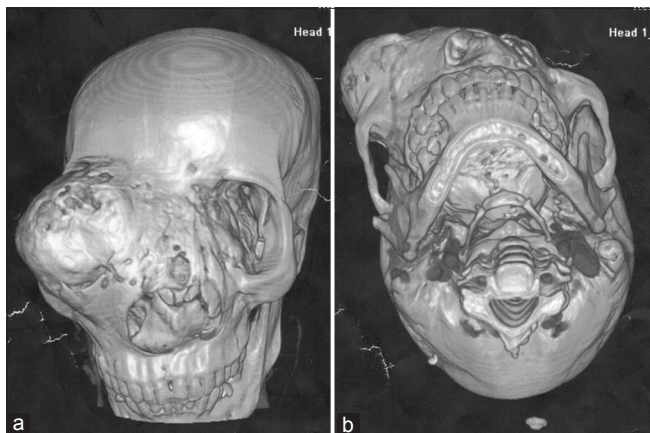


Figure 3: Preoperative radiographic images of disfiguring ossifying fibroma (Case 2). (a) Anteroposterior view, CT showing disfiguring tumor distorting the right eye. (b) Axial view, CT.

wall to decompress the lesion. The dura was never entered. Calvarial bone grafts were harvested by taking a large frontal parasagittal craniotomy.

A craniofacial reconstructive surgeon with the help of an oculoplastic specialist completed a complex reconstruction. We used the bone grafts to reconstruct a new orbit and mid-face. Repair of the skull bone and scalp was undertaken in a second surgery by our craniofacial/plastic surgeon.

Pathological examination

Histopathological examination demonstrated an ossifying fibroma with a proliferation of plump spindle cells, mostly hypocellular, with focal hypercellular areas. Production of woven bone was seen intermittently with the spindle cells. The spindle cells showed focal areas of storiform arrangement with focal osteoblastic rimming. The final diagnosis was fibrous dysplasia.

Postoperative course

The patient remained neurologically and physically stable throughout the follow-up period, with the exception of increasing headaches and occasional nosebleeds. Follow-up PET and MRI at 1 month and 10 months postoperative were stable and showed no evidence of tumor growth. The patient has been followed for 10 years with mild interval progression of bony changes but no clear evidence of tumor regrowth. During this time, she underwent one additional delayed cosmetic procedure for her left eye.

Case 3

A 39-year-old man presented with extensive bone and soft-tissue changes related to a large plexiform neurofibroma involving the left temporal region, underlying bone, and

dura. He had no neurological deficit but did have worsening headaches. The patient traveled from Ethiopia for surgical repair.

Imaging studies

CT scan of the brain including bone windows and thin cut imaging through the temporal bones demonstrated extensive bony erosion with scalloping of the bone edges compatible with the long-standing presence of the left temporal mass which itself involved the subcutaneous tissue, temporalis muscle, and dura. There was no appreciable intradural extension of the lesion. These findings were confirmed on MRI, which revealed the heterogeneously enhancing mass involving the soft tissue, bone, and dura of the left temporal region. Diagnostic angiography revealed enlargement of the superficial temporal and middle meningeal arteries, which were embolized with polyvinyl alcohol the day before surgery.

Operation

A single-stage operation was carried out. Our craniofacial team resected a large amount of redundant soft tissue and skin overlying the relatively hypervascular mass. The tumor was removed *en bloc*, and thickened underlying dura was also resected. The underlying bone was severely thinned, and titanium cranioplasty was used for reconstruction.

Pathological examination

Pathological examination was compatible with a plexiform neurofibroma.

Postoperative course

Postoperatively, the patient did well without new deficits. After 1 month's recovery and removal of all sutures, the patient was able to return home. Six-month follow-up was carried out remotely, and the patient was doing well without regrowth of tumor or new deficits. The patient was offered a second surgery to correct cosmetic appearance, but was happy with the result and declined to return.

Case 4

A 44-year-old woman from Nigeria presented with worsening headaches and balance difficulties.

Imaging studies

The patient traveled to our center where MRI revealed a 6 cm mass lesion involving the cerebellopontine angle with overlying bony hyperostosis. CT scan with bone windows demonstrated the marked hyperostosis resulting in deformity

of the overlying skull, which was well hidden by the patient's hair on physical examination. There was marked deformity of the cerebellum and brainstem by the extra-axial mass, which displaced the basilar artery and extended out along the internal auditory canal.

Operation

The patient underwent suboccipital craniotomy with resection of the tumor. At the time of surgery, the majority of the tumor was removed, but a small amount was left due to adherence to the basilar artery. The bony hyperostosis was aggressively drilled away, and titanium mesh was utilized to cover the defect.

Pathological examination

Pathological examination demonstrated a meningioma.

Postoperative course

The patient tolerated the procedure well, though she developed partial fourth cranial nerve palsy after the surgery. Otherwise, she suffered no new deficits. She returned home 6 weeks after surgery, and follow-up imaging at 18 months was stable. Her fourth cranial nerve palsy had fully resolved.

DISCUSSION

Here, we demonstrate the successful treatment of giant skull deforming tumors in 4 patients from Africa by a multidisciplinary team. Giant skull deforming tumors are exceptionally rare with limited cases reported in recent history.^[2-8,11,12,15] Since these tumors develop over the course of several years,^[2,7,8,11,12] they usually occur in patients who lack access to medical care or otherwise resist treatment during the early stages of tumor development.^[2,12] The majority of recent cases have been meningiomas,^[11,12,15] but cases of glioblastoma,^[5] squamous cell carcinoma,^[6] and osteosarcoma or ossifying fibroma, as seen here, have also been reported.

Common risks and symptoms associated with giant skull deforming tumors include headaches,^[6,15] cognitive and neurological impairment,^[7] and ulceration and subsequent infection of the scalp.^[2,11] However, some patients experience little to no symptoms beyond the skull protrusion itself,^[8,11,12,15] which may contribute to the delay in seeking treatment. Surgical complications include damage to existing brain structures and hemorrhage.^[2,8,11,12] As we experienced, the resection of highly vascular tumors can lead to substantial blood loss and must be anticipated and monitored during the procedure. In addition, reconstruction of the skull or facial structures adds a major challenge, necessitating consultation and careful planning with plastic surgeons.^[2,8]

Reported methods of reconstruction include the use of myocutaneous skin flap reposition,^[2] bone reconstruction with methyl methacrylate,^[11,15] or dura mater reconstruction with fascia lata,^[11] lyophilized dura,^[15] or titanium mesh as described here. The use of myocutaneous skin flap reposition represents an important technique to provide tissue coverage following resection of complex tumors. Importantly, when employing this approach, the flap should consist of sufficient subcutaneous tissue and vascularity to ensure it receives ample blood supply.^[2] In our experience, these giant tumors often act as "tissue expanders," leaving more than enough skin to provide tissue coverage and often necessitating resection of redundant tissue at the conclusion of the procedure.

Due to the complexity and rarity of giant skull deforming tumors, each procedure must be planned and performed on a case-by-case basis. Imaging studies should be undertaken to help establish the extent of the tumor and guide surgical resection. As we do in all cases, the goals of surgery balanced maximal safe surgical resection with a desire to avoid permanent neurological deficit or disability. Although we knew that the only chance to achieve a cure in these cases might be the initial surgery (given the barriers to having the patients return to the US for repeat operations), we also felt that a permanent disability would be an unacceptable complication, particularly for individuals returning to countries with limited medical and social support resources. Three of our patients had been "home bound" for years before surgery due to the disfiguring nature of their tumors. In these cases, it was hoped that simply restoring a "near-normal" cosmetic result would allow the patients to re-enter society, radically improving their quality of life.

Giant skull deforming tumors are exceptionally rare and there is no definitive consensus on treatment practices, particularly for patients from countries with such limited medical resources. Historically, neurosurgeons have traveled to areas in sub-Saharan Africa, Latin America, and the Caribbean to train neurosurgeons locally, where an estimated 22 million additional neurosurgical procedures are needed for patients.^[10,13,14] However, for challenging tumors like these that require a multidisciplinary team of specialists, care is facilitated by bringing patients to an area where such teams are already concentrated. The highly specialized nature of modern neurosurgery requires the collaboration of many practitioners to address the multiple aspects of these unique cases. These multidisciplinary teams must include physicians from different subspecialties, including cranial surgery, tumor resection, craniofacial reconstruction, and oculoplastic surgery. Complete surgical resection and skull reconstruction are feasible in these patients, but chances of hemorrhage and difficulties of skull reconstruction require a skilled, multidisciplinary team of clinicians.

Interestingly and unexpectedly, our provision of care to these patients received both positive and negative responses by the

lay community in the Twin Cities. Treatment costs for the patients described here ranged from \$125,000 to \$275,000. The cross-continental agreement allowed the surgeons and hospital to offer their services free of charge to the patient. When word of the surgeries was made public by the local media,^[1,9] much of the Twin Cities community responded positively, while others found it unfair to provide free care to noncitizens when there are US citizens who cannot afford treatment. Thus, the hospital has become averse to future ambitious endeavors because of risk of negative media attention.

CONCLUSION

Giant disfiguring cranial tumors are extremely rare, primarily occurring in patients who are unable or unwilling to access care during the early stages of tumor development. Resection of such large masses can lead to substantial blood loss, and reconstruction of the impacted skull region and facial features requires precise management. Here, we show the successful treatment of four giant disfiguring cranial tumors by a multidisciplinary team. With proper caution and resources to send patients to a center able to provide excellent care, simultaneous surgical resection and craniofacial reconstruction by a multidisciplinary surgical team are a viable treatment option for these rare cases.

Acknowledgments

The authors acknowledge Superior Medical Experts for research and drafting assistance.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

Financial support and sponsorship

This manuscript was supported by a grant from the United Hospital Foundation.

Conflicts of interest

Jillienne Touchette is CEO and has ownership interest in Superior Medical Experts. All other authors declare no conflicts of interest.

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How to cite this article: Nussbaum E, Graupman P, Lacey M, Harrison A, Lowary J, Touchette J, *et al.* Transport of patients with giant disfiguring cranial tumors from Africa to the US for collaborative multidisciplinary treatment. *Surg Neurol Int* 2021;12:175.