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Case Report

Post Operative Macroglossia after Craniovertebral Junction Surgery in Patient with Achondroplasia

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Abstract

Introduction: Post-operative macroglossia is a rare but devastating complication that can occur during any spinal surgery procedure. Very few cases have been reported regarding development of post-operative macroglossia after spine surgery. This complication can be more troublesome in cases of Achondroplasic patients, who are already at high risk because of pre-existing oral anomalies.

Case Report: Here we present a case of 22 year old Achondroplasic female with pre-existing oral cavity anomalies that develops post-operative life threatening macroglossia, after having undergone revision decompression surgery for craniovertebral junction. Occipto-cervical fixation with foramen Magnum decompression and D10 to L2 posterior fixation with decompression was performed in this patient in prone position with less than 500 ml blood loss in less than 4 hours. After failed conservative management and laser assisted tongue reduction, we had to resort to tracheostomy and shifting from oral endotracheal tube to tracheostomy tube to manage this complication. Additionally, the occipitocervical fixation was changed giving slight extension to reduce the venous pressure.

Conclusion: Patients with pre-existing oral cavity anomalies like down syndrome, achondroplasia etc. are at high risk of developing post-operative macroglossia, especially if they are undergoing craniovertebral junction surgery. In these patients, it is preferable to intubate through nasal route rather than oral route. Also, the length of surgery should be kept as short as possible. If two surgical procedures are required in the same patient, best would be to do them in staged manner on different days. Excessive neck flexion during the procedure should be avoided during positioning and craniovertebral fixation surgery, best is to do it in slight extension or neutral alignment of neck. Use of tongue blocks as a protective measure is also encouraged.

Keywords: Post-Operative Macroglossia; Craniovertebral Junction; Achondroplasia

Introduction

Post-operative macroglossia is a rare but devastating complication that can occur during any spinal surgery procedure, but most commonly has been described after posterior fossa and upper cervical spine surgeries [1]. This complication can be more troublesome in cases of Achondroplasic patients, who are already at high risk of having pre-existing anomalies like macrocephaly, depressed nasal bridge, a short posterior cranial base, maxillary hypoplasia, otolaryngeal system dysfunction [2]. Few case reports have also described pre-existing macroglossia in patients of achondroplasia [3]. In achondroplasia, macroglossia can be absolute or relative or both. Absolute macroglossia means conditions where tongue is increased in size but oral cavity is normal in size. Relative macroglossia (sometimes called as pseudomacroglossia) means conditions where tongue is normal in size but the oral cavity is either small or malformed.

Also, it is a well-known medical fact that patients with achondroplasia have foramen magnum stenosis leading to neurological manifestations [4], which frequently need surgical intervention. Very few case reports of this rare complication of post-operative macrglossia have been described in literature. Here we present a case of 22 year old Achondroplasic female with pre-existing oral cavities anomalies who develops post-operative life threatening macroglossia, after having undergone revision decompression surgery for craniovertebral junction.

Case Report

A 22-year-old Achondroplasic female was admitted under our spine surgery team with complaints of inability to walk due to progressive weakness in both legs and inability to control urine/stool for last 1 year. Two years back, she had undergone posterior cervical C1 laminectomy for established foramen magnum stenosis. Initially she showed neurological improvement after the first surgery but within 6 months of the surgery she started deteriorating again and presented to us in wheelchair with inability to stand/walk or even to use the both upper limbs effectively.

On examination she had findings of typical upper motor neuron lesion consisting of spasticity in all four limbs, increased tone, paraparesis with grade 1/5 in lower limbs and grade 3/5 motor power upper limbs, hyperreflexia, no muscle wasting, upgoing plantar reflex, absent superficial reflexes. She had indwelling urinary catheter due to inability to control urine and was wearing a diaper because of frequent fecal incontinence. She even had loss of cortical sensations and clonus was present in the left ankle. Her higher mental functions and cranial nerve examinations were within normal limits.

As per her previous medical records, dental examination showed that she had a tonsillar hyperplasia with abnormal dentition and mildly depressed nasal bridge.

She was further investigated by means of MRI brain with MRI of whole spine, dynamic x-rays of cervical spine and open mouth views of cervical spine. Her investigation lead us to the conclusion of severe foramen Magnum stenosis with instability with cord signal intensity changes representing long-standing compression with concomitant D11-12, D12-L1 stenosis (Figure 1). She was counseled and planned for occipto-cervical fixation with foramen Magnum decompression and D10 to L2 posterior fixation with decompression. She was accordingly worked up for the surgery by way of blood investigations, x-ray chest, cardiac work up and pre-anesthesia evaluation.

Surgery and management

On the day of surgery the patient was intubated with flexometallic cuffed oral endotracheal tube 7.0 mm ID and the patient was positioned prone with head held in neutral position with Mayfield head holder. A soft bite block was inserted after induction of anaesthesia. Two teams simultaneously worked together and performed occiput to C3 fixation with decompression and D11 to L2 posterior fixation with decompression respectively.

Figure 1: Whole spine MRI showing craniovertebral stenosis and dorsolumbar stenosis with myelomalacia.

The entire surgery was uneventful with less than 500 ml blood loss and was completed within 4 hours. However as soon as the patient was made supine it was observed that she had a huge tongue edema with tongue protruding well outside the oral cavity (Figure 2). After discussions with senior anesthetic it was decided not to extubate the patient but to wait for the tongue swelling to subside within next few days.

Since the tongue was protruding out, it was kept covered in moist gauge pieces, which were changed hourly. However, the tongue swelling did not subside even after 3 days and the patient could not be extubated. Postoperative x-ray and MRI of cervical spine was done which showed satisfactory fixation with neck and neutral alignment (Figure 3). An opinion from ENT surgeon was sought. Direct laryngoscopy revealed massively swollen tongue,

Figure 2: Clinical photograph showing post-operative macroglossia with tongue protruding out of mouth.

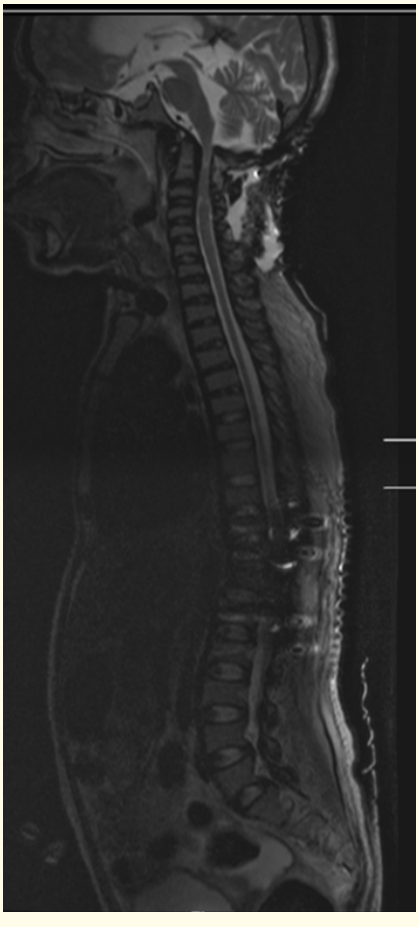

Figure 3: Postoperative MRI showing satisfactory alignment of the spine at Craniovertebral junction and dorsolumbar junction.

but larynx could not be viewed as the view was obstructed by massively swollen tongue. A trial of laser assisted tongue edema reduction was decided.

As decided, CO_2 laser assisted fenestrations made in tongue mucosa and adrenaline and dexona packing put over the tongue. The tongue edema did reduce, but still the tongue could not be put back into the oral cavity, the edema started increasing again after 2 days

After a lot of discussion and family counseling it was decided that it is not a feasible option to keep the patient intubated for so long. Also so there was a possibility of tongue necrosis due to constant pressure from the endotracheal tube.

Finally, it was decided to perform revision occipito-cervical surgery and to fix the neck in slight extension and stabilize it with a HALO brace. At the same sitting, the ENT surgeons performed a tracheostomy. The endotracheal tube was removed and a tracheostomy tube was inserted. In the immediate post-operative period, the tongue could be inserted back into the oral cavity (Figure 4a and 4b).

The patient was kept on ventilator. The tongue swelling gradually improved further and the ventilator support could be taken off on the 2nd post-operative day after gradual weaning. The patient was discharged on air with improved neurology (power in all four

Figure 4a: The tube was changed from oro-tracheal to tracheostomy. Immediately after surgery, the tongue could go inside the oral cavity.

Figure 4b: The tongue size reduced further in 2 days.

limbs 3/5 at the time of discharge). At this time the tongue was still enlarged but was comfortably placed inside the mouth. Patient was still not able to have oral feeds and the nutrition was still given through nasogastric tube.

At 1 month follow-up, the power was almost 4/5 in all four limbs; spasticity was less as compared to before. At the same sitting, ENT team performed a fibreoptic laryngoscopy and changed tracheostomy tube. They found vocal cords were mobile but edematous, arytenoids were edematous, trachea was collapsed and tracheomalacia. There was granulation tissue in stoma. Decannulation trial was given but patient couldn't tolerate because of collapse of trachea. The tongue was almost normal in size with patient able to tolerate oral feeds.

Figure 5: After revising the head position, the patient was kept in halo so that there is no movement at craniovertebral junction.

Discussion and Conclusion

It is a well-known fact that macroglossia is common in conditions like Down syndrome, Beckwith-Wiedemann syndrome, primary amyloidosis, and congenital hypothyroidism [5]. Few of the acquired conditions leading to macroglossia could be trauma, cancer, endocrine disorders or inflammatory or infectious diseases [6]. By definition, severe postoperative macroglossia or SPOM is a massive swelling of the tongue occurring within hours following any operative intervention like posterior fossa surgery or upper cervical spine or craniofacial surgery [3]. The first evidence of postoperative macroglossia was given by McAllister in 1974, where it occurred as a positional complication (sitting position) [7]. SPOM is a potentially life threatening complication and can present intraoperatively with decreasing oxygen saturation, in the immediate post-operative period with tongue protrusion and difficult extubation (as happened in our case) or as a delayed presentation with difficult breathing with decreased oxygen saturation [13].

However recent literature shows that, SPOM is most common after a neurosurgical or craniofacial procedure in high risk individuals like achondroplasia, Down syndrome or endocrinology disorders [8].

There has been extensive work done for investigating the cause of SPOM, but no conclusive cause(s) can be pin pointed. Factors that have been implicated can be classified as (1) local mechanical compression (by teeth, oropharyngeal airway, endotracheal tube, throat pack) interfering with venous or lymphatic drainage of the tongue; (2) regional venous obstruction (excessive neck flexion, head-down position); and (3) various combinations of these two factors. Moore., *et al*. [10] observed that the complication occurred only with posterior fossa surgery and proposed that macroglossia may be neurogenic in origin. As per them, SPOM could be due to abnormal autonomic nervous system discharges to tongue secondary to direct brainstem stimulation during neurosurgical or craniofacial procedure [10].

The lingual arteries run at base of the tongue, deep to the lingual veins that drain into the ipsilateral internal jugular vein or facial vein. In high risk patients the initial congestion of the venous system may lead to local ischaemia due to compression of lingual arteries. This is followed by reperfusion injury. The lymphatics follow the vessels and may play a contributory role. Venous obstruction from excessive flexion of the neck can occur when the patient's head is positioned in flexion either in prone position or in sitting position. In one case, the macroglossia was prevented by monitoring jugular venous pressure during surgery and keeping it negative throughout the surgery [13].

One study which was done by Bouaoud J., *et al*. points out that two most common reasons could be oral intubation in high risk individuals and surgery lasting more than 2 hours [3].

As per Chowdhury T., *et al*. the main reason for SPOM could be mechanical compression of the tongue during the procedure which would impair the venous/lymphatic drainage of the tongue ultimately leading to arterial impedance [9].

Immunological hypothesis behind SPOM were also proposed by few like Chan MT., *et al*. where hypersensitivity reactions secondary to drugs like droperidol and angiotensin-converting enzyme inhibitors word considered to be the culprits [11].

In our case, we feel the reasons for SPOM were a combination of many factors starting from oral intubation to long length of surgery (4 hours), surgical procedure involving foramen Magnum, Achondroplasic patient and C-V junction fixation in flexed position. The patient was already having relative macroglossia due to underlying achondroplasia. Oral intubation occupied the space within the oral cavity and thus leading to external protrusion of tongue. This lead to mechanical compression of veins. Further the flexed posture of the neck due to desired fixation of occipitocervical junction, further added to increased venous pressure leading to venous congestion and ultimately arterial compression and tongue edema. Therefore, when these factors were reversed in the second surgery which included adjusting the position of neck and changing the airway from oral endotracheal intubation to tracheostomy, it led to immediate involution of tongue within oral cavity and progressive decrease in tongue edema.

Literature highlights many treatment options for SPOM like oral toilet, bite blocks, pain relievers or even partial glossectomy [12].

Operative procedures like glossectomy, can be median type (along the mid-line) or peripheral types [12]. In both types, the main Idea behind the surgery is to debulk the tongue by resection of tongue tissue and the subsequently suturing the margins [12].

We feel in the best interest of the patient, it is important that in high risk patients (for SPOM) like down syndrome or achondroplasia or oral malignancies should always get a pre-operative ENT or dental opinion for any pre-existing tongue hyperplasia or maxillofacial anomaly.

Also, the intubation should be always through fibre optic nasal tubes and the length of surgery should be kept as short as possible. If two surgical procedures are required in the same patient best would be to do them in staged manner on different days. Also, excessive neck flexion during the procedure should be avoided during positioning or if C-V junction fixation is required, best is to do it in slight extension or neutral alignment of neck. Use of tongue blocks as a protective measure is also encouraged.

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