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Modifying the facial abnormalities in a patient with neurofibromatosis using 3D modelling and virtual surgery: A case report

Seyed Hossein Mortazavi[1], Farzin Sarcarat[1], Roozbeh Pahlevan[2*], Fatemeh Nasrollahi[2], Soroush Valinia[3], Farnaz Keyhanlou[4]

1. Craniofacial research center of Islamic Azad university, Dental branch, Tehran, Iran.

2. Oral and maxillofacial resident of Bouali hospital, Islamic Azad university, Dental branch, Tehran, Iran.

3. Mechanical engineering B. sc.,Craniofacial research center of Islamic Azad university, Dental branch, Tehran, Iran.

4. Dentistry student, Islamic Azad university, Dental branch, Tehran, Iran.

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**Corresponding author:*
Roozbeh Pahlavan
Oral and maxillofacial resident of Bouali hospital, Islamic Azad university, Dental branch, Tehran, Iran.

Tel: +98-21-22580933
Fax: +98-21-22580933
Email: Dr_r_pahlevan@yahoo.com

ABSTRACT

Neurofibromatosis is associated with various degrees of soft tissue and hard tissue involvement; among which tumor-related face ptosis is the main reason for serial exitional treatment. This case report demonstrates usage of analyzing mirror images technique for exact examination of abnormalities affecting one side of the face. The patient was a 48-year-old woman, diagnosed with Von Recklinghausen's disease with numerous surgeries in the past, who was seeking surgical treatment for the mass on the left side of her face and modifying deformity.

Materials and Methods: The procedure began with producing a 3D scan of patient's face by a high quality laser scanner and the scanner data were transferred to relevant softwares and then the postsurgical changes were applied to the patient' face. Thereafter, using engineering and graphical softwares such as CATIA and SolidWorks, the necessary surgical molds were designed. In the next steps, the molds were built using a 3D scanner several sizes. During surgery, the templates were first placed on the patient's face in each phase and debulking was achieved based on the relevant template so that favorable amount of debulking was finally reached using these surgical guides.

Conclusion: The simultaneous application of 3D surface laser scanner technology and reconstruction of the affected side based on the healthy side and analyzing the information obtained from this reconstruction, make an objective assessment over the precise prediction of the abnormal soft tissue volume, pre- intra- and post-operatively which leads to improved results.

Key words: Neurofibromatosis; 3D modeling; virtual surgery.

Introduction

Neurofibromatosis (NF) is a rare autosomal dominant disorder occurring 1 in 3000- 4000 births [1,2]. This disorder occurs in two types: NF type I, which is also known as Von Recklinghausen skin disease, and NF type II. The type I disease is characterized by the following symptoms: several neurofibromas (seen in 85-90% of cases) [3], cranio-orbital-temporal involvement in 1-4% of cases [4,5] with various degrees of soft tissue and hard tissue involvement accompanied with malformation and asymmetry in the face and ear areas, facial skin and upper eyelid ptosis with visual defects depending on the amount of tumor development and its spread to surrounding regions. In acute cases in which the involvement of eyelids and internal areas of eyes occurs, one can also expect visual defects and even blindness [6]. Neurofibroma mostly affects the soft tissue, however, it may include intrabony defects. The main reason for serial excision treatments in NF is the tumor-related face ptosis due to

These treatments are used to reduce the tumor volume. Due to the spread of tumor to surrounding structures and sometimes the intensity of involvement, even the surgery cannot remove considerable damages in most cases and the tumor remains usually tend to regrow and eventually lead to re-sagging of the region. This occurs especially in operations, in which only suturing is used to hold tissues in place against gravity. These patients often experience the reoccurrence of the problem within a short time after surgery and therefore have to undergo several surgeries for relative modification [7]. Surgical treatment, in the absence of hampering medical complications, is considered as the first choice in treating the patients with such clinical problems. Plexiform neurofibroma is usually deep and aggressive and it is impossible to be totally excised without damaging nontumorous tissues. Therefore, surgical treatment is only used to reduce the tumor volume with the aim of reaching an acceptable degree of beauty [8,9]. The 3D virtual surgery using CAD/CAM (computer aided design and manufacturing) technique has recently become popular in cosmetic surgery due to its ability to create a geometric model in 3D space. This facilitates determining planes for surgery, makes the results of the surgery, to some extent, predictable before operation, and helps to reduce the operation time [10]. Currently, the technology of 3D images and printing is effectively used in the treatment of soft tissue and bone defects. The technique of analyzing mirror images is used for exact examination of abnormalities affecting one side of the face. Analysis of mirror images renders an exact duplicate of an image in reverse form. In this analysis, the mid-sagittal plane is used to create symmetry. The amount of discrepancy in this method is investigated through superimposing the healthy anatomic part on the affected side [11]. The following steps are performed to build 3D templates which are used to control the symmetry during surgery: Initially, the patient's face is scanned using a 3D scanner with a resolution of 16 Megapixel and then a 3D model is produced. Designing a symmetrical form of the face is conducted using engineering softwares in the next step, i. e., the ideal form of the patient's face with complete symmetry is designed based on the form of the healthy side using special software. The next step is designing a 3D mask from the symmetrical form of the face. The 3D facial mask includes a vertical axis of symmetry that coincides with the (mid-sagittal) axis of symmetry of the face and six horizontal axes, symmetrical with respect to the face axis of symmetry, which are designed based on the most raised points of the face. These six axes are superposed during the surgery on the axis of symmetry to examine the symmetry of the relevant face part in each phase of surgery. After designing the phase, it is time to produce the 3D mask which is used to control the face symmetry. This 3D mask is made of PVC and is built using a 3D printer with a resolution of 0.3mm. Later, the 3D mask is sterilized by ethylene oxide and used in the surgery.

Case Report

The patient was a 48-year-old woman, diagnosed with von recklinghausen's disease with numerous surgeries in the past, who was seeking surgical treatment for the mass on the left side of her face. She stated that her disease was diagnosed after birth. She underwent the first surgery to reduce the mass volume at the age of eight but due to intensive pigmentation and hypertrichosis on the left side of the face, the skin grafting from the pectoral area accompanied by re-debulking operation was performed at the age of eleven. She underwent several operations after that for modifying the deformity. During her clinical examination at this center, she was diagnosed with craniofacial neurofibromatosis with intensive involvement of soft tissue on the left side of her face (cheek, orbit, eat, occipital and temporal fossae regions) accompanied by weakness of zygomatic and frontal branches of the seventh nerve due to previous surgeries and obvious symptoms like Lisch nodule and blurred vision. Numerous café au lait macules and freckles of various sizes were found in upper and lower extremities. It was reported in her anamnesis that her father also suffered this disease with less intensity. The performance of MRI and CT scan during paraclinical examinations revealed the tumor boundaries affecting the skull on the left side of the patient's face. The tumoral region was calcified and extended to small blood vessels and the soft tissue in the larynx area at the lower part. CT angiography revealed that these vessels supplying blood to the tumor originated from the left carotid artery.

Materials and Methods

Procedure began with producing a 3D scan of patient's face by a high quality laser scanner with a resolution of 16mp and a precision of 0.5mm. The head was laid at NPH position, a position that was easily reproducible, clinically. Lying in this position is very important for diagnosis and pre-surgical work-up. The scanner data were transferred to relevant softwares and then the postsurgical changes were step by step applied to the patient's face. Thereafter, using engineering and graphical softwares such as CATIA and SolidWorks, the necessary surgical molds were designed. The mid-sagittal plane of the face, as the reference plane, was created though three points: nasion, anterior nasal spine, and pogonion. In the next steps, the molds were built using a 3D "2035 Quantum" scanner with a precision of 0.3 mm in several sizes (considering the current size of the damage and the gradual volume decrease until the soft tissues on the left and right sides of the face become symmetrical). The molds included transverse, basal, axial, and final molds.

The surgery was performed under GA. First the local anesthetic (Lidocaine mixed with epinephrine 1:100000) and then Tumescene solution was injected into the left side of the face. The rhytidectomy cut was performed on the left side extending towards the previous scar lines in the submandibular area of the patient. Dissection was carried out in supra SMAS form and then the tumoral tissues of the area were removed individually. The templates were first placed on the patient's face in each phase and debulking was achieved based on the relevant template so that the favorable amount of debulking was finally reached using these surgical guides. The grade of symmetry was checked using the final splint followed by suturing. Later, the intraoral approach was performed to remove the tumoral tissue from the buccal mucous. A two-layer Medpore sheet prosthesis with a thickness of 1.5 mm was then inserted in the defected area of orbital floor. The genioplasty surgery was performed at the next phase to modify the chin position. A Medpore prosthesis of an average size was inserted in the right mandible angle.

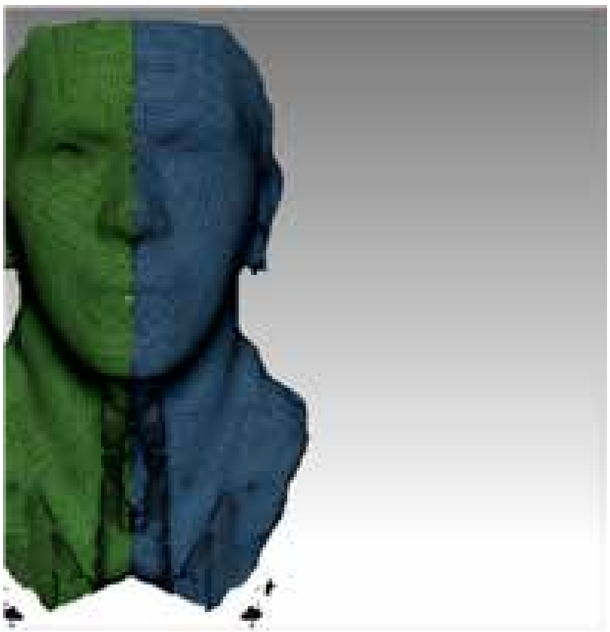
Discussion

Deformities and malformations due to NF type I are hardly treatable and most patients have to undergo several surgeries to reduce the tumor volume. Various studies on neurofibromatosis treatment protocols have shown that following partial removal of tumor, several operations will be necessary. This leads to more aggressive resections for more conclusive treatment of facial neurofibroma. Such aggressive treatments require more incisions, especially in the commissure and alar area of nose, to provide a better access to tumoral area. Finally, these sensitive areas have to be closed through precise techniques to prevent more aggravated deformities in these areas. In acute cases of disease with extensive involvements, a surgeon usually needs to remove the skin of the involved part for total resection of the tumor. Reconstruction is achieved using free flaps to modify these areas [12]. Some authors believe that the childhood years are the best time for treatment since the tumor hasn't yet been spread extensively at these ages. After adultness, there is higher possibility of tumor spreading and sagging increase due to gravity, and hence, the treatment will be more difficult. The reoccurrence rate of head and neck tumors is much greater than that found in other parts of body (60% compared to 29%). This is another reason for the difficulty of total tumor resection in these regions considering their anatomical structures. As a result, the possibility of reoccurrence after subtotal resection and debulking treatments (45%) is greater than that observed in total resection (with an occurrence rate of 20%) [13]. As the occurrence rate of tumor depends on tumor weight and the gravity of earth and while the tumor removal is difficult in cases of very aggressive tumors invading the surrounding tissues, the existence of a method to assess the amount of tumor resection for symmetrizing both sides of the face seems to be necessary. Numerous surgical treatments routinely used include several debulkings, lifts of tissues with sagging due to gravity, inserting prosthesis in healthy side, and other symmetrizing techniques.

Hivlin et al. (2010) applied the technique of marking both face sides using facial aesthetic unit to create a relative symmetry during operation and the level of patients' satisfaction (33 patients entered into this study) was reported to be good based on the global aesthetic score [14]. Chai et al. (2015) applied the scanning and 3D printing technique using computer softwares to assess the atrophy volume and the volume of adipose tissue needed for the reconstruction in seven patients suffering from facial atrophy and obtained considerably good results. At present, the patients' face images and 3D printing are very valuable tools in planning the treatments for face reconstruction. They are helpful in both defining planes for surgery and better symmetrizing the affected side with the healthy side of the face [10]. Harvold reported in his study that the line passing the anterior nasal spine and nasion coincides with patient's mid-sagittal plane in 90% of cases. He used the nasion, anterior nasal spine, and pogonion to create the mid-sagittal plane in his study [16]. The camera of the 3D laser scan can record a real image of the face similar to the 3D CT scan of facial surface tissue. The application of this camera reduces the amount of rays the patients are exposed to and spares the costs of CT scan. The volume difference between the affected and the healthy sides is measured by superimposition. To date, the MIA method has been used extensively for building craniofacial prostheses and implants. In our study, the volume of tumoral tissue was estimated before surgery and the operation was performed more precisely using surgical stents and through creating symmetry with the healthy side [16]. Such estimations have many advantages including the precise calculation of the amount of tissue to be resected, previewing the results of surgery during the operation, reduction of surgery time, and better assessment. This method, however, has limitations including the necessity for having a healthy side to create symmetry and also lack of precision in case of excessive facial edema during the surgical procedure [17].

Conclusion

The simultaneous application of 3D surface laser scanner technology and reconstruction of the affected side based on the healthy side and analyzing the information obtained from this reconstruction, make an objective assessment over the precise prediction of the abnormal soft tissue volume, pre- intra- and post operatively. Furthermore, the use of specialized computer software and 3D printers provides the possibility of performing a multiphase virtual surgery through designing and building surgical guides and molds based on healthy side before the real surgery. The application of these surgical guides and molds during surgery increases the precision and improves the final result of symmetrizing the affected side with the healthy side, makes the step-by-step assessment possible during the surgery, prevents irreparable errors of excessive tissue resection, and reduces the surgery time.



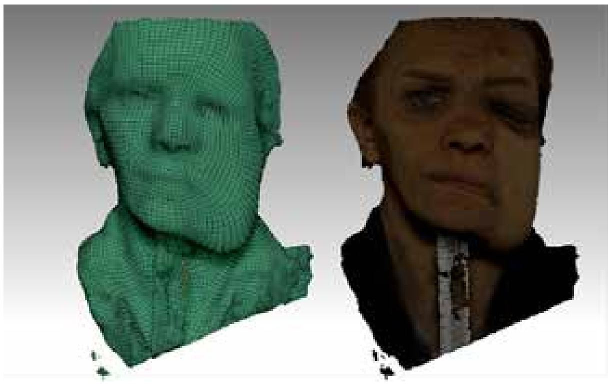


Figure 1. Graphical Software: CATIA V5 R21.

Figure 2. Facial view of the patient. A, Preoperative view, and B, Postoperative view at 6 months after the surgery.

Figure 3. Placement of templates on patient's face during the operation.

Conflict of Interest

The authors declare no conflict of interest.

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