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CASE REPORT
Craniofacial/Pediatric

A Case of Orthognathic Surgery for Jaw Deformity in a Patient with Spinocerebellar Ataxia

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Summary: Spinocerebellar ataxia (SCA) is a progressive neurodegenerative disease that can cause various ataxia symptoms. Here we report a patient with spinocerebellar ataxia who underwent orthognathic surgery to correct a mandibular protrusion with facial asymmetry. A 33-year-old woman was admitted to our hospital for orthognathic surgery. She started preoperative orthodontic treatment after a diagnosis of mandibular protrusion with facial asymmetry. Two and a half years later, after completing preoperative orthodontic treatment, she returned to our hospital after being diagnosed with spinocerebellar ataxia. After discussing the risk of surgery with the anesthesiologist and neurologist, we elected to perform orthognathic surgery after the patient provided informed consent. Sagittal split ramus osteotomy and intraoral vertical ramus osteotomy were performed under general anesthesia, but no remarkable perioperative complications occurred. After a 3-year follow-up, the occlusion has remained stable, and no postoperative relapse occurred. Whether we should provide surgical treatment for SCA patients is controversial. However, when long-term predictions were considered, altering an occlusion could improve a patient's quality of life in the present case. *(Plast Reconstr Surg Glob Open 2022;10:e4257; doi: 10.1097/GOX.0000000000004257; Published online 13 April 2022.)*

Spinocerebellar ataxia (SCA) is an autosomal-dominant inherited neurodegenerative disorder characterized by various slowly progressive symptoms of ataxia, such as ataxic gait and dysarthria.[1] Although some patients with SCA have undergone surgery under general anesthesia, orthognathic surgery in patients with SCA is unreported. Here, we report a patient with SCA type 3 (SCA3) who underwent orthognathic surgery to correct a mandibular protrusion with facial asymmetry.

CASE REPORT

A 36-year-old female patient was admitted to our hospital for orthognathic surgery. Extraoral examination revealed a concave profile and facial asymmetry. Intraoral examination revealed a class III malocclusion with an excessive negative overbite (−1 mm) and overjet (−1 mm) (Fig. 1). Mandibular midline deviated to the right. Cephalometric analysis revealed a class III skeletal relationship due to increased mandibular growth. The lateral inclination of the occlusal plane was not observed. Based on these findings, we diagnosed the patient with a skeletal class III malocclusion due to increased mandibular growth.

The patient presented to the department of otorhinolaryngology at our hospital because her symptom of dysarthria did not improve despite presurgical orthodontic treatment. Next, she came to the department of neurology at our hospital for suspected central nervous system disease. The patient underwent physical examination, neurological examination, brain magnetic resonance imaging, neuropsychological examination, and genetic analysis. Eventually, she was diagnosed with SCA3.

Neither the orthodontist nor our department knew about her SCA3 diagnosis during orthodontic preparation. When the patient presented to us again to plan orthognathic surgery after 2 years and 5 months, she informed us of her SCA3 diagnosis. According to an intraoral examination after presurgical orthodontic treatment, the mandibular dental midline deviated 3.5 mm to the right, and the overjet and overbite were −5 mm and 0 mm, respectively. After presenting the case to the neurologist and anesthesiologist, we discussed all the details of the surgery and suspected complications with the patient.

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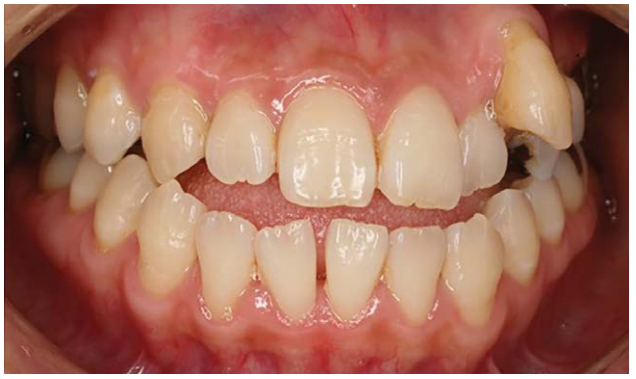

Fig. 1. Intraoral finding at the initial visit: intraoral photographs showed a class III malocclusion with excessive negative over bite (−1 mm) and overjet (−1 mm). There was deviation of the mandibular center to the left, maxillary center to the left.

As she still wanted to undergo orthodontic surgery, she provided written informed consent, and we scheduled an orthognathic surgery. In April 2015, the patient underwent right mandibular intraoral vertical ramus osteotomy and left mandibular sagittal split ramus osteotomy under general anesthesia. Induction was completed with propofol and remifentanil. Rocuronium bromide was added after airway was provided. For maintenance, desflurane 3%, oxygen and air were used. Sugammadex was administered after the operation was completed. According to the model surgery, we set back the mandible asymmetrically 4 mm on the right side, and 8 mm on the left side. Elastic traction was used for postoperative occlusal stabilization. Mastering the exchange of elastics required more time than healthy patients, but she could perform exchange alone. Three years after surgery at the last follow-up, the occlusion was stable with a harmonious skeletal relationship and pleasing profile (Figs. 2, 3).

DISCUSSION

Spinocerebellar ataxia type 3 (SCA3) is an autosomal-dominant inherited neurodegenerative disorder. It is one of the most common forms of SCA, especially in Japan.[2] The patient was diagnosed with SCA3 based on genetic testing in the present case. Various symptoms are associated with SCA, including gait instability, dystonia, difficulty with fine motor skills caused by cerebellar ataxia, and pyramidal and extrapyramidal signs. Severe dysarthria and dysphagia may also occur in advanced disease stages.[3] In this case, at the time of surgery, although the patient experienced mild dysarthria and mild limb ataxia with lower predominance, she maintained gait autonomy, and she was able to communicate with us, similar to other healthy patients.

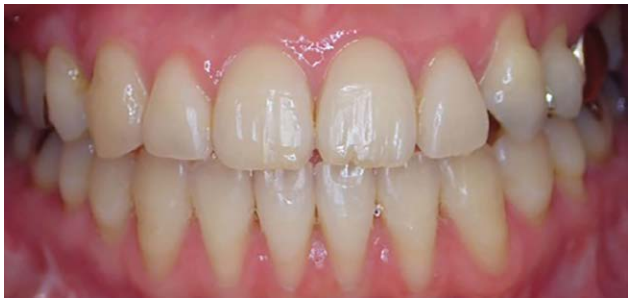

Fig. 2. Postoperative finding: intraoral photographs indicated class I canine and molar relationships were obtained.

Although some case reports describe surgery under general anesthesia in patients with SCA,[4] only one case of surgery under general anesthesia in a patient with SCA3 has been reported.[5] Moreover, orthognathic surgery has not been reported in any cases. Regarding orthognathic surgery for SCA3 patients, we should consider two major risks. One risk is general anesthesia management, while the second is the orthognathic treatment process management. In terms of anesthesia risk, airway stenosis can occur and worsen after general anesthesia in SCA3 patients due to respiratory muscle weakness, vocal cord paralysis, and aspiration. This condition can appear more intense in SCA3 patients with existing peripheral denervation. Consequently, airway evaluation is crucial during the perioperative period, especially immediately after extubation. In the present case, fiberscoptic laryngeal evaluation did not reveal impairment of the vocal cords before surgery, even immediately after extubation. Moreover, we used train-of-four monitoring to avoid residual neuromuscular block. In this case, the patient had no acute onset or exacerbation of symptoms after anesthesia with desflurane.

Moreover, circulatory variables, such as hypotension, bradycardia, and atrioventricular blocks, can occur in SCA3 patients due to dysautonomia.[6] Thus, a percutaneous radial arterial catheter was inserted to monitor hemodynamics accurately. Some previous publications reported that local anesthesia could be safer than general anesthesia in SCA3 patients[7] because of perioperative airway complications and circulatory risks. In this case, the patient did not have bulbar palsy, peripheral denervation, or dysautonomia, and therefore, we elected to administer general anesthesia after discussing every possible risk with the neurologist and anesthesiologist. Actually, no airway complications or circulatory instability occurred during the perioperative period.

In terms of considerations in the management of orthognathic treatment, we should consider the severity and speed of symptom evolution in SCA, including clumsiness, slurred speech, and gait ataxia. The treatment period for jaw deformity ranges from 3 to 5 years, including preoperative treatment and postoperative follow-up. Thus, the patient might experience difficulty in returning to the hospital and continuing follow-up due to a decline in activities of daily living. Although SCA progresses slowly, clinical symptoms including gait abnormality, difficulty performing fine motor skills, dysphagia, and respiratory muscle weakness appear gradually. Weakness in the perioral and tongue muscles might cause dysphagia and narrowing of the dental arch, open bite, and postoperative relapse. Therefore, appropriate information, including any potential complications and long-term prognosis, should be provided so that patients can make informed decisions.

Right intraoral vertical ramus osteotomy and left sagittal split ramus osteotomy were appropriate to surgically

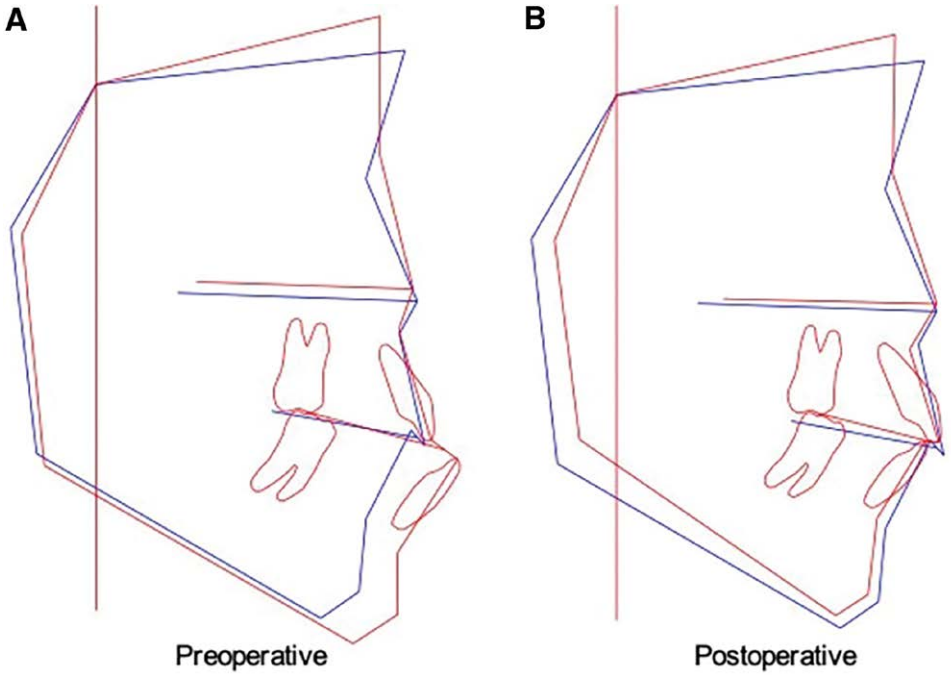

Fig. 3. Preoperative and postoperative profilograms. The posttreatment analysis revealed backward movement of the mandible (A, B). Blue line: age- and sex-matched Japanese norm. Red line: present case.

correct the patient's dentofacial deformity in the present case because the patient had TMJ clicking on the right side, and the setback was not large (4 mm). Intraoral vertical ramus osteotomy on the deviated side was also considered a technique to prevent condylar torque. In some cases, we should choose double jaw surgery depending on the setback and necessity of maxillary movement. However, we should consider that double jaw surgery has a higher risk of postsurgical airway complications in such cases and is associated with a more invasive approach due to longer operative time and more bleeding than single jaw surgery. Moreover, we predicted airway changes after mandibular setback using simulation software. Accordingly, the mandible setback of 4–6 mm can be done safely even if we take the airway space into consideration.

For these reasons, we chose mandible setback. Therefore, the choice of surgical procedure requires judicious attention in accordance with the patient's needs.

The treatment plan for patients with progressive degenerative diseases, such as SCA, is one of the most controversial issues. We performed orthognathic surgery after discussing the SCA stage, possible complications during the entire perioperative period, and the long-term prognosis with the neurologist, anesthesiologist, and patient. In conclusion, the treatment plan positively impacted patient satisfaction and quality of life, as aesthetics, oral function, and self-confidence were improved.

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