Blindness and basal ganglia hypoxia as a complication of Le Fort I osteotomy attributable to hypoplasia of the internal carotid artery: a case report

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Le Fort I osteotomy is used as a surgical procedure for correction of maxillofacial deformities. The common complications of this procedure are hemorrhage and infection, with incidence of 6% to 9%. Blindness associated with Le Fort I osteotomy was reported in 8 patients. An 18-year-old female complained of loss of sight in the left eye after recovery from hypotensive general anesthesia. The visual field of the left eye was dark and only perceived some movement. She presented with motor dysfunction and regressive behavior 2 weeks later as a result of hypoxia of bilateral basal ganglia. Two months later, her visual acuity recovered gradually and regressive behavior improved. Carotid angiography showed congenital hypoplasia of the left internal carotid artery. We suspected that hypoplasia could cause hypoxia of the central nervous system. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;104:e27-e33)

Le Fort I osteotomy is a surgical procedure for correction of maxillofacial deformities.1 Von Langenbeck first described surgery of the maxilla in 1859 for the removal of nasopharyngeal polyps. In 1901 Le Fort published his classic description of the natural planes of maxillary fracture.2 In 1927 Wassmund first described the Le Fort I osteotomy for the correction of midface deformities.3 Schuchardt advocated separation of the pterygomaxillary junction in 1942.4 Today, the Le Fort I osteotomy has become a well-established procedure to correct maxillary protrusion, cleft lip/palate, and for other craniofacial deformities.1 The reported complications associated with these techniques include intraoperative hemorrhage, infection, injuries of peripheral nervous system, airway compression, fistula formation, and bone necrosis.5 The incidence of complications is about 6% to 9%.1 There had been only 8 cases reported about the complications of visual impairment or other injury of the central nervous system related to the Le Fort I osteotomy.6,7 The possible reasons for these complications include ischemia/infarction of the ophthalmic artery as a result of unanticipated fracture, rupture of an ophthalmic aneurysm, or unknown cause.7 As far as we know this is the first report on hypoplasia of unilateral internal carotid artery that could be the main reason for this unforeseen surgical complication of Le Fort I osteotomy.

CASE REPORT
An 18-year-old female had maxillary retrusion and mandibular prognathism (Fig. 1). After 2 years of preoperative orthodontic treatment, she was referred to our department of oral and maxillofacial surgery for orthognathic surgery. She was healthy with no abnormal medical or family history, but occasional dizziness was noted when resting. Le Fort I osteotomy and intraoral vertical ramus osteotomies were performed in August 2003 under hypotensive general anesthesia. The mean arterial blood pressure was maintained between 60 and 70 mm Hg for 8 hours. The lowest mean blood pressure was 52 mm Hg (75/40 mm Hg) for 3 minutes immediately after pterygomaxillary separation. The estimated blood loss was about 1300 mL; 2500 mL of crystalloids and 500 mL of red blood cell concentrate were transfused during the operation. Her laboratory data showed preoperative hemoglobin of 13.3 g/dL with a hematocrit of 36.9%, and postoperative hemoglobin of 8.6 g/dL and hematocrit of 23.3%.

After recovery from general anesthesia, she complained of vision loss in her left eye. Preoperative corrected distance Snellen visual acuity of both eyes was 20/25, with myopia (diopter = 4.00 D). Ophthalmologic examination in the immediate postoperative period showed normal appearance of both eyes (Fig. 2), but absence of light sense in the left eye and left pupil dilatation (5 mm) without direct light reflex.

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Both fundi appeared normal. She was able to perceive hand movement from a 20-cm distance, and the Snellen visual acuity was below 20/1000. Computed tomography of the head demonstrated no abnormal finding such as intracranial/intraorbital hemorrhage, skull base fracture, or orbital fracture (Fig. 3). Magnetic resonance imaging (MRI) of the orbital region also revealed no injury of optic vessels (Fig. 4) and the basal ganglia were normal (Fig. 5). Intravenous administration of 300 mL of mannitol and 70 mg of dexamethasone (1 mg/kg) were given, followed by 500 mL of mannitol every 12 hours and 35 mg of dexamethasone every 6 hours.

Ophthalmology, neurology, and cardiology examinations were done the next morning. The color Doppler examination of the neck suggested reduced flow of the left internal carotid artery, increased flow of the both spinal artery and reverse flow of the left ophthalmic artery. No evidence of intima injury of the left internal carotid artery, and no thrombosis were found. It suggested hypoplasia of this vessel. Two days later, another Doppler examination did not detect any signal from the left internal carotid artery or the left ophthalmic artery, but normal blood flow of the spinal artery was noted. It was unclear why there was no signal from the left internal carotid and the left ophthalmic arteries. Minimal blood flow might not be detected by our color Doppler. Cardiac and orbital ultrasonography and fluorescein angiography (FAG) showed no abnormal finding. Ten days later, the patient developed direct light reflex in the left eye but the visual field was still abnormal. On August 19, the second day after discharge from our hospital, she presented with anxiety, insomnia, and uncooperative and regressive behavior. The nervous disorders similar to Parkinson’s disease, marked by muscular rigidity, tremor, and impaired motor control of tongue, jaw, or extremities, were noted. Extrapyramidal syndrome was observed, including parkinsonism, akathisia, dystonia, and tardive dyskinesia. It was suggested that injury of motor pathways other than the pyramidal tract was caused by hypoxia. After psychiatric consultation, a benzodiazepine drug (1 mg of lormetazepam before sleep) was prescribed. MRI demonstrated hypoxia of bilateral basal ganglia (Fig. 6) and no signal of left internal carotid artery were seen in the magnetic resonance angiogram (Fig. 7). All symptoms recovered gradually (Table I) and a carotid angiography was done on October 20, which confirmed hypoplasia of the left internal carotid artery (Fig. 8). Therefore, we concluded that the congenital hypoplasia of the left carotid artery when subjected to hypotensive anesthesia caused hypoperfusion and hypoxic injury to her optic nerve and the basal ganglia resulting in the visual loss and her psychiatric disorder. She continued to receive postoperative orthodontic treatment and ophthalmologic follow-up. The distance Snellen visual acuity is 20/450 in her left eye and part of the visual field recovered until June 2006, when this report was written.
DISCUSSION

Reported complications associated with Le Fort I osteotomy include intraoperative hemorrhage, infection, injury of peripheral nervous system, airway compression, fistula formation, or bone necrosis. Although visual or basal ganglia impairment are rare in orthognathic surgery, there have been case reports of visual loss in non-ocular procedures such as cardiopulmonary bypass surgery, abdominal surgery, intracranial surgery, spinal surgery, and neck dissection. The actual reasons for these injuries are not clearly known but visual impairment may occur from mechanisms of ischemic optic neuropathy, occlusion of central retinal artery, or cerebral ischemia. Ischemia may be provoked by factors such as atherosclerotic disease, hypertension, collagen disease, temporal arteritis, or diabetes. Hypoxia of basal ganglia is often induced by cardiac arrest or severe hypotension, and it may cause mental-rotation deficits, aphasia, acute movement disorder, and so forth.

The neurologic impairments of our patient are discussed as follows.
Visual impairment

Visual loss after the surgery may be caused by (1) ischemic optic neuropathy (anterior and posterior); (2) injury to the optic nerve by occlusion of the central retinal artery; or (3) damage to the optic area of the brain hemisphere, including pituitary apoplexy, optic tract, optic radiation, and cortex of occipital lobe.\(^{12}\)

The visual transmission travels from the retina, optic nerve, optic chiasm, optic tract, lateral geniculate body, optic radiation, and into the visual cortex in the occipital lobe. If injuries occur after optic chiasm, both eyes will be affected. In this case, the patient only lost her left visual acuity; therefore, initially we suspected that the retina or the optic nerve was injured. However, on ophthalmoscopic examination, the retina appeared normal. The direct pupil reflex of the left eye was negative, and the consensual pupil reflex was negative as well.\(^{13}\) Hence, damage of optic nerve was inferred. Two kinds of damage may occur, namely anterior ischemic optic neuropathy or posterior ischemic optic neuropathy. Anterior ischemic optic neuropathy results from either obstruction of the posterior ciliary artery, which supplies the optic papilla,\(^{14}\) or imbalance between the artery and intraocular pressure, attributable to systemic hypotension.\(^{15,16}\) If only one eye is affected, congestion and/or swelling of optic papilla, found on fundoscopy, may be present in the early stage and severe in the late stage. The direct pupil reflex of the affected eye is sluggish.\(^{17,18}\) Posterior ischemic optic neuropathy results from obstruction of the pial vessels, which come from the collateral arteries arising directly from the ophthalmic artery, supplying the posterior part of optic nerve,\(^{14}\) and it may cause (1) visual field defects sometimes combined with decreased visual acuity, (2) no direct pupil reflex, and (3) no edema or hemorrhage of the optic papilla and the retina of the affected eye.\(^{19}\)

According to the clinical examination, posterior ischemic optic neuropathy was highly suspected in this case. Visual loss is a rare but severe complication in orthognathic surgery. The contributing factors include unanticipated fractures, massive hemorrhage or severe anemia, hypotensive anesthesia, inappropriate pressure on the eyeball, or abnormalities of the internal carotid artery.

Unanticipated fractures. When performing Le Fort I osteotomy, inappropriate separation of the pterygomaxillary junction will result in fractures extending to pterygoid plates, sphenoid bone, orbital floor, optic canal, or the skull base.\(^{6,20-22}\) It will damage the optic nerve or its vascular supply.\(^1\) According to Renick and Symington\(^{23}\) and Robinson and Hendy,\(^{21}\) separation of the pterygomaxillary junction may cause unanticipated fractures in approximately 58% to 75% of cases and these fractures may not be seen in computed tomography. When an unexpected visual impairment occurs, we should consider this kind of problem first\(^24\) and empiric management must be prescribed as soon as possible.

Hemorrhage and anemia. Hemorrhage from the descending palatine artery or sphenopalatine artery in Le Fort I osteotomy may cause systemic hypotension.\(^{24}\) Hemorrhage from the pterygopalatine fossa may enter the orbital cavity through the inferior orbital fissure and compress the globe. Severe anemia will not cause ischemic optic neuropathy until it is combined with hypotension.\(^{25}\) We recommend using the drill to perform horizontal osteotomy of the lateral wall of the maxillary sinus, and the osteotome for pterygomaxillary separation, then manual pulling or wire traction\(^{26,27}\) to down-fracture the maxilla, instead of “disimpaction forceps.” It may prevent unanticipated fracture of pterygoid plates, which will cause massive bleeding from the descending palatine artery, or extend to the orbit.

Hypotensive anesthesia. Hypotensive anesthesia is helpful during a maxillofacial surgery for reduction of blood loss and improving the visibility in the surgical field.\(^6\) The blood flow to the globes may be altered by elevated intraocular pressure or decreased systemic blood pressure. Thus, hypotensive anesthesia may potentially decrease the blood supply to the retina and choroid,\(^{24}\) resulting in embolism of the vessels or infarction of the optic nerve. Is hypotensive anesthesia a risk factor for visual impairment? There have still been no associated reports. According to Brown et al.\(^{25}\) and Schobel et al.,\(^{28}\) the durations of hypotensive anesthesia
for patients who developed visual impairment after surgery were 15 minutes to 2 hours and 40 minutes to 5 hours, respectively. The influence of the duration length was not mentioned in their reports. However, if hypoperfusion, when performing orthognathic surgery or neck dissection, combined with other risk factors such as anemia or vascular disorders, visual loss is likely to occur.\textsuperscript{12}

\textit{Inappropriate pressure to the globes.} Sometimes rigid contact lenses are used to protect eyes during maxillofacial surgery, but this device may hamper the normal movement and expansion of the eyelids when edema occurs, and an undue amount of external pressure conducting to the ocular globe would result in increased intraocular pressure and visual damage eventually.\textsuperscript{30} The blood flow to the optic papilla was affected by the pressure gradient between the posterior ciliary artery and the intraocular pressure. Either elevated intraocular pressure or decreased systemic blood pressure may cause insufficient perfusion to the optic nerve.\textsuperscript{16}

\textit{Pathology of internal carotid artery.} Congenital abnormalities of the internal carotid artery include missing and hypoplasia, that underdevelopment part of this artery. The incidence of abnormalities is about 0.01\%. The statistics, until 1987, showed 20 cases of hypopla-
sia, 24 cases of missing, and 19 cases of them combined with aneurysm.31 Until 2001, fewer than 100 cases of abnormalities of internal carotid artery were reported. Because of compensation from the contralateral internal carotid artery or vertebral arteries, the patients with congenital abnormalities of internal carotid artery seldom have symptoms.32 Hypoxic change in the areas supplied by this abnormal artery, however, tends to occur spontaneously33 or when systemic hypotension or cerebrovascular accident happen.12

**Hypoxia of basal ganglia**

Hypoxia of basal ganglia may be the main reason for neuropsychiatric and motor disorders in this case. Basal ganglia act with an adjusting characteristic at cortical-striatal-thalamo-cortical circuits that connect motor function of the cerebral cortex and the behavior/emotional center of the limbic system (Fig. 9).34 As a result, pathological change of basal ganglia will cause motor disorders, e.g., extrapyramidal syndrome and emotional abnormality. Hypoxia of basal ganglia may also be induced by heart arrest, severe hypotension, or suffocation.35 Basal ganglia are easily injured by hypoxia because of the following:

1. cortical-striatal-thalamo-cortical circuits will produce a large amount of glutamate, when hypoxic, that will damage the basal ganglia;35
2. the vessel supplying the basal ganglia is the lateral lenticulostriate artery, which is long and narrow;8 hypoxia also affects the internal capsule and optic radiation results in visual impairment;
3. hypoxia may increase the permeability of vessels in basal ganglia, then reperfusion injury will occur.36

**SUMMARY**

Congenital abnormality of the internal carotid artery is extremely rare. There have been fewer than 100 cases reported worldwide so far. These patients usually have no symptoms. The abnormalities are usually discovered because of temporary brain ischemia caused by intracranial hemorrhage or cerebrovascular accident. When orthognathic surgery with hypotensive anesthesia is planned and the patient has experience with altered consciousness or syncope in past years, a Doppler examination of carotid arteries should be performed before orthodontic treatment. Proper surgical technique should help avoid unanticipated skull base fractures. In cases where there is evidence of compromised carotid blood flow, hypotensive anesthesia may be contraindicated.

**REFERENCE**


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